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Cardiology in the Young

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Fetal Cardiology

YIA-01
Postinterventional fetal aortic regurgitation: Good or bad?

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Background and Aim: Aortic valve regurgitation (AR) may occur in fetuses with critical aortic stenosis (CAS) after a successful effective aortic valvuloplasty (AV). Prenatal improvement of AR has been reported, but not systematically studied. The aim of this study is to assess the postinterventional incidence of AR in fetuses with CAS, the degree of in-utero aortic valve remodeling and its effect on prenatal LV growth.
Method: This is a retrospective study of all fetuses who underwent AV due to CAS at our center. Only fetuses with an available postnatal echocardiogram were included. Echocardiograms were reviewed on the first or second post-interventional day and the day of birth for AR severity, LV size and function and related to the balloon to aortic valve ratio. The degree of AR was analyzed by two experienced observers and defined as none/trivial, mild, moderate or severe based on the duration and extent of the AR jet and amount of retrograde flow in the aortic arch.
Results: Between 2001 and October 2023, 135 fetuses underwent 165 FAV at our institution. Postinterventional and postnatal echocardiograms were available for 61 patients. The incidence of AR is shown in Table 1. Fifty two fetuses (85%) showed variable degrees of AR directly after FAV. Balloon/valve ratios were significantly higher in moderate/severe AR fetuses (median: 1,09 (0.81-1.23) vs. 1,00 (0.85-1.18), p=0.0003). At birth, the number of patients showing some degree of AR decreased to 52% (p<0.0001). Fetuses with moderate/severe AR directly after FAV showed improved relative LV growth until birth compared to no/mild AR fetuses (median LV length-gain 23.2% vs. 11.6%, p=0.040).
Conclusions: Fetal AV led to AR in most patients. Higher degrees of AR were associated with larger balloon to aortic valve ratios and were well tolerated. Significant improvement of AR severity up to complete resolution is possible in prenatal life. More than mild AR was associated with improved LV development. Care should be taken to choose a large enough balloon (>=110% of the valve diameter) to effectively dilate the aortic valve. More than mild AR seems to be beneficial without precluding an eventual postnatal univentricular pathway.

Table 1: Aortic Valve Regurgitation after Fetal Aortic Valvuloplasty

	After FAV	At Birth	P
All	52/61	32/61	<0.0001
Severe AR	15	3	0.0011
Moderate AR	13	7	
Mild AR	24	22	

FAV: Fetal Aortic Valvuloplasty
AR: Aortic Valve Regurgitation

Keywords: Fetal Cardiac Interventions, Fetal Aortic Valvuloplasty, Critical Aortic Stenosis, Aortic Regurgitation

Congenital Heart Surgery

YIA-02
Central venous pressure augmentation against abdominal compression as a novel marker for cardiovascular load in the fontan circulation

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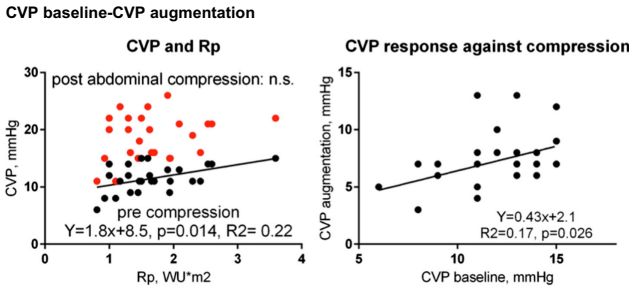
Background and Aim: Although augmentation of central venous pressure (CVP) by volume loading is highly variable in the patients with Fontan circulation, the mechanism is not clearly understood. Since renin-angiotensin-aldosterone system (RAAS) is highly activated and it closely relate with the serum markers for cardiovascular fibrosis in the Fontan circulation, we hypothesized that reactive augmentation of CVP is affected by circulating blood volume and activation of RAAS.
Method: Twenty-nine patients who underwent Fontan procedure more than 10 years ago were enrolled in this study (Age; mean



Marked increase of CVP against abdominal compression was observed in the patients with higher baseline CVP

16.2, 12.7-22.9 years). During cardiac catheterization, ventricular pressure, systemic blood pressure and CVP were simultaneously monitored during abdominal compression procedure for 5 second. Plasma volume was directly measured using dye dilution method. Hemodynamic change during abdominal compression and baseline hemodynamic property, plasma volume, RAAS activity as well as serum markers of cardiovascular fibrosis.
Results: The degree of CVP augmentation against abdominal compression was positively correlated with the baseline CVP (p=0.026). While the CVP augmentation was independent of pulmonary resistance, it was positively correlated with the plasma volume (p=0.0030). Interestingly, the CVP augmentation was also positively correlated with the plasma renin activity (p=0.0060), serum aldosterone level (p=0.011) and type IV collagen 7s (p=0.0074), suggesting close interaction with cardiovascular remodeling in the Fontan circulation. CVP augmentation was independent of EDP or blood pressure augmentations.
Conclusions: CVP augmentation against abdominal compression was influenced by the RAAS activation and its resultant modification of circulating plasma volume as well as cardiovascular fibrosis, suggesting CVP augmentation as the marker for venous adaptation to the Fontan circulation. The relation between CVP augmentation and outcome of Fontan circulation would be subject for future study.

Keywords: Fontan circulation, long term, cardiovascular remodeling, central venous pressure

General Cardiology

YIA-03
Red blood cell distribution width and platelet distribution width as novel biomarkers for fontan hemodynamics and associated end organ damage

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Background and Aim: Circulating blood cell count and cellular size are under homeostasis of biosystem. Red blood cell distribution width (RDW) and platelet cell distribution width (PDW) are known as surrogate markers for heart failure and microcirculation, respectively. Since the venous congestion coupled with reduced cardiac output are the common features of Fontan circulation and heart failure, RDW and PDW could be the marker for Fontan Failure. We tested our hypothesis that the RDW and PDW are the biological marker for Fontan circulation and related organ dysfunction.
Method: The consecutive 134 patients with Fontan circulation who were performed cardiac catheterization were enrolled in this study. The relationships between RDW, PDW and hemodynamic index as well as markers for heart failure, and end organ dysfunction was analyzed.
Results: RDW was positively correlated with central venous pressure (p<0.001) and pulmonary wedge pressure (p=0.002), suggesting unfavorable Fontan hemodynamics. Although higher cardiac index and lower systemic vascular resistance were also correlated with higher RDW, it was considered as the result of vasodilation often observed in the unfavorable Fontan circulation, since RDW was closely correlated with elevated serum levels of procollagen type III peptide (p=0.0024), type IV collagen 7s (p=0.001), the markers for hepatic fibrogenesis, even after adjusting for age. In sharp contrast with the RDW, PDW was independent of age, hemodynamic index and natriuretic peptides, but was positively correlated with liver fibrosis index Fib-4 index (p<0.001), total bilirubin (p=0.023), and albumin/IgG ratio. (p=0.0085). Interestingly, PDW was positively correlated with hepatic venous oxygen saturation (p=0.0054) and serum manganese level (p=0.027), and negatively correlated with hepatic venous wedge pressure (p=0.022), indicating that PDW was likely to be associated with resultant progression of portosystemic shunt originating from hepatic fibrosis.
Conclusions: RDW reflected venous congestion and related fibrogenesis in the Fontan patients. In sharp contrast, PDW appeared to have indicated Fontan related liver dysfunction. The pathologic interaction among PDW, platelet production and hepatic dysfunction needs to be elucidated in the future.

Keywords: Fontan circulation, biomarker, organ dysfunction, Fontan associated liver dysfunction.

Genetics, Basic Science and Myocardial Disease

YIA-04
Impaired relaxation in induced pluripotent stem cell-derived cardiomyocytes with pathogenic TNNI3 mutation of pediatric restrictive cardiomyopathy

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Background and Aim: Restrictive cardiomyopathy (RCM) is characterized by impaired diastolic function with preserved ventricular contraction. Several pathogenic variants in sarcomere genes, including TNNI3 are reported to cause Ca2+ hypersensitivity in cardiomyocytes (CMs) in overexpression models; however, the pathophysiology of induced pluripotent stem cell (iPSC)-derived CMs specific to a patient with RCM remains unknown.
Method: We established an iPSC line from a pediatric patient with RCM and a heterozygous TNNI3 missense variant R170W. We conducted genome editing via CRISPR/Cas9 technology to establish an isogenic correction line harboring wild-type

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TNNI3 as well as a homozygous TNNI3-R170W. iPSCs were then differentiated to CMs to compare their cellular physiological, structural, and transcriptomic features.
Results: CMs differentiated from heterozygous and homozygous TNNI3-R170W iPSC lines demonstrated impaired diastolic function in cell motion analyses as compared with that in CMs derived from isogenic-corrected iPSCs and three independent healthy iPSC lines. The intracellular Ca2+ oscillation and immunocytochemistry of troponin I were not significantly affected in RCM CMs with either heterozygous or homozygous TNNI3-R170W. Electron microscopy showed that the myofibril and mitochondrial structures appeared to be unaffected. RNA sequencing revealed that pathways associated with muscle development and structures were altered in RCM-iPSC-derived CMs.
Conclusions: Patient-specific iPSC-derived CMs could effectively represent the diastolic dysfunction of RCM. Myofibril structures including troponin I remained unaffected in the monolayer culture system, although gene expression profiles associated with muscle development were altered.

Keywords: Restrictive cardiomyopathy, iPSC, isogenic, motion analysis, diastolic dysfunction, cardiomyocytes

Cardiac Dysrhythmias and Electrophysiology

YIA-05
Radiofrequency ablation vs cryoablation of right atrial lateral accessory pathway in pediatric age: Acute and mid-term safety and efficacy profile

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Background and Aim: In literature, transcatheter radiofrequency ablation (RFA) of right lateral accessory pathways (APs) in the pediatric population results in 92-95% of acute efficacy, with about 72% of long-term success rate. Conversely, cryoablation shows excellent acute success (97%), although with a 20% recurrence rate. This study sought to determine acute and long-term success rate, along with predictors of recurrences, in right lateral APs ablation carried out with RF versus cryoenergy approach.
Method: Datas from 53 consecutive pediatric patients who underwent RFA or cryoenergy ablation of right lateral APs between January 2020 and April 2022 at Bambino Gesù Children Hospital were analysed. APs were grouped into: right lateral (RL group), right antero-lateral (RAL group) and right postero-lateral (RPL group).
Results: Out of 53 pts with at least 1 AP [57% males, mean age 12.16 ± 2.64 years, median weight 47 kg (IQR 36-59), height 155 cm (IQR 144 – 164), BSA 1.44mq (IQR 1.18-1.65)] all but one with Ebstein anomaly had structurally normal heart. Twenty-five (47.2%) belonged to the RL group, 14 (26.4%) to the RAL group, and remainders to the RPL group. Radiofrequency ablation was performed in 36 cases (68%), with a 86% acute success rate: 94.5% in the RL group, 100% in the RAL group, and 66.7% in the RPL group, respectively. Cryoablation was performed in 17 pts (32%) with a 82% acute success rate: 85.7% in the RL group, 87.5% in the RAL group, and 50% in the RPL group, respectively. Even including multiple redo procedures, over follow up (mean 28 ± 25 months) recurrences occured in 22% of RF cases (5 pts with RL APs, and 3 pts with RPL APs) vs 12% of cryoablations (2 pts with RPL APs). Median fluoroscopy time was 0.4 min (IQR 0.1-3.4) in RF vs 0.6 min (IQR 0.0-3.1) in cryoenergy,
Conclusions: Transcatheter ablation of right atrial APs shows higher success rate for lateral and antero-lateral localizations. This study confirms that right lateral APs ablation is feasible safe and effective in the pediatric population, regardless of the type of energy and vascular access chosen.

Keywords: right atrial lateral accessory pathway, transcatheter radiofrequency ablation, cryoablation

Cardiac Imaging

YIA-06
Natural shear wave elastography in fontan patients: a new tool for non-invasive assessment of diastolic function?

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Background and Aim: Non-invasive evaluation of diastolic function in Fontan patients is challenging since conventional parameters are not reliable. Ventricular compliance is known to have a direct impact on ventricular filling and therefore on diastolic function. Chamber compliance, in turn, is directly related to myocardial stiffness. The operational stiffness of the myocardium can non-invasively be assessed using shear wave elastography (SWE), a promising new echocardiographic modality based on measuring the velocity of naturally occurring shear waves using high frame imaging. SWE thus offers a novel tool to study diastolic function. The aims of this study are: to assess feasibility of SWE in Fontan hearts; to document shear wave (SW) speed after atrioventricular valve closure (AVVC); to consider whether SW speed shows correlation with filling pressures.
Method: We enrolled 47 Fontan patients (mean age 19 ± 11 years, range 3-46y). High frame rate parasternal long-axis views were acquired using an experimental scanner (1367 ± 270 frame/s). Images were processed offline by extracting tissue Doppler acceleration coded M-modes drawn in the middle of the wall related to the main atrioventricular valve and the outflow valve. We also collected records of pressure in the cavo-pulmonary conduit from recent heart catheterization or from peripheral intra-venous line.
Results: Measurement of SW speed after AVVC was feasible in 97% of patients. Average SW velocity was significatively higher than previously collected data from age-matched healthy volunteers (5.3 ± 1.5 m/s versus 3.54 ± 0.93 m/s, $p < 0.001$). There was no correlation between SW velocity and age ($r=0.092$, $p=0.54$) or association with underlying anatomy ($p=0.68$). Shear waves velocities after AVVC showed a good correlation with pressure in the cavo-pulmonary conduit ($r=0.55$, $p=0.002$), while no other conventional echo parameter showed correlation with filling pressures.

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Figure 1.

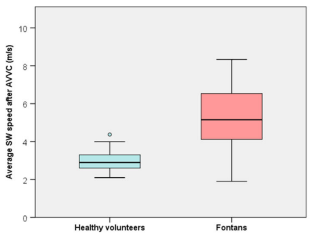

Fig. 1. Values of natural SW speed from Fontan patients are compared with healthy volunteers.

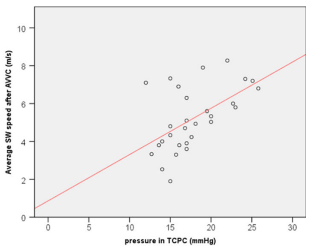

Fig. 2. Correlation between pressure in the TCPC and average SW speed after atrioventricular valve closure, end-diastole

Conclusions: Our findings show that measurements of natural SW is feasible in Fontan hearts. Their myocardium appears stiffer than normal. Whether this depends on an alteration of intrinsic myocardial properties (fibrosis) and/or on preload conditions, remains to be understood. Our data indicate that shear wave elastography may become a useful tool for non-invasive assessment of diastolic function in univentricular hearts.

Keywords: Fontan, Diastolic function, Natural Shear Wave Elastography

OP-001
CMR reference values in children and adolescents for global function index (GFI) and myocardial contraction fraction (MCF)

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Background and Aim: Cardiovascular magnetic resonance (CMR) imaging is becoming increasingly important in diagnostics and follow-up assessment of children with heart disease. Global function index (GFI) and myocardial contraction fraction (MCF) were identified as useful imaging markers to assess left ventricular (LV) cardiac performance and can provide prognostic information for several cardiac diseases. However, reference values for children and adolescents are lacking. Therefore, the aim of this retrospective study was to establish paediatric reference values for MCF and GFI.
Method: 155 CMR examinations of healthy children and adolescents (4-18 years) from two centers in the United Kingdom and Germany were included. LV end-diastolic, end-systolic and stroke volumes (LVEDV, LVESV, LVSV), ejection fraction and myocardial mass (LVMM) were measured using CMR short axis stacks. Results were used to calculate GFI and MCF by the following formulas:
-GFI=[(LVEDV-LVESV)/(1/2*(LVEDV+LVESV))+(LVMM/1.05))*100]-MCF=(LVSV/(LVMM/1.05))*100
Results: The mean age of the subjects was 13.8±2.8 years, 103 of them were male (66%). The mean values for GFI was 46.4±6.1% and for myocardial contraction fraction it was 111.0±20.5 %. Mean LVEF was 57.3±5.7%. Both, GFI and MCF decreased significantly with age (GFI: r=-0.29, p=0.0003; MCF: -0.28, p=0.0005). There was no difference between girls and boys for GFI and MCF (GFI: 46.8±5.2% vs. 46.2±6.5%; MCF: 113.7 ±20.6% vs. 109.6±20.4%). Strong correlations between GFI and MCF (r=0.79, p<0.001) as well as between GFI and LVEF (r=0.80, p<0.001) were documented whereas the correlation with MCF and LVEF was weaker (r= 0.33, p<0.001). Normal distributions for different age groups were assessed.
Conclusions: We provide CMR reference values for the new cardiac functional markers GFI and MCF in children and adolescents. They can serve to improve interpreting clinical CMR studies and can be used for future research studies.

Keywords: Cardivascular magnetic resonance imaging, reference values, global function index, myocardial contraction fraction

OP-002
Right ventricle function responses to acute exercise in adolescent athletes, pulmonary hypertension and tetralogy of fallot

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Background and Aim: Exercise stress echocardiography (ESE) is a valuable, but underutilised tool in assessing right ventricular (RV) function. We aim to evaluate the role of ESE in characterising RV function reserve in three groups with varying volume and/or pressure overload.
Method: Adolescent athletes (n=51), pulmonary arterial hypertension (PAH, n=11) and tetralogy of Fallot (ToF, n=25) patients were included, after matching for age and resting heart rate (HR). All participants underwent ESE (modified Bruce protocol), at age and fitness adjusted increments (20 to 50 W). Right ventricular free wall longitudinal strain (RVFW-Sl), peak systolic (RV-S') and diastolic (RV-E') longitudinal tricuspid annular velocity were measured. Exercise intensity was classified based on domains according to percentage from peak HR into low (<35%), moderate (<69%), high (<88%) and severe (<89%), and by absolute HR (continuous). The relationship between RV function and exercise intensity was analysed using random effects linear mixed models.
Results: Mean peak exercise parameters were as follows (athlete vs ToF vs PAH): oxygen consumption (43 ± 7 vs 37.8 ± 8.3 vs 19.7 ± 5.6 mL·min-1·kg-1), work-rate (21640 vs 102 ± 39 vs 69 ± 19 W) and HR (184 ± 12 vs 180 ± 11 vs 154 ± 23 bpm). Compared to athletes, both PAH and ToF showed overall reduced RV function reserve, worse at higher intensity exercise (Figure). The relationship between RVFW-Sl and HR in ToF showed lower baseline values, but a similar slope to athletes during exercise (Panel A, B). When compared to ToF, PAH showed an abnormal relationship between RVFW-Sl and HR, with dysfunction during

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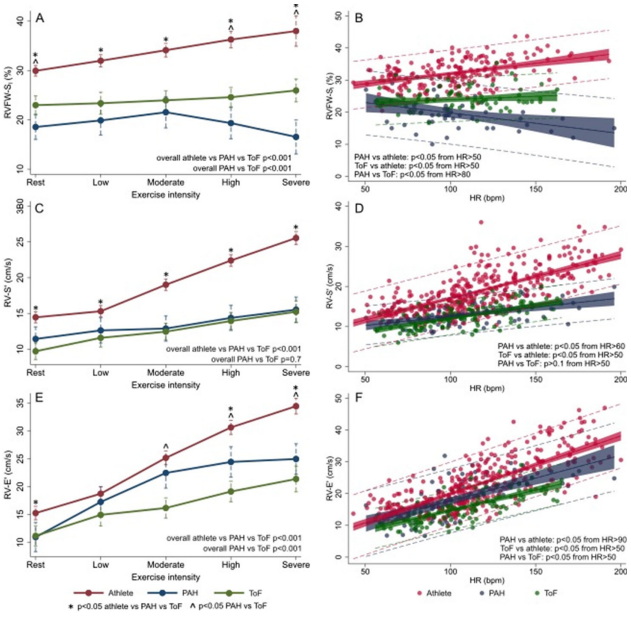
Figure



Relationship between right ventricle (RV) function and exercise intensity domain and heart rate (HR) exercise response, in athletes (red), pulmonary hypertension (PAH, blue) and tetralogy of Fallot (ToF, green). Panels A, C, E show linear mixed model (exercise intensity as fixed effect, participant cluster as random effect) margin estimates with 95% confidence intervals. Panels B, D, F show scatterplots of cardiac function parameters over HR (coloured dots), with linear regression best fit line (solid-coloured lines), 95% CI of linear prediction (coloured area) and 95% CI of linear forecast (dashed coloured lines, p values from linear mixed model (HR as fixed effect full interaction with group fixed effect variable, participant as random effect)

exercise (Panel A, B), but no differences observed in RV-S' (Panel C, D). ToF appears to have the worse diastolic function response among the three groups, in relation to exercise intensity domains and HR (Panel E, F).

Conclusions: ESE can be used to characterise RV reserve in detail and can detect subclinical changes such as systolic dysfunction in PAH or reduced diastolic reserve in ToF. RV assessment during ESE should be multimodal, speckle tracking echocardiography and tissue Doppler imaging being useful complementary tools for evaluating RV reserve.

Keywords: exercise testing, pulmonary hypertension, athletes, tetralogy of Fallot, ventricular strain, cardiac reserve

OP-003

Diastolic myocardial stiffness assessed by shear wave elastography in children with a fontan circulation compared to age-matched healthy volunteers

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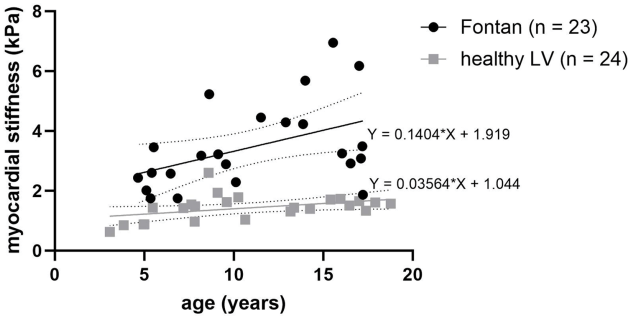
Background and Aim: Increased ventricular filling pressures have been associated to poor outcome after the Fontan palliation. Currently, there are no reliable non-invasive methods to assess diastolic function in Fontan patients. Using shear wave elastography,

Figure 1



Relationship between age and myocardial stiffness in both Fontan and healthy control LV's.

the aim of our study is to quantify the diastolic myocardial stiffness in Fontan population and to compare it to healthy controls.

Method: Cross-sectional study of pediatric Fontan patients (<18 year) presenting at the echocardiography lab. All patients underwent acoustic radiation force-induced shear wave imaging, to assess diastolic myocardial stiffness. Additionally a full functional echocardiogram was performed. Parameters from exercise testing, diagnostic catheterization and cMRI (within the last 3 years) were recorded retrospectively. Control subjects were age- and sex-matched from a database with children with normal hearts.

Results: 24 patients with a Fontan circulation and 24 controls were included. Diastolic myocardial stiffness was increased in Fontan patients (median 3,17kPa (IQR 2,44-4,29) versus in healthy left ventricles (median 1.46 kPa (IQR 1.17-1.64; p<0.001). The increase with age was more steep in Fontans versus in healthy controls (Figure 1). There was no significant difference in myocardial stiffness between single left, single right or 2 inseparable ventricles (p=0.263). Myocardial stiffness did not correlate to invasively measured ventricular end-diastolic pressure (n = 12; range 5-13 mmHg), extracellular volume % or indexed ventricular volume as measured by cMRI (n = 11) or any exercise testing parameter (n = 10). It did not correlate either to inflow doppler signals or tissue doppler velocities.

Conclusions: Pediatric patients with a Fontan circulation have increased diastolic myocardial stiffness, even if they do not have increased invasively measured resting ventricular filling pressures. Conventional parameters to assess diastolic function did not correlate with myocardial stiffness in our population. Future studies will need to determine the prognostic value of increased myocardial stiffness in Fontan patients.

Keywords: myocardial stiffness, Fontan

OP-004

Distinct radiomics feature signatures in LGE sequences of children and adolescents with myocarditis – results from the multicenter registry "MYKKE "

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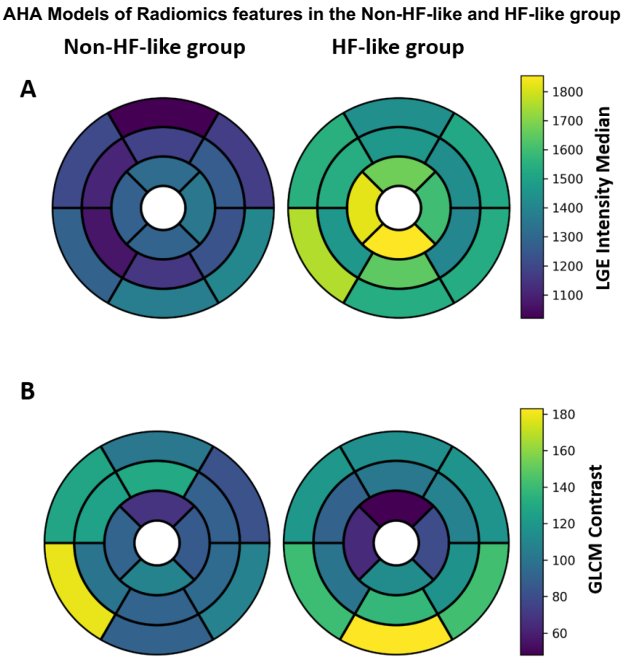
Background and Aim: Pediatric myocarditis can present with asymptomatic courses up to severe heart failure. Endomyocardial biopsy (EMB) remains the diagnostic gold standard, but cardiac magnetic resonance imaging (CMR) with its mapping methods and late gadolinium enhancement (LGE) is becoming increasingly important as a non-invasive diagnostic tool. In this context, radiomics tools enable a quantitative characterization of LGE intensity and texture patterns. We aimed to characterize pediatric patients, enrolled in the multicenter registry for myocarditis "MYKKE" using radiomics features.

Method: Short axis LGE sequences of patients with EMB-proven myocarditis were analyzed. Epi- and endocardial borders were detected automatically, corrected by a radiologist, and divided according to the AHA model. To account for different anatomical size, field of view and intensities in LGE images, a resampling and normalization on blood pool and myocardium was performed per patient. Fourty-eight radiomics features for each AHA segment were extracted with pyradiomics v3.0.1. Clinical characteristics were retrieved from the MYKKE database. Patients were divided into the groups heart failure-like (HF) and non-HF-like myocarditis based on clinical symptoms and ejection fraction.

Results: We included 88 patients with EMB-proven myocarditis in our analyses: 29 patients in the HF-like- and 59 in the non-HF-like group with a median age of 7.0 (2.0-14.0) and 16.0 (14.0-17.0) years, respectively. Troponin Ths values were significantly elevated in the non-HF-like group (554 vs. 85 ng/l, p=0.005). We detected significantly higher median intensity values in the HF-like group in the segments basal, mid-ventricular and apical septal as well as mid-ventricular and apical inferior (Figure 1A). The texture parameter GLCM (Grey Level Co-Occurrence Matrix) contrast, describing grey differences between two neighboring pixels as an expression of homogeneity, showed higher values basal and mid-ventricular inferior and basal lateral in the HF-like group, indicating lower homogeneity while in the non-HF-like group this was present in the septal region (Figure 1B).

Conclusions: We presented distinct spatially resolved radiomics profiles in LGE CMR images in different clinically characterized pediatric patients with EMB-proven myocarditis. This classification by radiomics features in an initial CMR could have an impact on the risk stratification and therapy management of these patients in the future.



AHA-Models of median features for the heart failure (HF)-like and Non-HF-like group. A: Presenting different distributions of LGE intensity and B: GLCM (Grey Level Co-Occurrence Matrix) contrast for homogeneity.

Keywords: Myocarditis, CMR, LGE, Radiomcis, heart failure

OP-005

Pediatric myocarditis: Cardiac MRI helps to predict adverse outcome - results from the "MYKKE"- registry

Theodor Uden[1], Franziska Seidel[2], Bernd Opgen Rhein[3], Felix Anderheiden[4], Bardo Wannenmacher[5], Axel Rentzsch[6], Katja Reineker[7], Martin Boehne[1], Gesa Wiegand[8], Tobias Hecht[9], Anna Eva Blank[10], Marcus Fischer[11], Michael Kaestner[12], Rudi Foth[13], Eva Gatzweiler[14], Inga Voges[15], Bettina Ruf[16], Dietmar Boethig[1], Thomas Pickardt[17], Stephan Schubert[18], Philipp Beerbaum[1] and Daniel Messroghli[19]

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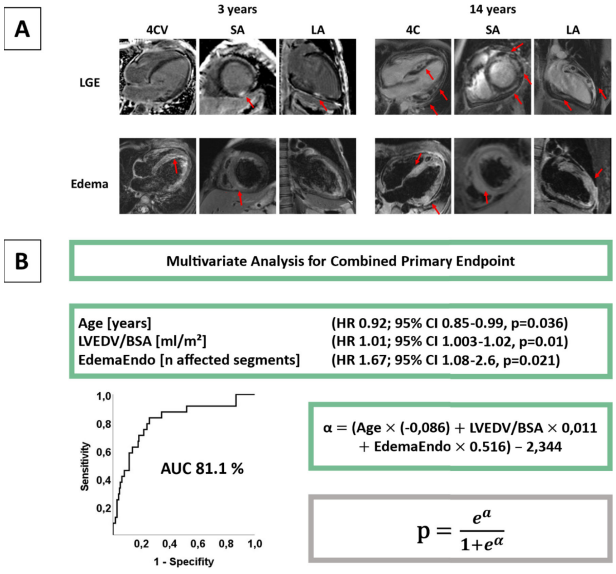
Background and Aim: Outcomes in children with myocarditis range from asymptomatic to severe heart failure. Late gadolinium enhancement (LGE) and edema in cardiac magnetic resonance imaging (cMRI) are widely used to assess myocardial affection. Specific LGE patterns are well-established in adult myocarditis, whereas data is limited in pediatric patients. Based on the prospective multicenter registry for suspected myocarditis in children and adolescents "MYKKE", our study characterizes LGE and edema patterns and evaluates their prognostic implications.
Method: Two examiners analyzed 214 initial good-quality cMRI examinations of pediatric patients with suspected myocarditis, which were performed between 2013 and 2020. Analyses included LGE, edema, cardiac function, and left ventricle volumetry. According to the AHA model, location and total number of affected segments and the affected layer within the wall were assessed for LGE and edema. Binary logistic regression´s main endpoint was adverse outcome, including death, resuscitation, heart transplantation, or mechanical circulatory support.
Results: Median age was 15.2 (IQR 11.5-16.5) years with a left ventricular ejection fraction (LVEF) of 60.9 (49.7-66.7). LVEF varied significantly regarding age ($p<0.001$), with patients aged ≤ 5 years demonstrating lower LVEF (27.4% (20.0-58.5) vs. 62% (57.2-67.1) in ≥ 12-year-old patients). Myocardial LGE was found in 128/214 patients (59.8%), and edema was detected in 70/214 patients (32.7%). The number of LGE- and edema-affected segments did not differ significantly regarding age groups. Patients ≥ 12 years presented with more epicardial-affected LGE segments than younger children. In 26 patients (12.1%) we observed the combined endpoint. In a multivariate model, the extent of endocardial edema, age, and the left ventricular end-diastolic volume relative to body surface area (LVEDV/BSA) predicted the primary endpoint with an area under the curve of 81.1%. The figure provides a formula allowing the patient-specific risk calculation. The formula needs further validation.
Conclusions: LGE and edema patterns, cardiac function, and outcome differed in children with suspected myocarditis depending on age. Endocardial edema extent, age, and LVEDV/BSA help to predict adverse outcome.

Keywords: myocarditis, children, late gadolinium enhancement, edema, pattern, risk

Representative Patient Cases of Different Age Groups and Multivariate Analysis Results of Significant Risk Factors



Section A shows late gadolinium enhancement and edema sequences of two patients of different age groups. The 3-year-old patient showed LGE findings only in the mid-inferior segment and edema in the neighboring mid-inferior segment. Clinically, this patient suffered from heart failure, with an LVEF of 11%. The 14-year-old patient showed LGE findings in the mid-inferoseptal and patchier in the basal and mid-anterolateral segments. The mid-ventricular short- and long-axis views show further affection in the mid-anterior, inferolateral, and inferior segments. Section B shows the results of the multivariate analysis. Age, LVEDV/BSA, and endocardial edema involvement significantly influenced the prognosis for the combined primary endpoint. The provided formulas calculate the risk to reach the endpoint. 4CV, 4-chamber view; SA, short axis view; LA, long axis view; LVEDV/BSA, left ventricular end-diastolic volume relative to the body surface area; n edema endo, number of endocardial edema-affected segments; e, Euler's number (~2,7182).

OP-006

Myocardial and liver T1 mapping and circulating fibrosis biomarkers in children with fontan circulation

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Background and Aim: Fontan patients are at risk of cardiac fibrosis from the underlying disease and cardiopulmonary bypass (CPB) surgeries, as well as liver fibrosis from chronically elevated hepatic venous pressure. To determine whether myocardial and liver T1 and circulating fibrosis biomarkers are related in this population.

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Method: Children in Fontan circulation prospectively underwent magnetic resonance imaging and blood tests of C-telopeptide for type I collagen (CITP), type III procollagen aminoterminal propeptide (PIIINP), insulin-like growth factor-binding protein 7 (IGFBP-7), tissue inhibitor of metalloproteinases (TIMP1, TIMP4), fatty acid binding protein 4 (FABP-4), growth differentiation factor 15 (GDF-15), soluble suppression of tumorigenicity 2 (sST2), protein delta homolog 1 (DLK1), and matrix metalloproteinase 2 (MMP2). T1 mapping with a modified Look-Locker sequence was performed at 1.5T before and after gadolinium administration, on a mid-ventricular short-axis slice and axially for the liver. Liver T1 was averaged over 3 regions of interest. Extracellular volume (ECV) was calculated using same-day hematocrit. Normal distribution was assessed, then Pearson's or Spearman's correlations were used. Groups were compared by t-test or Mann-Whitney-U test.
Results: Fourty-nine Fontan patients (30 male, age 13.5 +/- 3.1 (range 8-19) years) participated, 24 with a single left ventricle (LV) and 25 with a single right ventricle (RV). Patients had undergone a median of 3 (interquartile range 2-3) CPB surgeries.
Native myocardial T1 in Fontan patients (1023 +/- 47 ms) was within reference values and not age-dependent, but liver T1 was higher with increasing patient age (r=0.336; p=0.039). Native myocardial and liver T1 and myocardial ECV did not correlate with CITP, PIIINP, IGFBP-7, TIMP1, TIMP4, FABP-4, GDF-15, sST2, DLK1, or MMP2. Liver ECV correlated with sST2 (r=-0.393; p=0.032) only. Native myocardial T1 (1037ms vs. 1008ms; p=0.036) and liver ECV (49.4% vs. 40.7%; p=0.006) were higher in single RV than single LV anatomy, while no difference was found for myocardial ECV (28.6% vs. 27.8%; p=0.087) or for native liver T1 (730ms vs. 719ms; p=0.549).
Conclusions: Myocardial native T1 may be normal in Fontan patients and does not correlate with circulating fibrosis biomarkers. In contrast, liver T1 increases with age even during childhood, and liver ECV relates to ventricular morphology and sST2.

Keywords: T1 mapping, single ventricle, fibrosis, myocardium, liver

OP-007
Prognostic value of left atrial strain in pediatric cardiomyopathies: A multi-centre study

Jolanda Sabatino[1], Martina Avesani[2], Sara Moscatelli[3], Rosalba De Sarro[1], Nunzia Borrelli[3], Salvatore De Rosa[1], Daniela Concolino[1], Elena Reffo[2], Daniele Torella[1] and Giovanni Di Salvo[2]
[1]University of Catanzaro, Italy; [2]University of Padua, Italy; [3]Royal Brompton Hospital of London, UK

Background and Aim: The left atrium is an early indicator of left ventricular (LV) dysfunction. However, the prognostic significance of left atrial (LA) function, in children with dilated cardiomyopathy (DCM), hypertrophic cardiomyopathy (HCM), and restrictive cardiomyopathy (RCM) has not been well established.
This study aimed to assess the prognostic value of LA strain, measured using 2-D echocardiographic speckle-tracking analyses (2D-STE), in paediatric cardiomyopathies (CMP).
Method: The study has a multicentre retrospective design and involved children with cardiomyopathies, who had undergone standard echocardiographic examinations and 2-D speckle-tracking analyses, including LV longitudinal peak systolic strain (LS), and LA peak systolic strain. The primary endpoint was a combination of sudden or cardiac death, the need for a ventricular assist device, or cardiac transplantation.
Results: A total of 155 children were included in the study, with an average age of 8.8 ± 6 years, comprising 50 with DCM, 50 with HCM, 10 with RCM, and 45 healthy controls (CTRL). Twenty-two patients (14%) experienced the primary endpoint during a median follow-up of 5 years. LA peak systolic strain and strain rate values displayed a consistent and significant decrease with the severity of diastolic dysfunction. Several factors including left atrial volume index (LAVI), LV global longitudinal strain (GLS), and LA reservoir strain, but not left ventricular ejection fraction (LVEF) and LV global longitudinal strain (GLS), were associated with the outcome in univariate analysis (all $P < 0.05$). These independent variables were chosen according to univariable analyses and clinical relevance. LA reservoir strain emerged as a more robust predictor of the outcome compared to the other echocardiographic variables. In the multivariable model, LA reservoir strain remained significantly associated with the outcome (HR 0.84, 95% CI (0.72-0.97), p=0.016).
Conclusions: 2D-STE-derived LA reservoir strain serves as a robust and independent prognostic predictor in children with cardiomyopathies, surpassing the predictive capacity of LV GLS, LVEF, and LAVI. Therefore, considering LA strain in the management of children with CMP may enhance the risk stratification of the disease and facilitate the planning for early clinical intervention.

Keywords: cardiomyopathies, left atrial strain

Interventional Cardiology

OP-008
The practice of balloon atrial septostomy in the current ERA. A survey by the junior members of the interventional working group of the AEPC

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Background and Aim: Balloon atrial septostomy (BAS) is a time-sensitive procedure that improves oxygen levels in newborns with cyanotic congenital heart diseases. There is significant variability regarding procedural setting and technique among different centers and the recent unavailability of compliant Rashkind balloon catheters might have played a role in changing local BAS protocols. To investigate current BAS practice with non-compliant balloon.

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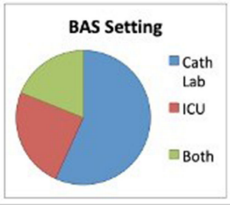
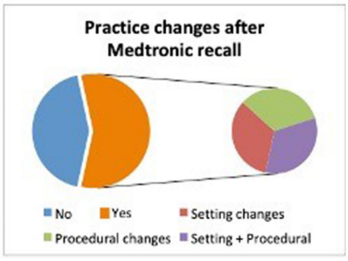
Method: AEPC Junior members created a 29-question survey, validated by pediatric cardiology experts. Distributed online, Excel was used to analyze responses.
Results: The survey collected responses from 36 centers in Europe and the UK. Most participants (83%) work in tertiary care centers. The majority of the respondents were consultant interventional cardiologists (38%) and interventional cardiology fellows (24%). BAS is performed by a general pediatric cardiologist in 13% of centers. 89% of them were under age 40. BAS is carried out exclusively in the cath lab (53%) or in the ICU (19%). 62% of centers perform BAS in the cath lab using fluoroscopy and 2D-echocardiography to guide the procedure. The femoral vein is the most common vascular access site (67%), with 75% of centers using an introducer sheath to gain access. Nearly half participants (51%) report the use of a diagnostic catheter and 78% the use of guidewire to access the left atrium. After the recall of the Medtronic catheter in August 2020, 57% of participants reported changes in the procedure of BAS regarding either the setting and/or the technical steps. The majority of respondents (78%) reported an intermediate, low or very low level of satisfaction with the currently available catheter balloons on the market.
Conclusions: There is significant variability in the BAS procedure as performed in different centers. It is performed mainly by the interventional cardiologists, in the cath lab. The recall of the last available compliant Rashkind catheter has been a significant factor in driving most centres towards this approach. Given the apparent variability and changes in BAS practices, it is essential to undertake a multicenter study to evaluate the safety and effectiveness of the procedure with non-compliant balloons.

Keywords: Balloon Atrial Septostomy, Survey, Junior AEPC members

Figure 1

Country	Number
Albania	1
Austria	1
Belarus	1
Belgium	2
Bosnia and Herzegovina	1
Czech Republic	1
Egypt	1
France	3
Germany	3
Italy	6
Latvia	1
Martinique	1
Poland	3
Portugal	3
Switzerland	1
Spain	2
United Kingdom	4
Uzbekistan	1
TOTAL	36





OP-009

Transcatheter atrial flow regulator device implantations in restrictive cardiomyopathy: Bridge to transplantation or destination therapy

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Background and Aim: The PRELIEVE study reported atrial flow regulator (AFR) implantations in patents > 65yrs old with HFpEF and HFrEF. We published 3 first-in-human cases of AFR-device implantation in restrictive cardiomyopathy (RCM), when LV dysfunction results in LA hypertension and postcapillary pulmonary hypertension (PH) (doi: 10.1016/j.jacc as.2022.05.010). Here, we report 1-4 year-clinical follow up of 7 RCM-post-AFR patients.
Method: Transcatheter AFR-implantations were performed in 7 children with RCM (age 3.5-13yrs) to create a restrictive LR-shunt (fenestration 6 or 8mm;Fig. 1). No procedure-related complications were observed.
Results: Patient #1 (P1; 13yr old F in NYHA-FC 3) greatly benefited clinically, and she had reduction of LA volume by 52% (CMR) 2 weeks after AFR-implantation. 8 months later, her mPAP had decreased from 30 to 19mmHg (no PH). 27 months after AFR-implantation, P1 is in excellent condition (NYHA-FC 2, interatrial dP=8mmHg), on eplerenone, clopidogrel and iron. P2 was a 11yr old girl with RCM/genetic syndrome in NYHA-FC4. The AFR immediately reduced PAWP from 29 to 21mmHg, and the patient improved clinically. She was not a Tx candidate and died 25 months after the procedure. P3 (6yr old F) had clinical and hemodynamic improvement after AFR-implantation, and converted from HLTx to HTx listing. She successfully underwent HTx 17 months after AFR-implantation. P4 was a 4yr old girl who decreased her mPAP from 38 to 33 mmHg and PAWP from 29 to 23 mmHg, 10min. after AFR-implantation. P5 (3.5yr old F) had mPAP 27 and LVEDP 25 mmHg at the time of AFR-implantation (pending 1yr-follow up). P6 was a 11yr old girl who was on BVAD for 22 months. AFR decreased

Transcatheter AFR device implantation

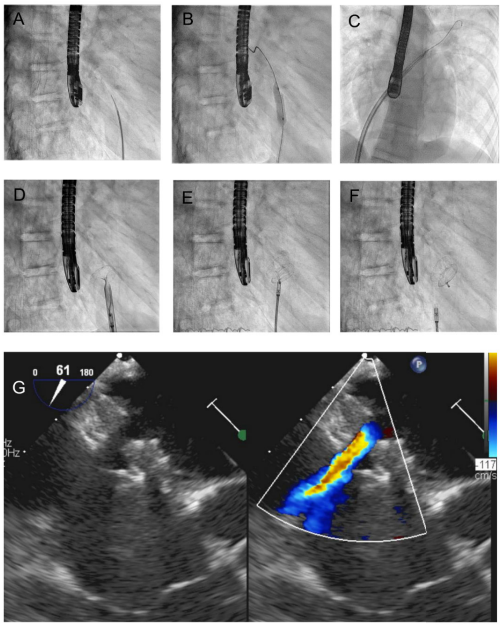

Figure 1

Interventional, percutaneous implantation of the Atrial Flow Regulator-Device in a 13 year old female with restrictive cardiomyopathy, heart failure with preserved ejection fraction (HFpEF), and mild pulmonary hypertension (PH).

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mLAP from 28 to 18 mmHg. She underwent successfull HTx 19 month later. P7 (15yr old F) had clinical and hemodynamic improvement after AFR implantation. Re-dilation of AFR 3 years later reduced mLAP from 17 to 13 mmHg. She is currently evaluated for HTx-listing.
Conclusions: AFR-implantation in RCM is safe and can prevent progressive postcapillary PH/PVR-elevation that would preclude heart transplantation (HTx) and require HLTx-listing. AFR-device implantation can be considered as bridge to HTx in young patients with fatal RCM, and destination therapy in those who are not HTx candidates.

Keywords: restrictive cardiomyopathy (RCM), Atrial Flow Regulator (AFR), interatrial device, heart failure, interventional cardiology

OP-010
30 Years' experience in percutaneous pulmonary artery interventions in transposition of the great arteries after the arterial switch operation

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Background and Aim: Pulmonary artery (PA) stenosis is common after arterial switch operation (ASO) for transposition of the great arteries (TGA). The effects of percutaneous interventions and differences between balloon angioplasty (BA) and stents on right ventricular and PA pressures are fairly unknown.We described our 30-years' experience in percutaneous PA (re)interventions after ASO.
Method: All TGA patients who underwent ASO between 1977 and 2022 in two Dutch congenital heart centers were included in this multicenter retrospective study. Perioperative ASO data, pre and post-interventional invasive pressures, gradients and estimated echocardiographic gradients were analyzed.
Results: Seventy-seven out of 960 (8%) TGA patients underwent 186 site-specific percutaneous PA (re)interventions after ASO. ASO time era's 1977-1989 (OR 0.207;95% CI 0.082-0.525; p=0.001) and 2010-2022 (OR 0.356;95% CI 0.197-0.640; p=0.001) were independent risk factors for PA interventions. Invasive post-interventional pressures and gradients were lower after stents compared to BA (RV pressure:47±14 vs 58±11; right PA (RPA)-PA gradient:11±11 vs 25±12, p<0.05; RV/systemic ratio:0.4±0.1 vs 0.6±0.2, p<0.001). Stents were associated with successful PA (re)interventions (OR 6.650;95% CI 2.660-16.623;p<0.001). In addition, 77% of the patients with unilateral PA stenosis (LPA 41%, RPA 59%) showed increased RV pressure (>30mmHg) and RV/systemic ratio improved post-intervention (0.5±0.2 vs. 0.6±0.2, p<0.05). We reported 19 complications (14.5%). Most complications were minor. Two post-procedural deaths were reported.
Conclusions: PA stenosis is common in TGA patients after ASO and stent treatment is superior to BA. Unilateral PA stenosis can already impact RV pressures. PA (re)interventions can be performed safely but caution for serious complications is warranted.

Keywords: transposition of the great arteries, arterial switch operation, intervention, pulmonary stenosis

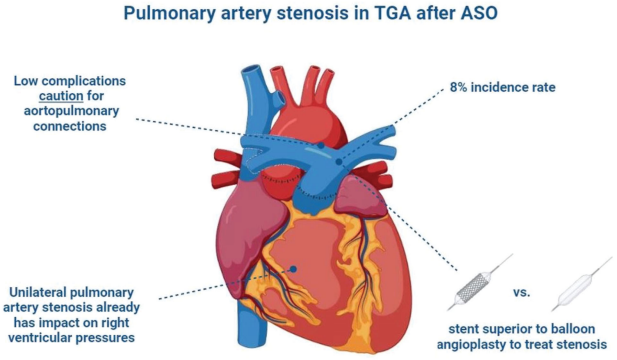

Figure 1. Pulmonary artery stenosis in TGA after ASO

ASO= arterial switch operation; TGA= transposition of the great arteries

OP-011
Acute changes in pulmonary capillary wedge pressure physiology following transcatheter pulmonary valve replacement

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Background and Aim: After transcatheter pulmonary valve replacement (TPVR), changes in left ventricular loading conditions can result in increased pulmonary capillary wedge pressure (PCWP). Understanding and defining this hemodynamic effect may allow more accurate prediction of hemodynamic instability, effect the resulting management plans, and may lead to an overall improvement in clinical outcomes. Our aim was to describe the relationship between pre- and post-procedural PCWP, or acute changes in PCWP, and outcomes after TPVR.
Method: We present a single center, retrospective cohort of patients that underwent TPVR at the University of Colorado in whom we recorded a pre- and post-TPVR PCWP measurement. Three outcomes were considered: need for ventilatory support, prolonged length of stay (>48 hours) and death. Abnormal wedge physiology was defined as an elevated pre-PCWP (>10 mmHg) or an increase (delta-PCWP) of at least 5 mmHg after TPVR. ROC analysis was conducted for each outcome with four test variables: pre- and post-PCWP, delta-PCWP, and percent change in PCWP.
Results: A total of 76 patients were included with a median age at procedure of 20 years of age (IQR 14.0-35.8, and range 6-69). Of all patients included, 36 (42.1%) were considered to have abnormal wedge physiology, and 44 (57.9%) were considered to have normal wedge physiology. In the ROC analysis, there was a significant association between the need for post procedural ventilatory support and pre-PCWP >10.5 mmHg (AUC 0.724, sensitivity 67%, specificity 82.12%, +LR 3.76, -LR 0.4) and post-PCWP >11.5

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mmHg (AUC 0.804, sensitivity 100% and specificity 68.5%, +LR 3.17, -LR 0.4), as well as a weak association with delta-PCWP >4.5 mmHg (AUC 0.687, sensitivity 67%, specificity 82.2%, +LR3.76, -LR 0.4).
Conclusions: Understanding the relationship between PCWP and short-term outcome aids clinical decision-making and patient care. Both pre- and post-PCWP, as well as an acute change, are measurements that may offer predictive value in assessing the need for post-procedural death, ventilatory support, and prolonged length of stay. However, further studies are necessary to validate these findings.

Keywords: transcatheter pulmonary valve replacement, congenital heart disease, pulmonary capillary wedge pressure.

Figure 1

A. Demographic, Clinical and Procedural Characteristics

Variable	Total n=76	Abnormal wedge physiology n= 32	Normal wedge physiology n=44	p value
Sex, female, n (%)	35 (46.1)	15 (46.9)	20 (45.5)	0.544
Age, years, median (IQR)	20 (14.0-35.8)	22.5 (13.5-41.3)	18.5 (14-32)	0.199
Weight, kg, median (IQR)	65.9 (46.8-87.0)	70.6 (46.8-89.9)	60.4 (47.0-77.0)	0.141
Height, cm, median (IQR)	163.3 (150.3-175)	161 (150.3-177.3)	165.5 (150.8-174.2)	0.797
Pre-implant hemodynamics				
Systolic RV pressure, mmHg, median (IQR)	36.3 (50-61.8)	54.0 (38.5-61.8)	48.0 (30.0-63.0)	0.267
RVEDp, mmHg, median (IQR)	8.5 (6.0-10.8)	10 (8.0-14)	7.0 (5.3-10)	0.002
Mean PA pressure, mmHg, median (IQR)	17 (13-20.8)	20.0 (17.0-24.0)	16.0 (12.0-17.5)	<0.001
Pressure gradient RVOT, mmHg, median (IQR)	20.5 (5.0-33)	18.0 (5.0-30.0)	24.0 (5.0-34.8)	0.253
Pre-PCWP, mmHg, median (IQR)	7.0 (6.0-10.0)	10 (8.3-12.8)	6.0 (5.0-7.0)	<0.001
CO, L/min/m2, median (IQR)	3.3 (2.7-3.8)	3.0 (2.5-3.5)	3.4 (2.8-4.3)	0.037
PVR, Woods/1/m2, median (IQR)	2.6 (1.9-3.3)	2.9 (2.0-3.5)	2.5 (1.8-3.1)	0.260
Post-implant hemodynamics				
Systolic RV pressure, mmHg, median (IQR)	30.0 (27.0-38.0)	37.0 (32.5-47.8)	28.0 (24.0-32.0)	<0.001
RVEDp, mmHg, median (IQR)	6.0 (5.0-9.0)	8.5 (6.0-11.8)	6.0 (4.0-8.0)	<0.001
Mean PA pressure, mmHg, median (IQR)	18.0 (15.0-23.8)	23.5 (19.3-27.0)	16.0 (13.3-18.0)	<0.001
Pressure gradient RVOT, mmHg, median (IQR)	3.0 (0.0-5.0)	3.0 (0.0-5.0)	2.0 (1.0-6.0)	0.983
ICE- velocity across the PV, m/s, median (IQR)	1.6 (1.4-1.9)	1.6 (1.4-1.9)	1.5 (1.2-1.8)	0.205
Post-PCWP, mmHg, median (IQR)	9.5 (7.3-13.0)	13.0 (11.0-16.8)	8.0 (7.0-9.0)	<0.001
Delta PCWP, mmHg, median (IQR)	2.0 (1.0-4.0)	4.0 (1.0-6.0)	2.0 (1.0-3.0)	0.020
% change in PCWP, %, median (IQR)	25.0 (9.1-40.0)	25.0 (7.7-41.7)	25.0 (11.5-37.5)	0.866
Need for ventilatory support, n (%)	3.0 (3.9)	3.0 (9.4)	0	0.071
Prolonged legnth of Stay, n (%)				0.892
48 hours	4.0 (5.3)	2.0 (6.3)	2.0 (4.5)	
>48 hours	4.0 (5.3)	2.0 (6.3)	2.0 (4.5)	
Death, n (%)	3.0 (3.9)	2.0 (6.3)	1.0 (2.3)	0.381

B. ROC Curve Analysis: Death, Ventilatory Support and Prolonged Length of Stay

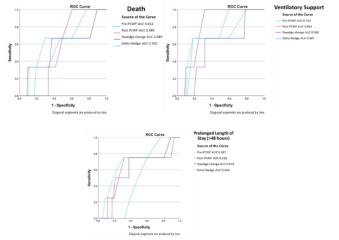

Figure 1A. Abnormal wedge physiology was defined as pre-PCWP>10 mmHg or delta-PCWP>5 mmHg. IQR: interquartile range; RV: right ventricle; RVEDp: right ventricular end diastolic pressure; PA: pulmonary artery; RVOT: right ventricular outflow tract; PCWP: pulmonary capillary wedge pressure; CO: cardiac output; ICE: intracardiac echocardiography. ROC analysis for three outcomes (death, ventilatory support and prolonged length of stay) with four test variables: pre-pulmonary capillary wedge pressure (PCWP), post-PCWP, percent change in PCWP and the difference between pre and post PCWP (Delta PCWP). Figure 1B. Ventilatory support was defined as any type of high flow oxygen device or advance ventilatory support; prolonged length of stay was defined as more than 48 hours after procedure.

OP-012

Treatment of RVOT dysfunction in the ERA of self-expandable valve: Initial results from the italian registry

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Background and Aim: The introduction of self-expandable valves could have a significant impact on the feasibility of transcatheter treatments of RVOT dysfunction. Venus P valve is the first autoexpadable Valve that received CE mark in 2022. Here, we report on the data about patients needing pulmonary valve implantation at the beginning of the autoexpandable valves era in Italy.
Method: Seven italian Centers were included in the study. The data of the patients who underwent a surgical or percutaneous valvolization from May 2022 to September 2023, were collected. Indications for valvolization was based on recently published guidelines.
Results: During this period, 231 Patients underwent pulmonary valve implantation, 124 (54%) surgically and 107 (46%) percutaneously. Among 107 patients, who underwent a percutaneous pulmonary valve implantation, 54 (51%) patients received a Sapien Valve, 16 (15%) Patients a Melody valve, 3 patients a Myval (3%) and 34 (31%) patients an autoexpandable valve. A venus P valve was implanted in 33 patients, while an Alterra was implanted in one patient through a compassionate use. Mean age was 35±14 years, and mean weight 63 ± 22 Kg. Almost all the patients were affected by Tetralogy of Fallot. At sizing balloon, mean RVOT diameter was 34 ± 7 mm. Only one procedural complication occurred: one patient had pulmonary bleeding that resolved spontaneously. At follow up of 50 ± 90 days all the patients are in good clinical condition. Nine patients underwent a CT scan at three months and in 4 patients a partial thrombosis of one of the valvar cusps was detected with no haemodynamic.
Conclusions: At the beginning of self-expandable valves almost 31% are treatable by using a transcatheter approach with good results. Further studies are needed to analyze how this technology will change the impact of transcatheter approach in treating RVOT dysfunction.

Keywords: RVOT dysfunction, Autoexpandable valves

OP-013

Outcome of manually modified microvascular plugs to pulmonary flow restrictors in various congenital heart disease

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Background and Aim: The development of microvascular plugs (MVPs) has enabled novel transcatheter deliverable endoluminal pulmonary flow restrictors (PFRs) with the potential to treat newborns and infants with life-threatening congenital heart diseases (CHDs) in a minimally invasive manner. We present our experience to evaluate the efficacy of this concept in controlling pulmonary blood flow in various CHDs.
Method: Retrospective clinical data review of patients with CHD and pulmonary over-circulation who received bilateral PFRs percutaneously.
Results: Twenty-eight PFRs (7 MVP-5Q, 12 MVP-7Q, and 9 MVP-9Q) were finally implanted in 14 patients with a median age of 1.6 months (IQR, 0.9–2.3) and a median weight of 3.1 Kg (IQR, 2.7–3.6). Nine patients had large intra-cardiac left-to-right shunts (including 3 with fatal trisomy and palliative programs), 2 had borderline left ventricles, 2 had Taussig-Bing anomaly, and one had a hypoplastic left heart. Four patients had concomitant ductal stenting. Two MVP-5Qs were snare-removed and upsized to MVP-7Q. Patients experienced a significant drop in oxygen saturation and Qp/Qs. All patients were discharged from

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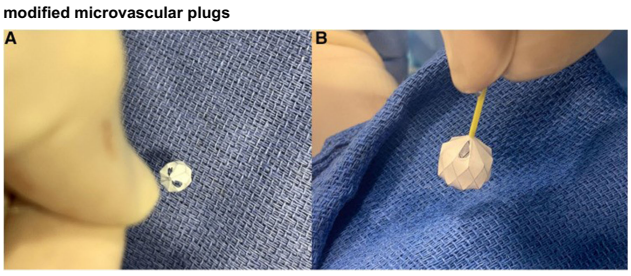
the ICU after a median of 3.5 days (IQR, 2–5.8) postoperative. Five patients had routine inter-stage catheterization and no device embolization or pulmonary branch distortion was seen. Fourteen (50%) PFRs were surgically explanted uneventfully on a median of 4.3 months (IQR, 1.2–6) post-implantation during biventricular repair in 6 patients and stage-2 palliation in one patient. The latter died 1 month post-operative from severe sepsis. Four patients are scheduled for surgical PFR removal and biventricular repair. Two patients with trisomy 18 died at 1 and 6.8 months post-procedure from non-cardiac causes. One patient with trisomy 13 is alive at 2.7 months post-procedure.
Conclusions: It is feasible to bespoke MVPs and implant them as effective PFRs in various CHDs. This approach enables staged left ventricular recruitment, comprehensive stage-2 or biventricular repair with lower risk by postponing surgeries to later infancy. Device explantation is uneventful, and the outcomes afterward are promising.

Keywords: congenital heart disease, microvascular plug, pulmonary artery band, pulmonary flow restrictor, transcatheter intervention



OP-014
Off-label use of the lifetech konar-MF VSD occluder for transcatheter closure of patent ductus arteriosus: A multicentre experience

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Background and Aim: Transcatheter patent ductus arteriosus (PDA) closure can be challenging in patients with large PDAs and/or unusual ductal anatomy. We aim to assess efficacy and safety of the off-label use of the new multifunctional occluder (MFO, Lifetech Konar-MFTM) in PDA closure.
Method: Patients in whom PDA closure was attempted with the MFO device in 11 tertiary pediatric cardiology centers (4 countries, 2018-2023) were retrospectively analyzed to review procedural outcomes.
Results: A total of 45 interventions were recorded in 43 patients (3 adults) with a hemodynamically significant ductus. Of the 41 (91%) children [median age: 0.7 year (range:0.2-17.3), median procedural weight: 6.9kg (range:1.7-32)], 7 (16%) were ex-preterm and 18 (40%) weighted <6kg. PDA was large [ductal diameter: 3.5mm (range:1.6-9)], hypertensive in 21 (49%, pulmonary artery (PA) pressures >2/3 systemic) and mainly conical (Krichenko type A: 58%). Of note, PDA was window-type or complex in 36% (Type B: 18%, Type D: 18%). Procedure was performed under both fluoroscopy [median fluoro time: 10.4min (range:2.0-34.3)] and transthoracic echocardiography guidance, with mainly both arterial and venous femoral access (86%) and either antegrade (73%) or retrograde (27%%) delivery of the device [MFO 5/3: 4 (9.3%), 6/4: 14 (32.6%), 7/5: 5 (11.6%), 8/6: 4 (9.3%), 9/7: 2 (4.6%), 10/8: 10 (23.2%), 12/10: 3 (7.0%), 14/12: 1 (2.3%)]. An alternative device had been previously attempted in 28% cases. Successful device implantation was achieved in 41 (96%) patients with no peri-procedural complication. Early migration of a 7/5 MFO to the right PA occurred in a 4kg infant with successful percutaneous retrieval and successful implantation of a 10/8 MFO. A 3.8kg infant had significant residual shunt after 8/6 MFO implantation, which was percutaneously snared and retrieved 5 weeks later with successful implantation of a 9/7 MFO and complete PDA closure. After a median follow-up of 2.6 years (range:0.4-5.9), 2 (4.6%) patients required surgical revision due to device-related obstruction of the left PA, 6 and 8 months later. The outcome was otherwise uneventful.
Conclusions: In the first multi-institutional series, off-label use of the MFO device for transcatheter closure of challenging PDAs may be a safe and efficient therapeutic strategy.

Keywords: Patent ductus arteriosus, multifunctional occluder, transcatheter closure

OP-015
Overexpansion of the edwards sapien 3 in the native right ventricle outflow tract: A retrospective single center experience

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Background and Aim: Transcatheter pulmonary valve implantation (TPVI) is recommended to treat the dysfunctional right ventricular outflow tract (RVOT) but, not infrequently, native RVOTs with borderline dimensions may require valve overexpansion. We herein sought to evaluate TPVI feasibility with Edwards Sapien 3 (S3) valve overexpansion in the native enlarged RVOT.
Method: Retrospective analysis was performed on 43 patients undergoing TPVI with S3 valve in the dysfunctional native RVOT, retrieving baseline characteristics, procedural TPVI data

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and follow up records. If available, in patients submitted to TPVI with overexpanded 29 mm S3 valve, preoperative computer tomography (CT) was retrieved to pinpoint the specific characteristics of the RVOT anatomy.
Results: TPVI with S3 valve expanded at nominal inflation volume was performed in 29 patients (group 1) while the remaining 14 patients (group 2) underwent TPVI with S3 valve overexpansion; one case of valve embolization was noted. Baseline characteristics and pre-TPVI evaluation were comparable, but RVOT size was larger (p=0.001) at balloon interrogation in group 2. TPVI success (p=0.15) and 30-day mortality (p=0.33) remained comparable, with no clinically relevant differences at follow up. CT-based analysis revealed that hourglass (Type V) landing zone (LZ) shape was predominant in patients from group 2: the focal LZ dimension was compatible with the nominal S3 size while LZ cross-sectional dimensions remarkably exceeded the nominal S3 area and perimeter moving towards LZ proximal (p≤0.02) and distal (p=0.004) extremities.
Conclusions: TPVI with S3 valve overexpansion is feasible in enlarged native RVOTs with adequate anatomical substrate and comprehensive characterization of LZ geometry may guide the decision-making process.

Keywords: Native right ventricle outflow tract, overexpansion, transcatheter pulmonary valve implantation

Paediatric Cardiovascular Intensive Care

OP-016
Local pulmonary administration of factor VIIA (RFVIIA) in massive pulmonary haemorrhage in post-operative cardiac infant

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Background and Aim: Diffuse pulmonary haemorrhage is an ominous condition that has a high paediatric mortality rate. Recombinant activated factor VIIa (rFVIIa) is a powerful haemostatic agent which has been used intravenously in life-threatening haemorrhage in variety of conditions in which conventional medical or surgical therapy are unsuccessful.
Method: We report off-label use of endotracheal rFVIIa for massive life-threatening respiratory haemorrhage following aspiration and cardiopulmonary resuscitation in a 3-month-old infant who was anticoagulated with enoxaparin following corrective cardiac surgery with other comorbidities.
Results: Off-label administration of endotracheal rFVIIa permitted rapid safe control of massive pulmonary haemorrhage and prevented further detrimental decline in respiratory function with satisfactory outcome.
Conclusions: Off-label use of local rFVIIa, in addition to standard measures, can help in achieving rapid control of severe pulmonary haemorrhage.
- Our report promotes an alternate local form of administration of pro-thrombotic agent (rFVIIa) in a novel fashion to control life-threatening pulmonary haemorrhage.
- It adds to the existing literature on the increasing use of rFVIIa in the paediatric cardiac surgical population.

Keywords: Pulmonary haemorrhage, recombinant factor rFVIIa, cardiac surgery

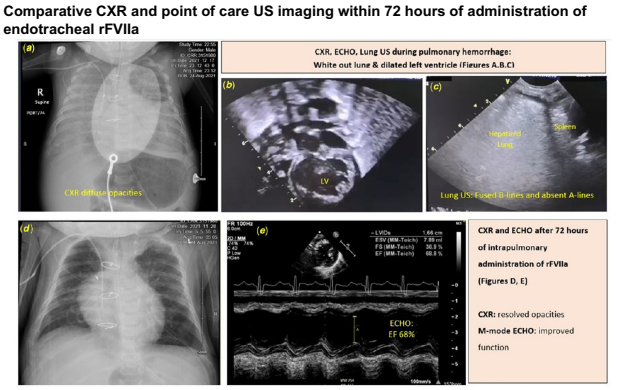

Figure 1. Comparative CXR and point of care US imaging within 72 hours of administration of endotracheal rFVIIa following cardiac arrest and pulmonary haemorrhage, in 3-month-old infant with repaired congenital heart diseases. EF: ejection fraction; LV: left ventricle; US: ultrasound.

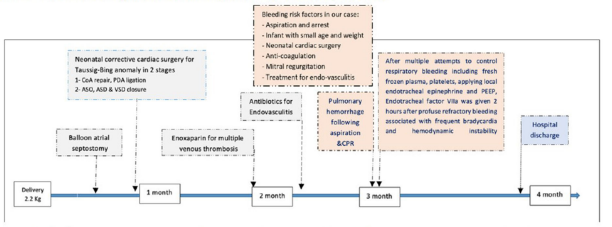

Figure 2. Timeline diagram outlining patient's hospital clinical course over [4] months period in 4-month-old cardiac infant post neonatal corrected complex cardiac surgery with pulmonary haemorrhage treated with local endotracheal rFVIIa. ASO: arterial switch operation; ASD: atrium septum defect; CoA: CoArctaion repair; CPR: cardiopulmonary resuscitation; PDA: patent ductus arteriosus; Peep: positive end expiratory pressure; VSD: ventricular septum defect.

Figure 1. Comparative CXR and point of care US imaging within 72 hours of administration of endotracheal rFVIIa following cardiac arrest and pulmonary haemorrhage, in 3- month-old infant with repaired congenital heart diseases. EF: ejection fraction; LV: left ventricle; US: ultrasound. Figure 2. Timeline diagram outlining patient's hospital clinical course over [4] months period in 4-month-old cardiac infant post neonatal corrected complex cardiac surgery with pulmonary haemorrhage treated with local endotracheal rFVIIa. ASO: arterial switch operation; ASD: atrium septum defect; CoA: CoArctaion repair; CPR: cardiopulmonary resus citation; PDA: patent ductus arteriosus; Peep: positive end expiratory pressure; VSD: ventricular septum defect

Congenital Heart Surgery

OP-017
Assessment of the pulmonary arterial growth and results of two-stage repair in infants with severe form of tetralogy of fallot

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Background and Aim: Assess of the pulmonary artery growth and the outcomes of complete repair after palliative treatment in infants with critical form of Tetralogy of Fallot.
Method: This was prospective randomized two-center study (2019 to 2022). Its included infants with Tetralogy of Fallot who underwent palliation with either stenting of the right ventricular outflow tract (stent group, n=21) or modified Blalock-Taussig shunt (shunt group, n=21).

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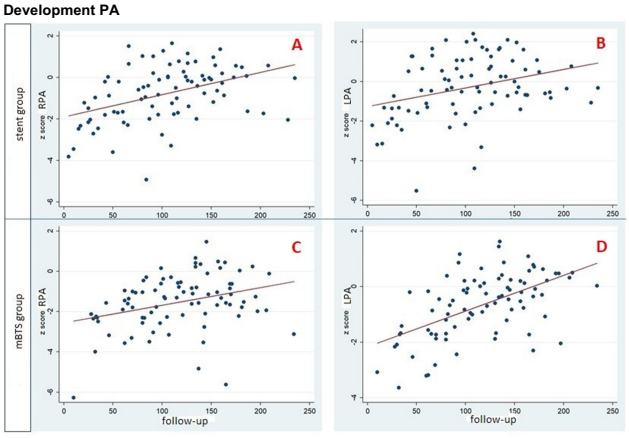
Results: In the stent group Nakata index increase from median 104.2 to 208.6 mm2/m2, while in the shunt group, it increased from 107.3 to 169.4 mm2/m2 ($p<0.01$). Mixed model analysis showed that in stent group the right pulmonary artery growth rate was 2.05*10-2 z scores/day, which was 3.01 times higher than in the shunt group. In stent group the left pulmonary artery growth rate was 2.3*10-2 z scores/day, which was 1.47 times higher than in the shunt group. In the stent group, there was one noncardiac-related mortality during the intermediate period. Transannular patch repair of the right ventricular outflow tract was performed in 12 patients (60%) in the stent group and in 15 patients (71.4%) in the shunt group ($p=0.52$) during complete repair. At 8 cases (40%) in the stent group and 6 cases (28.6%) in the shunt group, pulmonary artery replacement was performed. Time to surgical repair was shorter in the stent group ($p=0.046$), while the aortic cross clamp time ($p<0.01$) and cardiopulmonary bypass time ($p<0.01$) were significantly shorter in the shunt group.
Conclusions: Right ventricular outflow tract stenting provides hemodynamic stabilization and uniform growth of the pulmonary artery compared to modified Blalock-Taussig shunt

Keywords: tetralogy of Fallot, pulmonary artery hypoplasia, two-stage repair, stenting of the right ventricular outflow tract, modified Blalock-Taussig shunt



OP-018
Cardiopulmonary bypass priming with allogeneic red blood cells – analysis of immune mediators and their alterations following hemofiltration

Aylin Poertecene[1], Svea Kleiner[1], Leonie Trachte[1], Christoph Kammeyer[1], Sebastian Tiedge[2], Joerg Optenhoefel[2], Alexander Horke[2], Dennhardt Nils[3], Nicole Ruebsamen[4], André Karch[4], Christine Susanne Falk[5], Philipp Beerbaum[1] and Martin Boehne[1]
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Background and Aim: The unfavourable ratio between blood volume and filling volume in paediatric cardiopulmonary bypass (CPB) systems often requires priming with red blood cell concentrate (RBC). Our study focused on the RBC-triggered release of immune mediators and how hemofiltration affects these mediators during the priming process.
Method: In a prospective study involving 22 children (3.0-10.5 kg) undergoing congenital heart surgery, a standardized CPB system was primarily primed with 125 ml of RBC (age: 7-28 days) and bicarbonate-buffered hemofiltration solution. Hemofiltration of the priming was performed before cannulation. 50 immune mediators in the used RBCs, the priming solution before and after hemofiltration, as well as in the filtrate and preoperative patient samples were analysed.
Results: Out of 50 mediators, 29 were detectable in over half of the used RBCs. No correlation was observed between RBC storage duration and cytokine load. Following circulation in the CPB system, the majority of mediators showed a relevant increase in concentration and cytokine load. After hemofiltration, while the concentration of numerous immune mediators remained stable in the priming, its cytokine load decreased significantly and many cytokines were detectable in the filtrate. Most cytokines were found at higher concentrations in patients than in the final priming solution.
Conclusions: Regardless of the storage duration, numerous immune mediators were present in the RBCs and increased during the processing. Hemofiltration as a pretreatment for priming results in a reduction in cytokine load. The pathophysiological role of the remaining inflammatory mediators as well as potential strategies to further optimize the priming solution should be evaluated in subsequent studies.

Keywords: cytokines, cardiopulmonary bypass, hemofiltration, red blood cell, priming

OP-019
Bridging infants with single ventricle physiology by pig cardiac xenotransplantation – a new therapeutic possibility

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Background and Aim: The results of cardiac allotransplantation in neonates and infants with complex congenital heart disease are better than in any other age group, with an approximate 60% survival at 25 years. However, the lack of suitable deceased human donors is associated with a high mortality of infants on the waitlist even if a mechanical assist device is used to support the circulation. The results of staged palliative surgical procedures remain suboptimal. We are investigating whether an infant could be supported by the orthotopic transplantation of a gene-edited pig heart until a deceased human heart becomes available, which in the USA is a mean period of approximately 4 months.
Method: Pigs with 10 or more genetic modifications are now available. These gene edits are directed to preventing injury of the heart by the human immune response and consist of (i) deletion of expression of the three known pig xenoantigens against which humans develop cytotoxic antibodies and (ii) insertion of transgenes for human protective proteins, e.g., complement- and coagulation-regulatory proteins. Novel immunosuppressive therapy based on blockade of the CD40/CD154 T cell co-stimulation pathway successfully prevents T cell rejection and de novo

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antibody production. In addition, survival of the pig graft may be enhanced by (i) the infant's relatively immature (and 'flexible') immune system and (ii) the thymectomy that is commonly carried out during heart transplantation.
Results: In our experience to date, the combination of the transplantation of a gene-edited pig heart and co-stimulation-based immunosuppressive therapy has resulted in graft survival extending to a maximum of 8 months, a sufficient time to enable an allograft to be obtained. Current evidence is that, even if the recipient becomes sensitized to pig antigens, this would not be detrimental to the outcome of subsequent allotransplantation.
Conclusions: Successful bridging would provide valuable experience on which to progress to pig cardiac xenotransplantation as destination therapy.

Keywords: bridging, heart, pig, xenotransplantation

OP-020
Management and outcomes of ebstein anomaly in neonates and infants: Own experience
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Background and Aim: Ebstein's anomaly is a complex congenital heart disease in which the septal and posterior cusps of the tricuspid valve (TV) are tightly linked to the adjacent myocardium and there is displacement of the functional TV ring towards the apex of the heart. To analyze UCCC experience of management and outcomes of Ebstein anomaly in neonates and infants.
Method: From 1999 to 2023 in UCCC 21 neonates with Ebstein's anomaly was admitted in ICU department in a critical condition. The mean age of patients was 7.5 + 3.3 days. Preoperative diagnosis was made by the anatomy of the tricuspid valve: type "B" in 6 patients, type "C" in 11 patients, type "D" in 4 patients. During the same time period 10 infants with EA were operated on. Preoperative diagnosis was made by the anatomy of the tricuspid valve: type "A" in 2 patients, type "B" in 4 patients, type "C" in 4 patients.
Results: In the group of newborns, it was possible to stabilize the condition thanks to ICU protocols in 9 patients. 3 of them were operated on in infancy, and 6 - after 1 year without mortality. 12 newborns were operated on in neonatal period due to their unstable condition. In 7 (58.3%) of them EA was combined with pulmonary blood flow obstruction of various degrees, and in 1 (8.3%) - with coarctation of the aorta. Early postoperative mortality in this group was 58.3% (7 patients). In the group of infants early mortality was 30% (3 patients). Average postoperative follow-up was 3.5 + 2.8 years. During the follow-up visit the clinical condition of the patients was examined, evaluated data of ECG, echocardiography, chest X-Ray and magnetic - resonance imaging of the heart. In late postoperative period reoperation was performed in 3 (60%) patients in newborn's group and 1 (14.2%) patient in infants group. There was not any late death.
Conclusions: Surgical interventions in newborns and infants have high operative mortality, while patients operated after one year and in adulthood have a low mortality rate.

Keywords: Ebstein's anomaly, neonatal period, infancy.

OP-021
Long term outcomes of infant mitral valve surgery
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Background and Aim: Outcomes associated with mitral valve surgery in infants are poorly described. Previous studies have indicated high mortality and re-intervention rates following mitral valve replacement. Very few studies have investigated the long-term consequences of valvuloplasty in this specific age group.
Objectives: Evaluate patient outcomes with mitral disease (stenosis, regurgitation or both) who had mitral valvuloplasty or replacement in the first year of life.
Method: Descriptive monocentric retrospective study including all children with mitral valve repair or replacement under 1 year of age over a period of 22 years (2001-2023). The outcomes assessed were: early mortality (at 30 days), late mortality, need for re-intervention.
Results: 56 patients were identified. Median age at surgery was 147 days and median weight was 5.1 kg. The most frequent mitral malformation was commissural fusion (50%). 39 patients underwent mitral valve repair and 17 mitral valve replacement. 24 patients had associated repairs. Mean duration of ICU stay was 7 days (2-87). Overall median hospital stay was 11 days. 11 patients had delayed sternal closure and one had veno-venous ECMO. Ten patients died (mortality rate: 17,9%). Mortality rate before discharge was 7.1%. Overall survival was 96% at 30 days, 86% at 1 year 82% at 3 years. Overall survival was significantly higher in patient with mitral valve repair compared to mitral valve replacement ($p=0.039$). The only mortality risk factor identified was replacement of the mitral valve compare to repair, HR 9 (1.3-94.8; $p=0.038$). 23 patients (41.1%) needed re-intervention: 16 had mitral valve replacement, 6 mitral valvuloplasty and 2 pacemaker implantations. 7 patients (12.5%) had more than one re-intervention. The re-intervention free survival rate after repair was 81% at 1 year; 65% at 5 years and 46% at 10years. The re-intervention free survival rate after replacement was 74% at 1 year; 55% at 5 and 10years. A log-rank test showed no difference in re-intervention free survival between mitral valve repair or replacement.
Conclusions: Mitral valve surgery in infants carries particularly high risks and is associated with a high rate of reinterventions. Mitral valvuloplasty has better outcomes in terms of mortality but only postpones the need for valve replacement.

Keywords: cardiac surgery, mitral valve, valvuloplasty

Paediatric Cardiovascular Intensive Care

OP-022
Circuit and membrane change in pediatric ECMO in cardiac surgery, experience over 10 years
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Background and Aim: Data on membrane and circuit change on pediatric patients with extracorporeal membrane oxygenation

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(ECMO) support are still limited. The outcome and indication on circuit and membrane change remains controversial.
Method: A descriptive retrospective monocentric review included all patients underwent ECMO from 2013 to 2022 in cardiac intensive care. The primary outcome was survival after ECMO weaning and secondary outcome was neurological complications and hemorrhagic complications on ECMO.
Results: Over 10 years, 131 patients received ECMO support in cardiac intensive care. 118 ECMO were veno-arterial and 13 were veno-venous. 43 patients (40%) underwent one or more membrane or circuit change (MCG), and 77 patients underwent none. First MCG happens on average 6 days and second MCG 13 days after ECMO support. Main indications for MCG were hemolysis and intra device thrombi. MCG was associated with longer ECMO duration (5 days versus 14 days) increased hemorrhagic complication and hemodialysis use, but less neurological complication. Hemorrhagic and neurological complications were the only identified independent mortality predictor, but not MCG.
Conclusions: Membrane and circuit change are associated with longer ECMO run but not with poorer outcome.

Keywords: ECMO, pediatric, neonatal, cardiac surgery, circuit

Congenital Heart Surgery

OP-023
Safe and effective use of apixaban in paediatric patients with heartmate 3 ventricular assist device

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Background and Aim: The growing population of children relying on the HeartMate 3 (HM 3) ventricular assist device (VAD) requires effective and safe anticoagulation. Apixaban's efficacy as an anticoagulant is being increasingly recognized. These cases demonstrate the safe and effective utilization of apixaban in paediatric patients with the HeartMate 3 VAD.
Method: Since June 2023, all paediatric patients who received a HM 3 VAD at our centre were prescribed Apixaban. The initial dose was 2.5 mg twice daily, adjusted based on Apixaban-specific anti-Xa chromogenic analysis trough levels (goal > 50 ng/dl). Additionally, all patients received daily aspirin.
Results: Three patients who received a HM 3 VAD were initiated on Apixaban. The median age of VAD implantation was 9 years (range, 9 – 12), with a median weight of 25 kg (range, 18 – 36) and a median body surface area of 1.00 m^2 (range, 0.78 – 1.22). The indication for VAD implantation in all cases was dilated cardiomyopathy. Apixaban was started at a median of 13 days after VAD implantation. There was a total of 345 days of VAD support with Apixaban anticoagulation, with a median time on Apixaban of 143 days. No thrombotic complications, bleeding, death, or stroke occurred. All three patients remain on VAD support, awaiting heart transplantation.
Conclusions: The use of apixaban was found to be effective, with minimal hemocompatibility-related events. These cases provide valuable insights into the successful application of apixaban as an anticoagulant in paediatric patients with HM3 VADs, demonstrating its potential as a promising alternative to traditional anticoagulant therapies.

Keywords: Apixaban, Heartmate 3, VAD, paediatric, anticoagulation

Cardiac Dysrhythmias and Electrophysiology

OP-024
Arrhythmias in congenital heart disease: A nationwide cohort study

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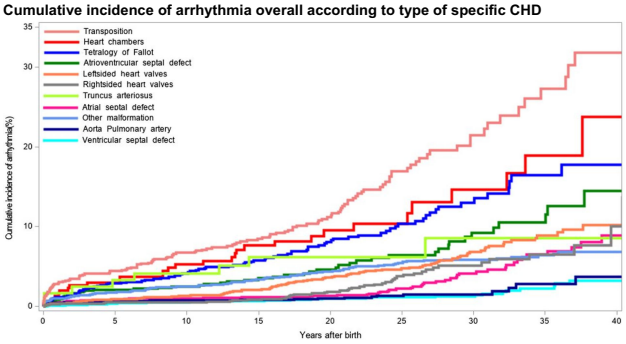
Background and Aim: An increasing number of patients with congenital heart disease (CHD) survive into adulthood, resulting in an expanding population of adults with CHD and an emerging number of complications including arrhythmias. However, the long-time risk of arrhythmias remains sparsely investigated and needs further research.
Method: In this Danish observational cohort study, all patients with CHD born from 1977 to 2018 were identified using nationwide registries and followed from date of birth until occurrence of first-time arrhythmia, emigration, death, or end of follow-up (December 31, 2018). The risk of arrhythmias was assessed among patients with CHD and compared with age- and sex-matched controls from the background population.
Results: A total of 23,464 patients with CHD (50.0% men) were identified and matched with 93,856 controls from the background population. During a median follow-up of 17.6 years, 3.1% of patients with CHD and 0.1% of controls developed arrhythmias – corresponding to incidence rates (IR) of 1.8 (95% CI 1.6-1.9) and 0.1 (95% CI 0.05-0.1) per 1,000 PY, respectively, and a hazard ratio of 32.0 (95% CI 25.5-40.2). The most common arrhythmias in patients with CHD were advanced atrioventricular block (IR 0.6 [95% CI 0.6-0.7] per 1,000 PY) and atrial fibrillation/flutter (IR 0.6 [95% CI 0.5-0.7] per 1,000 PY). Patients with



The figure shows the cumulative incidences of arrhythmias from day of birth stratified according to type of CHD. CHD: congenital heart disease

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transposition of the great arteries, malformations of the heart chambers, tetralogy of Fallot, and atrioventricular septal defect were at highest risk of arrhythmias (Figure).
Conclusions: Patients with CHD are associated with a significantly higher risk of arrhythmias than the background population. Those with complex CHD are at particular high risk and warrant increased attention.

Keywords: Congenital heart disease, arrhythmias, epidemiology

OP-025
Introduction of the low voltage bridge mapping for the AVNRT RF ablation in children improves outcomes and safety of the procedure

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Background and Aim: The low voltage bridge mapping (LVBM) is an approach to AVNRT ablation that has been postulated in recent years to facilitate and improve safety of the procedure. It has been well described by Drago et al. in combination with cryoablation and proved to be both very effective and safe. In our center AVNRT ablations were performed using traditional approach with use of RF and CARTO mapping system. LVBM in children has been well described with cryoablation but not with RF. LVBM has been introduced in our center from the April 2023 and is now routinely used for all AVNRT RF ablations. The aim of this study was to analyze how the introduction of the LVBM influenced results of the pediatric AVNRT ablations using the RF current.
Method: We performed a retrospective analysis of all AVNRT ablations performed in our center using LVBM (April to November 2023) compared to all AVNRT ablations that were performed using traditional approach, (January 2020 to March 2023). In our center both approaches consist of using CARTO mapping system.
Results: The study group (LVBM) consisted of 19 patients and control group consisted of 43 children. Both groups did not differ significantly with regard to age (median 15.1 years in LVBM group vs 15.1 years in control) and body weight (median 61,5kg in study group vs 60kg in controls). In the LVBM group total time of RF applications was significantly shorter than in the control group: median of 108sec vs 210sec ($p<0.05$). Number of RF applications was smaller in the LVBM group: median of 5vs10 in controls ($p<0.05$). Total procedural time was not significantly shorter in the study group: mean of 71min vs 79min ($p=0.26$). Acute procedural success rate was 100% in LVBM vs 93% in controls ($p=0.55$). There were no complications reported in neither of groups.
Conclusions: The introduction of the LVBM in RF AVNRT ablations in children allows to limit the number of RF applications required and shortens the total RF application time, without prolonging the procedure time. Our experience shows LVBM can be effectively used also in RF ablations of AVNRT.

Table 1

	LVBM			CONTROL			
	n = 19			n = 43			p value
	range	mean	median	range	mean	median	
Age [years]	9.6 – 17.9	13.8	15.1	10.9 - 17.8	15.1	15.1	p = 0.4
Gender M/F	8/11			17/26			p = 0.9
Total RF application time [sec]	59-529	155	108	53-3480	345	210	p <0.05
Number of RF applications	1-16	6	5	1-47	11	10	p < 0.05
Procedure time [min]	50-95	71	70	38-150	79	75	p = 0.26
Acute success rate	100%			93%			P = 0.55
Reported complications	0			0			

Keywords: AVNRT, low voltage bridge mapping, RF ablation

OP-026
Clinical characteristics and outcomes of pediatric patients with long QT syndrome: A korean multicenter cohort study

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Background and Aim: Congenital long QT syndrome (LQTS) is the most common inherited arrhythmic syndrome, which can results in fatal arrhythmia and sudden cardiac death in the young age. Despite numerous studies, reports on Asian patients with long QT syndrome are scarce. The aim of this study was to elucidate the clinical characteristics and outcomes of patients with long QT syndrome from a Korean multicenter study.
Method: We conducted a retrospective multicenter cohort study for the pediatric patients with inherited arrhythmia syndrome, including 16 pediatric cardiology centers in Korea from March 2022 to October 2023. Among the 350 patients with inherited arrhythmia syndrome diagnosed at < 19 years, 227 LQTS patients were identified.
Results: Fifty two percent of patients were male. The mean age of patients at diagnosis was 7.8±5.6 years with a mean follow-up duration of 6.0±5.2 years. Among the 193 patients who underwent genetic testing, 62% had pathogenic or likely pathogenic variants in LQTS-related genes. The most common variant was KCNQ1 (56%), followed by KCNH2 (20%), SCN5A (12%), KCNJ2 (6%), CACNA1C (4%), and CAML2 (1%). Jervell and Lange-Nielsen syndrome were found in 7 patients. Symptoms at diagnosis included unexplained syncope (34%), aborted cardiac arrest (12%), and seizure (9%). Twenty-six percent were

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asymptomatic. An implantable cardioverter-defibrillator (ICD) was implanted in 40 patients (17%) at the age of 11.8±6.9 years for primary prevention in 7 (17%) and secondary prevention in 33 (83%). Appropriate shocks were delivered in 20 patients (50%). Life threatening arrhythmic event (LAE) occurred in 27 patients (12%); aborted cardiac arrest in 8, appropriate ICD shock in 15, ventricular arrhythmia in 8, and sudden cardiac death in 3. Increased LAEs were associated with LQTS 3 (HR 7.897, 95% C.I. 1.711-36.444), female (HR 2.658, 95% C.I. 1.200-5.888), corrected QT interval at diagnosis (HR 1.009, 95% C.I. 1.009-1.019), and aborted cardiac arrest (HR 2.997, 95% C.I. 1.348-6.661).
Conclusions: LQTS patients had a significant risk for LAEs. This study showed LQTS 3, female sex, increased QTc, and aborted cardiac arrest at diagnosis were risk factors for LAE in Korean patients. Longer term follow-up and international collaborative data acquisition are required for predicting outcomes and managing LQTS patients.

Keywords: long QT syndrome, pediatric, sudden death, implantable cardioverter-defibrillator

OP-027
Multisystemic management in children with andersen-tawil syndrome: Beyond the QT prolongation

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Background and Aim: Andersen-Tawil Syndrome (ATS) is a ultra-rare autosomal dominant (AD) condition due to likely pathogenic or pathogenic variant in KCNJ2, coding for potassium channel Kir2.1. Classically ATS is characterized by the triad of QT prolongation/ventricular arrhythmia, susceptibility to episodic flaccid muscle weakness and skeletal abnormalities. Arrhythmic involvement is clinically relevant and represented by long QTc (LQT), ventricular tachycardia (VT), polymorphic VT and premature ventricular contractions (PMCs). Muscular manifestations include weakness and periodic paralysis with vary variable frequency, duration, and severity. At skeletal level affected individuals may present fingers defect as clinodactyly or syndactyly, scoliosis, short stature, and joint laxity. Other less frequent possible manifestations are distinctive facial features, dental anomalies, and mild learning difficulties. Few studies in literature are available regarding multisystemic involvement of ATS in children. The aim of this study is to investigate and describe ATS's clinical implications focusing not only on the cardiac spectrum but also on multiorgan level.
Method: Our tertiary academic paediatric center represents the referral point for the center and south of Italy for the multidisciplinary management of patients with Channelopathies. In this study we include data from 6 patients with ATS who had multisystemic clinical assessment, multiorgan screening and genetic study through (Next Generation Sequencing) sequencing.
Results: We focused on ATS manifestations in children, reporting the main multisystemic signs and symptoms to be considered at diagnosis and during follow up, focusing on the arrhythmic spectrum manifestations and the multiorgan features as muscular, skeletal and dental involvement. We collected data related to clinical and instrumental screening and surveillance: electrocardiogram, 24-Hour ECG holter monitoring, cardiac stress test, echocardiography, clinical symptoms as palpitations or muscle weakness, growth measures, dysmorphic features, etc.
Conclusions: In the management of ATS pediatric population, it is important to consider not only the major signs classically associated with the disease such as arrhythmic spectrum disorders (i.e. QT prolongation) but also other multisystemic aspects that can have a significant impact on prognosis in particular at musculoskeletal level. We underline the importance of the multidisciplinary, personalized, and longitudinal in time approach to children with ATS.

Keywords: Andersen-Tawil Syndrome, QT prolongation, periodic paralysis, multisystemic assessment

OP-028
The correlation between symptoms and event recorder in detecting recurrences in pediatric cases undergoing ablation for supraventricular tachycardia

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Background and Aim: Documentation of supraventricular tachycardia (SVT) can be challenging in pediatric age group complaining with palpitations. In this particular group, who are learning to perceive and describe symptoms, long-term monitoring applications such as event recorder devices are also possible to document tachycardia burden.The aim of this study was to define significance of event recorders in detecting symptom/recurrence correlation after SVT ablation.
Method: Among 1682 patients who underwent successful SVT ablation at our center between 2013 and 2023, the following cases were included in the study: patients 1) with atrioventricular nodal reentry tachycardia (AVNRT), concealed accessory pathway (cAP), and focal atrial tachycardia (FAT) after excluding those identifiable on surface ECG as Wolf-Parkinson-White and Mahaim groups; 2) without congenital heart disease; 3) above 10 years of age capable of describing symptoms; and 4) presenting with palpitation post-ablation without SVT detected on ECG and 24- ambulatory Holter monitoring but with event recorder indication.
Results: A total of 706 patients were included in the study (487 AVNRT, 171 Concealed AP, 48 FAT). Among a total of 104 patients reporting palpitations (15%), event recording data of 73 patients was available, and SVT was documented in 8 (8/73, 11%) cases (4 AVNRT, 1 Concealed Ap and 3 FAT). Sinus tachycardia was detected on event recording when symptoms were reported in 9 (9/73, 12%) patients. Analysis based on subgroups revealed that in symptomatic patients requiring event recorder, SVT burden were detected in 0.8% of the total AVNRT group, 0.5% of the total concealed AP group, and 6% of the total FAT group.

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Conclusions: In symptomatic cases with SVT substrates like AVNRT, concealed AP, and FAT,that may not be detected by ECG and Holter monitoring, though the symptom/record correlation appears low, the use of event recorder is recommended in clinical practice for detecting recurrent SVT episodes after initial ablation in individuals when ECG and Holter alone are indecisive.

Keywords: ablation, event recorder, tachycardia,

OP-029

Catheter ablation of tachyarrhythmias in patients with atrial redirection surgery for d-transposition of the great arteries – 22 years of experience

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Background and Aim: Dextro-transposition of the great arteries (d-TGA) is the second most common cyanotic congenital heart disease. Mustard- and Senning-type (M/S) atrial redirection surgery was the standard palliation until the late 1980's. Long-term follow up is typically complicated by sinus node dysfunction, failure of the subaortic right ventricle and atrial tachyarrhythmias.
Method: All patients (pts) with d-TGA after M/S surgery who underwent catheter ablation for any tachyarrhythmia between 2001 and 2023 were included. Spectrum of arrhythmias, success rates of catheter ablation and complications were analyzed.
Results: In 98 pts (34 females (34.7%); mean age at first procedure 33.1 y; Mustard-type surgery 51 (52.0%)) 145 ablation procedures were performed. A total of 190 tachyarrhythmias were identified, 4 (2%) of ventricular and 186 (98%) of supraventricular origin (55 (38%) cavotricuspid isthmus (CTI)-dependent reentrant tachycardia, 3 (2%) non-CTI-dependent reentrant tachycardia, 66 (46%) non-sustained atrial tachycardia, 35 (24%) non-automatic focal atrial tachycardia, 7 (5%) focal atrial tachycardia, 3 (2%) atrial fibrillation, 2 (1%) junctional ectopic tachycardia, 15 (10%) AV-nodal reentrant-tachycardia). Clinically documented tachycardia was non-inducible in 26 procedures (18%). Catheter ablation was acutely successful in 105/124 (84.7%) interventions with identified tachycardia-mechanism, -substrate and -localization. Within a mean follow up of 6.1 y [0.06 – 20.2 y] 97 clinical tachyarrhythmia recurrences were observed, 10 (9.5%) of which were identical to the previously targeted tachyarrhythmia in 39 re-do procedures. There were 6 (4.1%) major procedure-related complications (2 femoral artery aneurysms, 2 arterio-venous fistulas, 1 hematuria, 1 sinus node dysfunction likely procedure related with subsequent pacemaker implantation). Additionally, intraprocedural development of fast ventricular tachycardia led to hemodynamic compromise entailing short cardio-pulmonary resuscitation with good clinical outcome in two procedures.
Conclusions: We present one of the largest series of catheter ablations in patients with d-TGA after M/S surgery. CTI-dependent macro reentrant tachycardia was the predominant arrhythmia. Catheter ablation has shown to be feasible with high success and low complication rates despite challenging postoperative electro-anatomical preconditions.

Keywords: dextro-transposition of the great arteries (dTGA), Mustard, Senning, catheter ablation

OP-030

The impact of vitamin d supplementation for PVCS with an RVOT morphology compared to pvcs with an LV fascicular morphology

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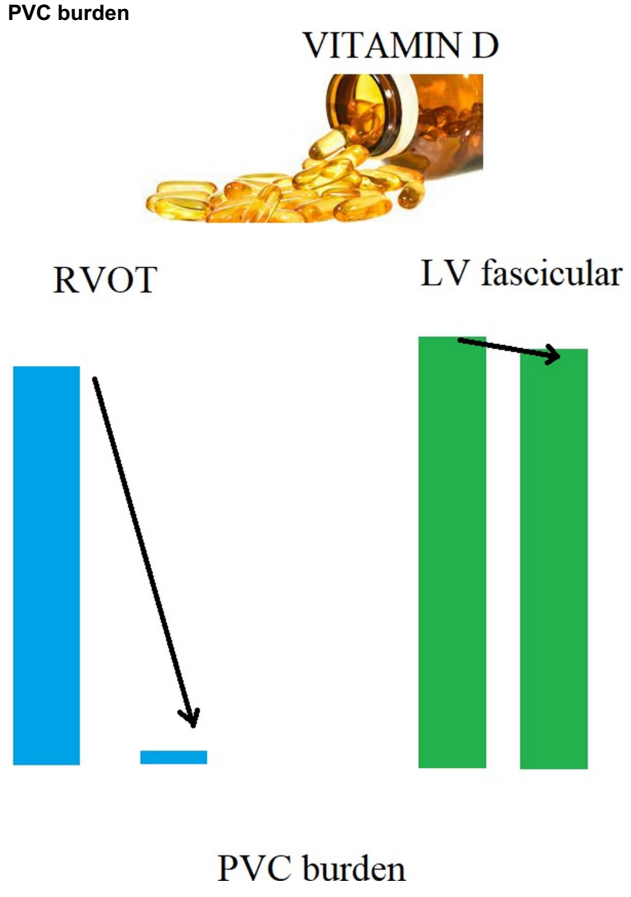
Background and Aim: Outflow tract PVCs are benign arrhythmias that usually occur in individuals without any structural heart disease. In some children and adults PVCs have been demonstrated to be associated with vitamin D deficiency. In our study we examined the efficacy of vitamin D supplementation in children with high burden PVCs and distinct ECG morphologies: RVOT and LV fascicular.
Method: We included 46 patients (mean age 10.6+/-4,1 years, 26% females) with high burden monomorphic PVCs (mean=18925/24 hours) and vitamin D deficiency. We compared 36 patients with RVOT morphology with 10 patients with LV fascicular morphology. RVOT morphology was characterized by an LBBB pattern, inferior frontal axis, and precordial transition to V3–V4 and QRS duration >150 ms. (n=36). LV fascicular morphology was characterized by an RBBB pattern with rsR' in lead V1, left axis (n=8) or right axis (n=2).and QRS duration of 100-110 ms.
Results: In the RVOT morphology group, mean age was 12,7 +/-2.7 years 75% males, with a PVC burden of 18343.7+/-13836.2 and 25OHvitamin D level of.23.5+/-9.4 ng/ml. After 3 month of supplementation with oral vitamin D there was an increase in the level of vitamin D to 41.6+/-6.3 with significant decrease of PVC burden to 3628.0 +/-2347.2. Overall there was 80% decrease in PVC burden in this group of children. In the LV fascicular morphology group, mean age was 6.9+/-5.5 years,71% males, with a PVC burden of 20535.3+/-20867.9. and vitamin D level of 25.8 +/-7.1ng/ml. After 3 month of supplementation with oral vitamin D, there was an increase in the level of vitamin D to 65.8+/-42.8 ng/ml but without significant change in PVC burden 19207.1+/-22807.8.
Conclusions: PVCs with RVOT morphology can effectively be treated with vitamin D supplementation in case of vitamin D deficiency. Since vitamin D deficiency is a pandemic. vitamin D should be considered before antiarrhythmic drugs or catheter ablation in children with monomorphic high burden PVCs.

Keywords: PVCs, vitamin D, outflow tract, fascicular

PVC burden



General Cardiology

OP-031

Adverse cardiac events among children, adolescents, and young adults with psychopharmacological treatment in sweden - descriptive study

Howaida Elmowafi[1], Estelle Naumburg[1], Jenny M Kindblom[2], Linda Henriksson[3] and David Gyllenberg[4]

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Background and Aim: Psychotropic medicines are associated with life threatening adverse cardiac events, but the number of these events in young people and in real clinical setting is unknown. In Sweden, an increased prescription rate of psychotropic medications to young people occurred at the same time as there has been an increase in the number of young people diagnosed with cardiac related symptoms. The aim of this study was to assess the type and proportions of life-threatening adverse cardiac events in young people exposed to psychotropic medicines in Sweden.

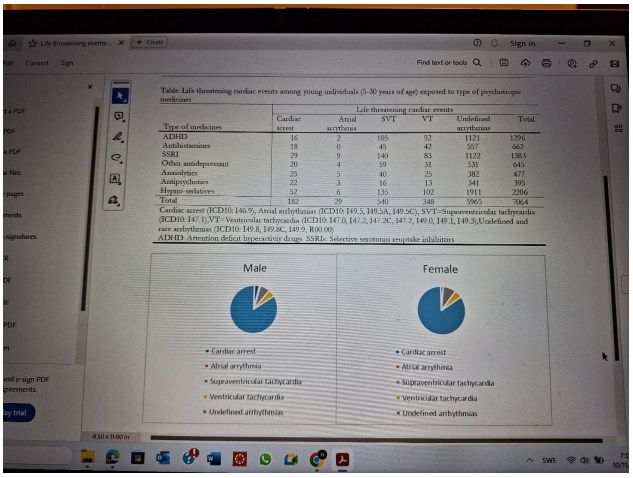
Method: Individuals aged 5-30 years old who received at least one prescription of psychotropic drug (ADHD, antihistamines, SSRIs, other antidepressants, anxiolytics, antipsychotics, and hypno-sedatives) between 2006 to 2018 were retrieved from Swedish drug registers. From patient registers, at least one episode of cardiac arrest with successful resuscitation, atrial arrhythmia, supraventricular or ventricular tachycardia, and undefined arrhythmia as a main diagnosis was considered a potential adverse life-threatening event. Information on death was retrieved separately from the Swedish death register. By linkage between the registers using the personal identification number, descriptive statistics determined type of event, event proportion in relation to type of psychotropic medicines and stratified by age and gender.

Results: Over 1, 600 000 individuals, aged 5-30, were exposed to psychotropic drugs between 2006-2018. Among them 7064 adverse life-threatening cardiac events occurred. The proportions of these events were; cardiac arrest with successful resuscitation 2.6%, atrial arrhythmias 0.4%, supraventricular tachycardia 7.6%, ventricular tachycardia 5% and undefined arrhythmias 84.4% which were more common in males. Hypno-sedatives medicines had the highest event prevalence for adverse cardiac events, followed by SSRIs and ADHD drugs, especially in participants aged 20-30 years. There were eight cases (0.0005%) with sudden cardiac death, and this was seen among the individuals exposed to SSRIs and hypno-sedatives followed by ADHD drugs, mainly in males.

Conclusions: Life-threatening cardiac events may occur among individuals aged 5-30 years exposed to psychopharmacological treatment, especially among those with SSRIs, hypno-sedatives, and ADHD drugs. Further research in real clinical settings is warranted to assess the risk to cardiac events linked to psychotropic drugs.

Keywords: ADHD, SSRIs, children, Young people, Anxiolytics, hypnotics

Life threatening cardiac events among the exposed to psychopharmacological treatments aged 5-30 years in Sweden during 2006-2018.



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Paediatric Cardiovascular Intensive Care

OP-032
Peri-procedural changes in lung ultrasound score following pediatric patent ductus arteriosus ligation

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Background and Aim: Patent ductus arteriosus (PDA) is a common congenital heart condition in neonates and infants, often requiring percutaneous/surgical closure when hemodynamically significant. Peri-procedural management and monitoring of these patients is crucial due to related comorbidities. This study investigates peri-procedural changes in lung ultrasound score (LUS).
Method: An observational and retrospective analysis was conducted. Neonates and infants admitted to a Neonatal and Pediatric Intensive Care Unit from 2013 to 2023 with clinically significant PDAs that underwent successful closure were included. Echocardiographic criteria comprised transductal diameter>1.5mm with unrestrictive pulsatile left-to-right flow and reverse end-diastolic flow in the descending aorta. Exclusion criteria were major structural heart disease. Clinical, analytical, and echocardiographic data were collected at several time-points. Lung ultrasound was performed by a single operator. Post-PDA ligation syndrome was defined as the need for cardiovascular support due to hemodynamic compromise within 24h post-ligation.
Results: This study included 20 patients (55% male): 6 term and 14 preterm babies, with a median gestational age of 27 weeks and median age at procedure of 28 days. Median birth weight and median weight at the procedure were 980g and 1525g, respectively. Eight infants were initially managed pharmacologically. Two of these received a second course. Eighteen patients underwent surgical ligation, and two percutaneous closure. Trends in cardiorespiratory stability indices and LUS are shown in Table 1. Post-interventional LUS were lower than pre-interventional LUS, reaching a minimum score post-72h (vs. 6-12h ($p<0.001$), 24h ($p<0.001$), 48h ($p<0.001$) and 72h ($p<0.001$)). Differences in stroke volume were less consistent (vs. 6-12h ($p=0.012$) and 48h ($p<0.001$)). Four patients developed post-PDA ligation syndrome and were managed with fluid support and dopamine. These patients presented significantly higher LUS pre-procedure and 6-12h post procedure than others, respectively (17 vs. 9, $p=0.022$; 16 vs. 9, $p=0.029$), however no significant differences were observed in age or weight at procedure.
Conclusions: We identified a significant decrease in LUS post-PDA ligation, particularly 6-12h post-procedure. Additionally, patients developing post-PDA ligation syndrome exhibited significantly higher LUS pre-procedure and 6-12h post-procedure. These findings suggest that lung ultrasound may be a valuable tool in recognizing early indicators of hemodynamic compromise and monitoring pulmonary disease following PDA ligation.

Keywords: Patent Ductus Arteriosus, ligation, Pediatric Intensive Care Unit, Lung Ultrasound Score, Post-PDA ligation syndrome

Table 1

Table 1. Changes in peri-procedure cardiorespiratory stability markers and LUS subsequent to PDA ligation.

	Baseline		1-6h	6-12h	24h	48h	72h
FiO2 - median (IQR)	0.24 (0.21-0.30)		0.30 (0.25-0.40)	0.28 (0.25-0.30)	0.23 (0.27-0.21)	0.25 (0.21-0.30)	-
MAP, mmHg - median (IQR)	47 (41-52)		53 (40-59)	54 (45-64)	57 (48-62)	52 (42-70)	-
SBP, mmHg - median (IQR)	64 (53-76)		67 (57-86)	66 (57-84)	72 (65-85)	70 (57-96)	-
pH - median (IQR)	7.36 (7.30-7.43)		7.33 (7.27-7.39)	7.38 (7.30-7.41)	7.39 (7.36-7.44)	7.40 (7.26-7.46)	-
Base deficit, mmol/L - median (IQR)	3.80 (1.88-9.23)		1.60 (0.70-8.20)	3.25 (-1.65-8.83)	2.00 (-2.20-9.80)	5.25 (-0.98-7.98)	-
Lactate, mmol/L - median (IQR)	1.30 (0.93-1.60		0.90 (0.60-1.80)	1.15 (0.75-2.93)	0.90 (0.70-1.60)	0.80 (0.63-1.15)	-
VTI, cm - median (IQR)	13 (11-14)		12 (10-13)	12 (10-14)	14 (11-15)	15 (14-16)	-
Cross sectional area of LVOT, cm^2 - median (IQR)	0.16 (0.15-0.22)		-	-	-	-	-
Stroke volume, mL - median (IQR)	2.10 (1.82-2.78)		1.96 (1.66-2.51)	2.05 (1.66-2.51)	2.13 (1.84-2.70)	2.49 (2.13-3.42)	-
p value		vs.	0.012	0.243	0.050	<0.001	-
LUS - median (IQR)	13.00 (12-14)		13 (12-14)	12 (9-15)	8 (7-11)	6 (5-8)	3 (2-4)
p value		vs.	0.070	<0.001	<0.001	<0.001	<0.001

FiO2: Fraction of Inspired Oxygen; IQR: Interquartile Range; LVOT: Left Ventricle Outflow Tract; LUS: Lung Ultrasound Score; MAP: Mean Arterial Pressure; SBP: Systolic Blood Pressure; VTI: Aortic Velocity Time integral

Changes in peri-procedure cardiorespiratory stability markers and LUS subsequent to PDA ligation.

Cardiac Imaging

OP-033
Identifying high-risk thoracic aortopathy using positron emission tomography: The aortas study

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Background and Aim: In patients with bicuspid aortic valve, the majority of aortic dissections occur below aortic diameter thresholds that trigger prophylactic surgical repair. There is therefore an urgent unmet need to develop novel methods for better risk stratification. In thoracic aortopathy, vascular smooth muscle cell stress and extracellular matrix remodelling results in microscopic calcification deposition on elastin fibres which later disappears with loss of elastic fibres. We aimed to establish whether positron emission tomography (PET) using 18F-sodium fluoride – a radiotracer than binds to vascular microscopic calcification - could detect high-risk patients who may benefit from early surgery.
Method: In a prospective cohort imaging study, patients across Scotland diagnosed with bicuspid aortic valve and age- and sex-matched control volunteers underwent baseline hybrid PET-CT (Biograph mCT, Siemens) followed immediately by PET-MRI (3.0T Biograph mMR, Siemens) after injection of 250 MBq 18F-sodium fluoride. Baseline maximum ascending aortic microcalcification activity was measured as maximum standardised uptake value adjusted for blood pool activity. Ascending aortic diameter was quantified at the level of the right pulmonary artery. In patients, a repeat PET-MRI was performed at 12-24 months. Annualised change in ascending aortic size index (diameter/body surface area) was calculated. Annualised change in maximum aortic microcalcification activity was calculated and stratified into tertiles.

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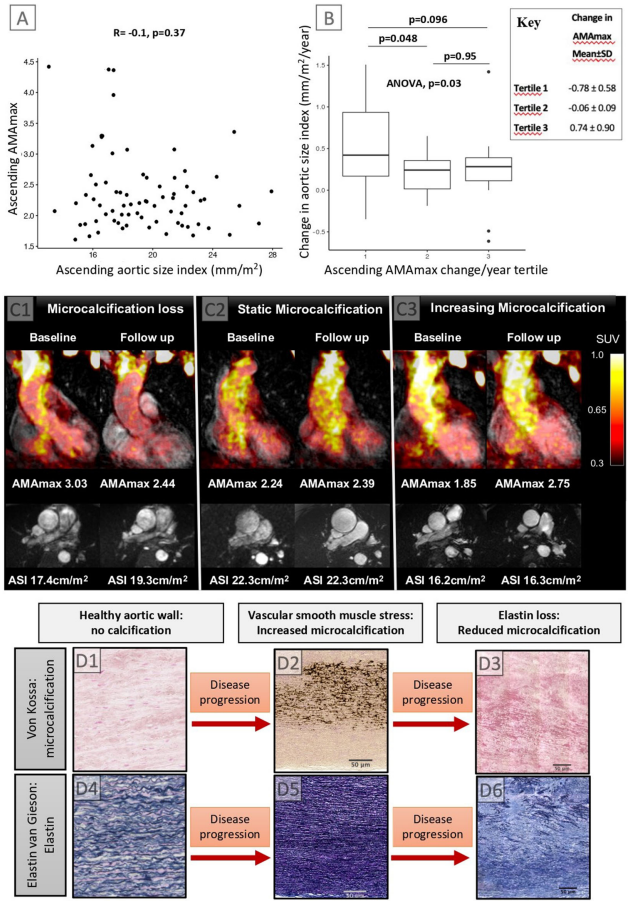
Representative ascending aortic wall histological samples were stained for microcalcification (von Kossa) and elastin (elastin van Gieson).

Results: Seventy-five patients with bicuspid aortic valve (52.5±7.5 years, 24% female) and 18 control subjects (50.6±6.2 years, 27% female) were recruited. The maximum aortic microcalcification activity was higher in patients than control subjects (2.41±0.69 versus 1.94±0.35, P<0.01). In patients, baseline maximum aortic microcalcification activity was not associated with baseline aortic diameter (p=0.37). Those with a loss of aortic microcalcification activity (tertile 1) experienced the greatest change in diameter (0.52±0.51 mm/m^2/year) compared to those with static (tertile 2, 0.21±0.21 mm/m^2/year) or increasing activity (tertile 3, 0.25 ±0.42 mm/m^2/year; p=0.03).

Figure



(A) A scatterplot of bicuspid aortic valve cohort demonstrating no association between baseline ascending maximum aortic microcalcification activity (AMAmax) and ascending aortic size index. (B) Bicsupid aortic valve patients were split into tertiles based on the annualized change in maximum aortic microcalcification activity AMAmax; Tertile 1 – reducing microcalcificaiton (elastin loss phase); Tertile 2 – static microcalcification; Tertile 3 – increasing microcalcification (see key for detailed data). Those who lost microcalcification activity had the most rapidly expanding aneurysms. (C1-3) Representative PET-MRI (coronal view of ascending aorta, top row of images) and MRI (axial view at level of right pulmonary artery, bottom row) examples from each tertile comparing maximum aortic microcalcification activity and aortic size index at baseline (right) and at 12-24 months (left). (D) Demonstrating representative microcalcification (von Kossa, D1-D3) and elastin (Elastin van Gieson, D4-D6) histological examples from healthy (D1+D4), mild/moderate aortopathy (vascular smooth muscle cell stress drives microcalcification deposition onto elastin fibres as part of remodelling, D2+D5), and severe aortopathy (elastin loss results in significant reducing in overall aortic wall microcalcification, D3+D6).

Conclusions: 18F-Sodium fluoride PET can detect and track microscopic calcium deposition within the aortic wall of patients with bicuspid aortic valve. Late loss of aortic microcalcification activity over time is a concerning feature and appears to identify those at greatest risk of disease progression.

Keywords: bicsupid aortic valve, aortopathy, positron emmision tomography, MRI, CT, dissection

OP-034
Early detection of arvc phenotype in family screening of ARVC: A CMR study in pediatric population

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Unité médico-chirurgicale de cardiologie congénitale et pédiatrique, centre de référence des maladies cardiaques congénitales complexes - M3C, Hôpital universitaire Necker-Enfants Malades, Université de Paris, France

Background and Aim: Early detection of familial Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC) is a challenging task. Novel imaging techniques represent an incremental value in ARVC family screening. Feature- tracking cardiac magnetic resonance (FT-CMR) allows quantitative global and regional myocardial strain, which has been shown to increase diagnostic value for ARVC disease detection. However, the value of FT-CMR derived strain in family screening remains unknown. The purpose of this study is to evaluate the role of FT-CMR in early detection of patients that will develop the disease according to task force (TF) criteria during follow up.

Method: First-degree relatives of patients with ARVC were evaluated according to 2010 TF criteria. We analysed retrospectively the serial CMR studies and we included 68 patients. CMR data were: left ventricle (LV) and right ventricle (RV) 's morphological values, RV and LV FT derived strain (global longitudinal/circumferential strain, GLS/GCS). We have considered data at baseline (T0) and at second evaluation (T2) during the follow up. Disease progression was defined as the development of new 2010 TF criteria during follow up that was absent at baseline (phenotype – vs phenotype +).

Results: The mean age at T0 was 13 +/- 2.8 years, 36.7% of patients were carriers of familial mutation. 42 patients underwent a second evaluation. At baseline there was a significant statistical difference in terms of RV deformation parameters between children with no progression to disease (phenotype -) to patients with transition to ARVC (phenotype +).We found difference in terms of the endocardial GCS, myocardial myo-GLS and endocardial GLS, respectively: -14,9 +/- 4,1 vs 11,14 +/- 2,4 (p 0.03); -24,6 +/-4,5 vs 19,9 +/- 6,4 (p 0.02); -25,8 +/- 8.8 vs 16,2 +/-1,7 (p 0.03).

Conclusions: We found reduced strain values at first CMR between the group of patient that developed the disease compared to the group of patient with a negative phenotype. FT-CMR can allow early detection of first degree relatives at risk of developing the disease. If confirmed in a larger prospective study, it could be useful to stratify the risk of life threating events and the recommendation for physical activity during childhood and adolescence.

Keywords: Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC) Familial screening Genetic CMR Global longitudinal strain

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OP-035

Use of beta-blockers and angiotensin receptor-inhibitors in paediatric bicupsid aortic valve with ascending aorta dilatation: An option for the future

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Background and Aim: Bicuspid aortic valve (BAV) confers a higher risk for ascending aorta (AscAo) dilatation and dissection, which may be secondary to the damaging impact on the aortic wall of abnormal flow dynamics but could also be due to intrinsic increased wall strain. Betablockers (BB) and angiotensin receptor-inhibitors (ARI) could become an option to decrease wall shear stress and therefore potentially decrease dilatation progression. Our aim was to assess whether aorta biomechanics by echocardiography are altered in BAV paediatric patients compared to matched controls and whether treatment has a short-term impact on biomechanics.

Method: Patients were prospectively included from our aortopathy clinic. Treatment, BB or ARI, was offered if ascending aorta dilatation (≥3 z-score) was found. Aortic biomechanics (impedance, elasticity and stiffness indexes, beta-modulus, pulse wave velocity and strain) were assessed by echocardiography before and 3-months after treatment.

Results: We included 77 children, 59 BAV (36 had AscAo dilatation), and 18 controls. Significant aortic regurgitation in 36/59 patients, who showed higher AscAo diameters by z-score ($p=0.011$). Input and characteristic impedances were significantly lower in BAV patients ($p<0.001$ and 0.007), and in dilated compared to non-dilated BAV ($p<0.001$). Elasticity and biomechanical strain did not show higher stiffness on the affected nor dilated BAV subjects. Pulse wave velocity (PWV) was significantly increased in the dilated-BAV compared to the non-dilated BAV($p=0.017$), controls also showed higher PWV than BAV-subjects ($p<0.011$). Speckle tracking strain did not differ amongst groups. Those treated with BB or ARI had a significant increase in characteristic impedance ($p=0.025$). When compared against those without treatment there was a decrease on impedance ($p=0.021$) and stiffness ($p=0.030$), with a trend towards improvement on elasticity parameters. Characteristic impedance differences were also observed if only analysing dilated patients ($p=0.022$).

Conclusions: The increased stiffness parameters in the BAV-aortic wall are both related to the ascending aorta dilatation and the disease itself, with higher impact on the latter. Treatment with BB or ARI improves impedance and stiffness indexes. Their use in BAV with enlarged AscAo could be a potential strategy to diminish the risk for ascending aorta dissection. Further research to assess haemodynamic changes after treatment is mandatory.

Keywords: Bicuspid aortic valve, ascending aorta dilatation, beta-blockers, angiotensin receptor-inhibitors, stiffness, elasticity.

Bicuspid aortic valve biomechanic changes after treatment and group differences

BIOMECHANICAL PARAMETERS AND STRAIN DIFFERENCES BETWEEN GROUPS

BIOMECHANIC AND HAEMODYNAMIC PARAMETERS	BAV vs CONTROLS				BAV. DILATED vs NON-DILATED				NON-DILATED BAV vs CONTROLS
	BAV	CONTROLS	*p-value*	EFFECT INTENSITY	DILATED BAV	NON-DILATED BAV	*p-value*	EFFECT INTENSITY	*p-value*
INPUT IMPEDANCE	0.045 +/- 0.024	0.084 +/- 0.029	**<0.001 ***	0.765	0.034 +/- 0.0179	0.0612 +/- 0.022	**<0.001***	0.698	**0.005***
CHARACTERISTIC IMPEDANCE	280.637 +/- 356.526	448.082 +/- 398.057	**0.007 ***	0.423	188.720 +/- 133.770	420.510 +/- 517.478	0.125	0.241	0.165
ELASTIC MODULUS (YOUNG)	558.980 +/- 713.386	450.593 +/- 272.232	0.525	0.103	551.997 +/- 730.382	569.163 +/- 703.270	0.896	0.021	0.557
AORTIC BIOMECHANICAL STRAIN	15.668 +/- 9.587	11.852 +/- 4.974	0.101	0.220	15.619 +/- 9.675	15.739 +/- 9.664	0.994	0.002	0.229
BETA STIFFNESS INDEX	5.895 +/- 0.856	5.923 +/- 0.535	0.525	0.103	5.877 +/- 0.850	5.912 +/- 0.863	0.896	0.021	0.557
BETA 2 LOGARITMIC STIFFNES INDEX	-0.105 +/- 0.028	-0.102 +/- 0.020	0.950	0.012	-0.101 +/- 0.027	-0.112 +/- 0.029	0.163	0.227	0.35
AORTA DISTENSIBILITY INDEX	0.007 +/- 0.005	0.006 +/- 0.003	0.794	0.043	0.007 +/- 0.006	0.007 +/- 0.004	0.742	0.052	0.713
PWV WHOLE THORACIC AORTA	520.849 +/- 433.924	639.404 +/- 682.000	0.777	0.046	470.006 +/- 250.282	598.497 +/- 619.920	0.623	0.080	0.604
PWV PLAX ASCENDING AORTA	665.584 +/- 256.333	885.875 +/- 550.000	**<0.011***	0.414	717.373 +/- 2512.049	562.902 +/- 730.841	**0.037***	0.400	**0.021***
SPECKLE TRACKING STRAIN ASCENDING AORTA	-27.44 +/- 15.22	-23.660 +/- 11.540	0.507	0.105	29.114 +/- 17.035	24.780 +/- 11.658	0.247	0.127	0.685

EVOLUTION OF BIOMECHANICAL PARAMETERS AND STRAIN AFTER TREATMENT

BIOMECHANIC AND HAEMODYNAMIC PARAMETERS	TREATED PRE vs POST			TREATED vs NON-TREATED	DILATED TREATED vs NO-TREATED
	PRE-TREATMENT	AFTER TREATMENT	*p-value*	*p-value*	*p-value*
INPUT IMPEDANCE	0.029 +/- 0.009	0.037 +/- 0.024	0.274	**0.021***	0.731
CHARACTERISTIC IMPEDANCE	197.196 +/- 173.673	420.005 +/- 537.496	**0.025***	0.137	**0.023***
ELASTIC MODULUS (YOUNG)	667.941 +/- 982.143	368.437 +/- 182.144	0.597	0.966	0.731
AORTIC BIOMECHANICAL STRAIN	15.637 +/- 11.258	13.019 +/- 6.419	0.86	0.371	0.329
BETA STIFFNESS INDEX	5.961 +/-0.988	5.792 +/- 0.507	0.669	0.966	0.731
BETA 2 LOGARITMIC STIFFNES INDEX	-0.094 +/- 0.027	-0.093 +/- 0.020	0.903	**0.030***	0.133
AORTA DISTENSIBILITY INDEX	0.006 +/- 0.004	0.007 +/- 0.006	0.495	0.986	0.832
PWV WHOLE THORACIC AORTA	509.634 +/- 300.241	624.843 +/- 456.495	0.664	0.197	0.187
PWV PLAX ASCENDING AORTA	1654.950 +/- 4092.113	625.792 +/- 920.980	0.577	0.849	0.093
SPECKLE TRACKING STRAIN ASCENDING AORTA	-26.002 +/- 11.223	-23.461 +/- 18.935	0.252	0.552	0.934

Mean +/- SD. Differences by U Mann Whitney Test. Paired T student Test
** = statistically significant*
BAV=bicuspid aortic valve; PWV= pulse wave velocity

Biomechanics and strain parameters differences after treatment and comparisons between groups

Paediatric Cardiovascular Intensive Care

OP-036

Cerebral autoregulation monitoring after cardiopulmonary bypass surgery for congenital heart disease in the paediatric intensive care unit

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Background and Aim: Despite improved survival rates in infants with congenital heart disease, many patients suffer from impaired neurological outcome. Dysfunctional cerebral autoregulation may lead to a loss of this protective mechanism and an increased risk for hyper- and hypoperfusion, with consequent risks for ischaemia, haemorrhage and delirium. Infants with congenital heart disease undergoing cardiac surgery with cardiopulmonary bypass and intensive care unit stay face a cumulative risk of impaired autoregulation. Current blood pressure recommendations are largely based on empirical data and optimal management is poorly established.

Method: Autoregulation was prospectively monitored in 83 neonates and infants following open-heart surgery with cardiopulmonary bypass after admission to the paediatric intensive care unit. Autoregulation indices were determined using near-infrared spectroscopy, correlating regional cortical oxygen saturation and local haemoglobin levels with invasive mean arterial pressure to calculate cerebral oxygenation index (COx) and haemoglobin volume index (HVx). Intact autoregulation was defined as $COx < 0.4$ and $HVx < 0.3$.

Results: The mean age of the patients was 111.6 ± 85.3 days and the mean weight was 5.0 ± 1.8 kg. The optimal mean arterial blood pressure for autoregulation was 56.3 ± 7.6 mmHg (COx) and 54.8 ± 6.6 mmHg (HVx). The mean lower limit of autoregulation was 45.7 ± 6 mmHg (COx) and 46.0 ± 6.5 mmHg (HVx). The mean upper limit was 64.6 ± 9.1 mmHg (COx) and 64.7 ±

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7.7 (HVx). The LLA, MAPopt and ULA could be determined in 66.27%, 95,18% and 78.31% by HVx and in 75.9%, 95.18% and 61.45% by COx. During 84.1 ± 8.2% of the monitored time, the COx was below 0.4, and during 77.16 ± 9.83% of the monitored time, the HVx was below 0.3, representing the time with intact cerebral autoregulation.
Conclusions: Cerebral autoregulation is individual, and the autoregulation parameters identified show a narrow range between lower and upper limits, together with significantly higher optimal mean arterial pressures compared with empirical blood pressure recommendations. Therefore, individualized real-time monitoring may be required to identify specific blood pressure targets to optimize autoregulation and prevent neurological injury.

Keywords: Congenital heart defect, Cerebral autoregulation, NIRS, Cardiopulmonary bypass

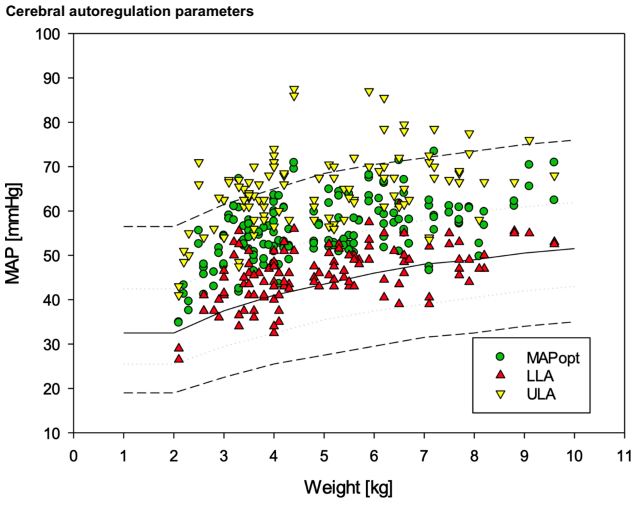

Figure 2: Illustration of all measured LLA, MAPopt and ULA. The empirical blood pressure data from de Graaff (2016) are plotted in the background. MAP: Mean arterial pressure, MAPopt: optimal MAP regarding to intact autoregulation, LLA: Lower limit of autoregulation, ULA: Upper limit of autoregulation. DE GRAAFF, J. C., PASMA, W., VAN BUUREN, S., DUIJGHUISEN, J. J., NAFIU, O. O., KHETERPAL, S. & VAN KLEI, W. A. 2016. Reference Values for Noninvasive Blood Pressure in Children during Anesthesia: A Multicentered Retrospective Observational Cohort Study. Anesthesiology, 125, 904-913.

General Cardiology

OP-037
Pediatric cardiac care – from developing countries to a cardiac reference centre in portugal – a 11 year experience

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Background and Aim: Globally about 1.35 million children are born with congenital heart disease (CHD) each year. Although most are expected to be treated within a defined period, in developing countries an early diagnosis and timely intervention is only possible in a minority of cases. Through bilateral agreements, Portugal and its ex-colonies have a well-established evacuation program, in which our Centre participates actively. Children are screened either through telemedicine or on-site missions, and then transferred for surgical or percutaneous management. We aim to describe and analyse our centre's experience in the evacuation of children with cardiac disease over the past 11 years.
Method: The authors carried on an observational and retrospective study over an 11 year period (2013 to 2023), which included paediatric patients with cardiac disease. Demographic data, type of heart disease and treatment, time until intervention and until discharge, and mortality rate were analysed.
Results: A total of 468 paediatric patients were included, 249 were female, the medium age was 5.7 years. Patients were mostly from Cape Verde (191) and from Guinea Bissau (186). The number of evacuated patients has been increasing throughout the years. Two hundred and twenty-eight patients had shunt related diseases, of which 28% were patent arterial duct, 32% interventricular septal defects, 15% atrial septal defects, 14% endocardial cushion defects and 12% had a combination of shunt defects. Eighty-four patients had cyanotic heart disease of which 56% were Tetralogy of Fallot. Twenty-seven patients had obstructive CHD, mostly pulmonary stenosis (59%). Of the 95 patients with acquired heart disease, 98% had valvular rheumatic heart disease. Most patients received surgical treatment (57%), 3% had both surgical and percutaneous interventions, and 18% were treated percutaneously. Of these interventions, 7.6% were palliative. Overall cardiac mortality rate was 3.6%, and 1 month after procedure was 2.19%. Medium time from arrival to the intervention was 2.2 months and medium time of discharge was 2.9 months.
Conclusions: Our Centre has a well-established program in the treatment of evacuated children with cardiac disease which has proven to be beneficial, ultimately improving the quality of life of almost 500 children. An appropriate screening is a vital for the program effectiveness.

Keywords: congenital heart disease, heart surgery, pediatric cardiac care, pediatric cardiology

Education/Training

OP-038
Global partnership to support the incorporation of cardiovascular screening in the national school health program in a low-income country

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Background and Aim: Cardiovascular diseases are among the most important causes of poor performance at school, but are not addressed in school health programs. We designed a program to strengthen Mozambique's national school health program to improve awareness, increase knowledge, promote cardiovascular screening, and support the prevention and control of rheumatic fever (RF) and rheumatic heart disease (RHD).
Method: The study was implemented in Maputo City between April/2021 and March/2023. Training materials were delivered in a 3-hour theoretical and practical training module to primary

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school teachers, that included rationale for the project, natural history of RHD, preventive measures and pathways for referral. Teachers educated grades 1-5 children who learned using colouring books in the classrooms 1-2 times/week for 10 minutes for a period of 1 year; children were stimulated to complete the activity with their caregivers at home. Class prefects and interactive students from grade 6-7 (3 per class) were further trained in a 30-45 min session to use a guidebook and become peer-educators through facilitating educational activities using RHD walls painted in each school. Finally, experienced patients living with RHD participated on education workshops to be able to support the team in school-based activities, and provide peer-support to children found to have RHD, including workshops with their parents.
Results: Teachers: 672 were trained in 11 training sessions on Saturdays, and delivered their training at least ten minutes/week. Children: 58,303 were educated: using colouring books 40 minutes/month (38,948), through peer education using the outdoor painted walls (19,355) and peer-educators trained in 28 sessions scheduled for morning and afternoon students (755). Patients Ambassadors: 4 trained patients living with RHD supported teacher training sessions, conducted educational lectures and talks at the community level. Parents: 291,515 family members were indirectly reached through discussions around the colouring books and also through community lectures.
Conclusions: Colouring books, RHD walls and peer-educators facilitation created an enabling environment to support RF/RHD education in a friendly and sustainable way. By engaging teachers, parents and experienced patients our strategy reached the communities, allowing the share of RF/RHD preventive.

Keywords: Rheumatic Fever, Rheumatic Heart Disease, Education

Congenital Heart Surgery

OP-039
Cardiac output drop reflects circulatory attrition after fontan completion: Serial cardiac magnetic resonance study

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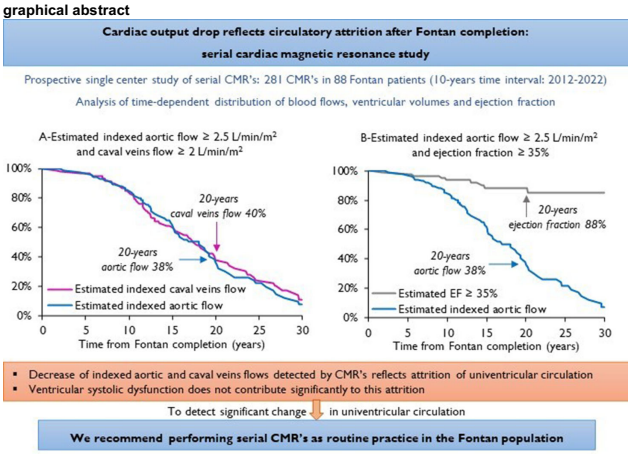
Background and Aim: Cardiac magnetic resonance (CMR) imaging is a main diagnostic tool in follow-up of Fontan patients. However, the value of serial CMR's for evaluation of Fontan attrition is unknown. This is a prospective study of serial CMR's in patients after Fontan completion. The aim of this study is the analysis of time-dependent evolution of blood flow distribution, ventricular volumes and function.
Method: Prospective single centre study of 281 CMR's (between 2012-2022) in 88 Fontan patients with distribution of blood flows, measurements of ventricular volumes, ejection fraction and heart rate. Since 2012 in our department Fontan patients from the age of 7 years old, underwent CMR with standardized flow measurements as part of regular follow-up at intervals of 2-3 years. Linear mix model regression for repeated measurements was used to analyse changes of measurements across serial CMR's. The survival curve for all indexed flows, volumes, ejection fraction and heart rate with associated point-wise linear 95% confidence intervals was estimated using the Kaplan–Meier estimator.
Results: During a time interval of 10 years, the median number of CMR's per patient was 3 (range 1-5). Indexed flows of ascending aorta, caval veins, pulmonary arteries decreased significantly across serial CMR's. Although a decrease of mean indexed aortic flow (3.03 ± 0.10 L/min/m^2 at first CMR versus 2.36 ± 0.14 L/min/m^2 at fourth CMR, p<0.001) was observed, ejection fraction did not decline (50 ± 1% at first CMR versus 54 ± 2% at fourth CMR, p=0.070). Indexed ventricular volume did not differ significantly across serial CMR's. The Kaplan Meier curve for estimated indexed aortic flow ≥ 2.5 L/min/m^2 after Fontan completion was: 98% at 5 years, 85% at 10 years, 62% at 15 years and 38% at 20 years.
Conclusions: The decrease of indexed aortic and cavo-pulmonary flows reflects the attrition of univentricular circulation and can be detected by means of serial CMR's. Ventricular systolic dysfunction does not contribute significantly to this attrition. In order to detect significant change of indexed aortic flow, we recommend performing serial CMR's as routine practice in the Fontan population.

Keywords: single ventricle physiology, cardiac magnetic resonance, fontan follow-up

graphical abstract



Adult Congenital Heart Disease

OP-040
Effect of exercise habits on fontan circulation

Shin Ono, Tomoaki Taruya, Fumiya Inoue, Daichi Hosokawa, Shinichiro Sakaki, Takeshi Ikegawa, Takuya Wakamiya, Sadamitsu Yanagi and Hideaki Ueda
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Background and Aim: Peripheral venous pressure increases during exercise in patients after Fontan surgery. Furthermore, there is a negative correlation between peak peripheral venous pressure:

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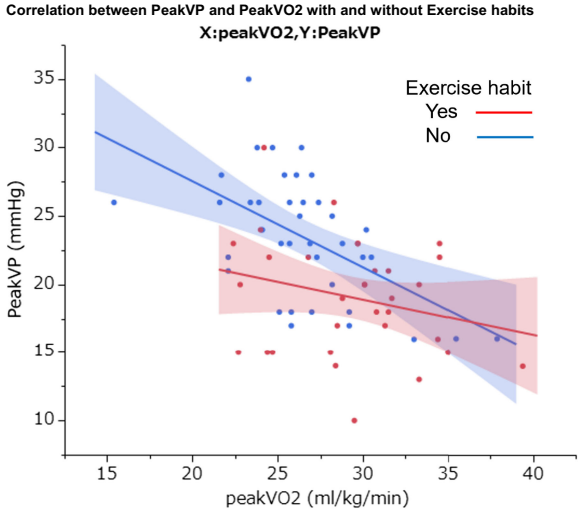
PPVP(mmHg) during exercise and PeakVO2 (ml/kg/min). We performed cardiopulmonary exercise stress tests in postoperative Fontan patients and compared the correlations between PPVP and peakVO2 between postoperative Fontan patients with and without exercise habits to investigate the effect of exercise habits on the Fontan circulation.
Method: In 70 postoperative Fontan patients (32 with exercise habits), PPVP and peakVO2 were measured by cardiopulmonary exercise testing using a Treadmill while peripheral venous pressure was measured. Multiple regression analysis with interaction was performed using PPVP as the objective variable and peakVO2 and exercise habit as the explanatory variables.
Results: Both PeakVO2 ($p=0.0005$) and exercise habit ($p=0.003$) independently contributed to peakVP. The estimated equations for those with and without exercise habits were below, indicating that PPVP was significantly lower in postoperative Fontan patients with exercise habits ($p < 0.0001$).
Ex habit (+): PeakVP = 33.4 - (0.44 × peakVO2) – 1.6 +

[0.18 × (peakVO2 - 27.8)]
Ex habit (-): PeakVP=33.8 - (0.44 × peakVO2) + 1.6 +

[- 0.18 × (peakVO2 - 27.8)].
Conclusions: Exercise habits may reduce elevated venous pressure during exercise in postoperative Fontan patients. This feature suggests that exercise habits may improve long-term prognosis in postoperative Fontan patients by reducing elevated venous pressure during daily activities and reducing organ congestion.

Keywords: Fontan, exercise, peripheral venous pressure



Exercise-induced elevation of venous pressure in postoperative Fontan patients is milder in those with exercise habits than without exercise habits

OP-041
Right ventricle unloading by transcatheter creation of bidirectional cavopulmonary connection in adults

Dunja Renner[1], Peter Ewert[1], Daniel Tanase[1], Kristina Borgmann[1], Jürgen Hörer[2], Andreas Eicken[1] and Stanimir Georgiev[1]
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Background and Aim: A bidirectional cavopulmonary connection is a well-established surgical procedure in young children with univentricular hearts (UVH). In adults, however, it is rarely performed and if so, then as an adjunct to other cardiac operations. Is there a role for the transcatheter creation of a bilateral PCPC (cathPCPC) in adults with low pulmonary vascular resistance, right ventricle volumeoverload but compromised pulmonary blood supply? We attempted to improve the haemoynamic situation of patients with medical limitation for surgery, due to several reasons, by excluding the upper body blood volume from the RA by connecting the SVC to the pulmonary arteries.
Method: Seven patients were selected for a cathPCPC: three had Ebstein's Anomalie or tricuspid valve dysplasia, three had UVH, and one right ventricular hypoplasia. Patients' ages were 18 to 65 years (median 53 years). Median mean pulmonary artery pressure was 15 mmHg (8 to 20mmHg). The PCPC was performed in two steps: First the implantation of a bare metal stent in the superior vena cava (SVC). Second, several weeks later, direct needle puncture from the SVC to the right pulmonary artery (RPA) and, consecutively, the implantation of covered stents between SVC and RPA.
Results: The procedure was technical successful in all patients. Mean follow-up period is 23 months (6 to 40 months). One patient died 2.5 months after the procedure as a result of his endstage cardiac disease, and one patient showed recurrent cardiac decompensation due to rhythm disturbances. Three patients reported an improvement of exercise tolerance. In one patient with zyanosis due to a LSVC without bridging vein the cathPCPC and the occlusion of collaterals led to a 10 % point increase of oxygen saturation; one zyanotic patient with untreated UVH improved 10 % in oxygen saturation. NT-proBNP levels improved in four of seven patients.
Conclusions: Percutanous PCPC in adults is feasible and shows an improvement of hemodynamics in some patients. A controlled study or registry with defined criteria for inclusion and evaluation should be the next steps to explore the potential of a cathPCPC as a palliative measure in adults with low pulmonary vascular resistance, right ventricle overload and compromised pulmonary blood flow.

Keywords: percutaneous bidirectional cavopulmonary anastomosis, reduced pulmonary blood flow, volume unloading of the right ventricle

OP-042
Digital twins for diagnosis and treatment planning in patients with univentricular palliation

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Background and Aim: Congenital heart defects often require surgical or interventional treatments. In univentricular defects, the goal is typically biventricular repair, but complex cases may necessitate univentricular palliation. This involves connecting the superior and inferior vena cava to the pulmonary artery through staged surgeries, known as total cavopulmonary connection (TCPC). Due to the heterogeneity in anatomy, hemodynamics, and treatment history, diagnosis and treatment decisions for these patients are challenging. This retrospective study investigates whether digital twins of univentricular patients can enhance anatomical and functional understanding, thus supporting personalized diagnosis and treatment planning.
Method: Anatomical digital twins were reconstructed from MRI, CT, and angiography data. Computational fluid dynamics were utilized to calculate patient-specific blood flow within the

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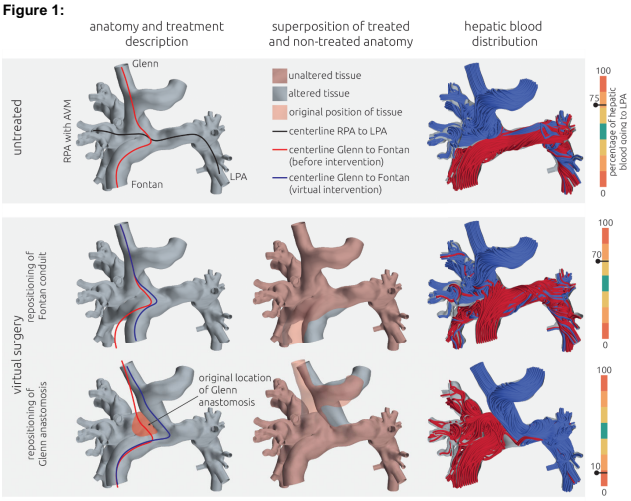
TCPC, enhancing the digital twins by hemodynamic information, such as pressures and velocities. By virtually altering the anatomical digital twins, different treatment scenarios, such as the implantation of covered stents or vascular grafts, could be mimicked. Additionally, the hemodynamic outcomes of different strategies were assessed and compared against each other.
Results: Digital twins were created for 10 univentricular patients, addressing specific aspects such as optimizing hepatic blood flow distribution and reducing energy loss in Fontan stenosis. Various treatment strategies, derived from heart team recommendations or individual expert opinions, were virtually tested, yielding at least one viable treatment option per patient. The detailed insight into patient-specific anatomy and pre-interventional hemodynamics was deemed valuable for treatment planning.
Conclusions: Digital twins hold significant potential in addressing the varied challenges in univentricular palliation patients. They offer a pathway to personalized treatment planning and risk stratification by simulating the outcomes of different interventions. However, further standardization of methodologies is necessary for prospective clinical application and broader adoption.

Keywords: Fontan, digital twins, univentricular physiology, modelling and simulation, therapy planning, digital medicine

Figure 1:



Exemplary illustration of a digital twin of a Fontan-palliated patient with uneven hepatic blood flow distribution towards the left and right pulmonary artery, resulting in pulmonary arteriovenous malformations in the right lung (top row). The lower rows show two virtual surgeries, which failed to achieve even distribution of hepatic blood.

OP-043
Fontan circulatory failure: Relevance of incorporating cpetecho in a dedicated fontan clinic

Aleksandra Cieplucha[1], Youri Bekhuis[2], Thomas Rosseel[2], Joeri Van Puyvelde[2], Els Troost[3], Pieter De Meester[3], Werner Budts[3] and Alexander Van De Bruaene[3]
[1]Poznan University of Medical Sciences, Poznan, Poland & KU Leuven, Department of Cardiovascular Sciences, Leuven, Belgium; [2]University Hospitals UZ Leuven, Department of Cardiology, Leuven, Belgium; [3]KU Leuven, Department of Cardiovascular Sciences, Leuven, Belgium & University Hospitals UZ Leuven, Department of Congenital and Structural Cardiology, Leuven, Belgium

Background and Aim: Fontan circulatory failure is a syndrome with a heterogeneous clinical presentation. Exercise echocardiography with simultaneous peripheral venous pressure measurement (CPETecho-PVP) may provide superior insights in the pathophysiology of Fontan circulatory failure. This study sought to assess the hemodynamic correlates of peak oxygen consumption (percent predicted peakVO2) using CPETecho-PVP in Fontan patients.
Method: Forty-one consecutive Fontan patients (LV dominance in 32) were evaluated with echocardiography, cardiopulmonary exercise testing and simultaneous peripheral venous pressure measurements. A multipoint PVP/CO slope was calculated as a linear approximation using linear regression analysis from pressure-flow plots for each patient. Spirometry and blood work, including N-terminal prohormone of brain natriuretic peptide (NTproBNP), was performed.
Results: Compared to Fontan patients with a peakVO2 >50% predicted (n=25; 61%), those with a peakVO2 <50% predicted (n=16; 39%) were more likely to have protein losing enteropathy (p=0.045), had lower forced vital capacity (66±13 vs 76±16 % pred; p=0.049), a higher NTproBNP (376±295 vs 133±116 ng/L; p=0.048), and lower albumin level (38±10 vs 47±4 g/L; p=0.001). On CPETecho-PVP, they had a lower heart rate reserve (+76±35 vs +100±37 bpm; p=0.041), a lower increase in cardiac index (2.0±1.2 vs 2.9±1.0 L/min.m^2; p=0.014) and a steeper PVP/CO slope (3.1±1.7 vs 1.8±1.1 mmHg/l.min; p=0.006). Age (31±9 vs 29±10 years; p=0.498), saturation at rest (95±4 vs 96±3%; p=0.387) and peak exercise (90±6 vs 90±5%; p=0.664) and PVP at rest (18±5 vs 17±4 mmHg; p=0.780) and peak exercise (29±5 vs 27±6 mmHg; p=0.318) were similar between groups. Of interest, PVP tended to increase more with leg raise (6±5 vs 3±1 mmHg; p=0.052) in those with peakVO2 <50% predicted.
Conclusions: Although exercise intolerance appears multifactorial, a steeper PVP/CO plot is associated with decreased exercise capacity whereas absolute PVP values are not. This suggests the value of implementing CPETecho-PVP in the setting of a dedicated Fontan clinic.

Keywords: Fontan circulatory failure, exercise intolerance, CPET-echo, peripheral venous pressure, non-invasive exercise assessment, peak oxygen consumption

CPETecho-PVP

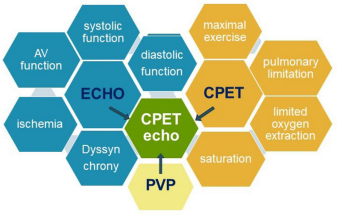


Fig. 1. CPETecho-PVP is a multifactorial non-invasive assessment of exercise intolerance in Fontan patients

KU LEUVEN

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OP-044

Microfon study – differences in the microcirculation in fontan adults compared to healthy controls; - preliminary results

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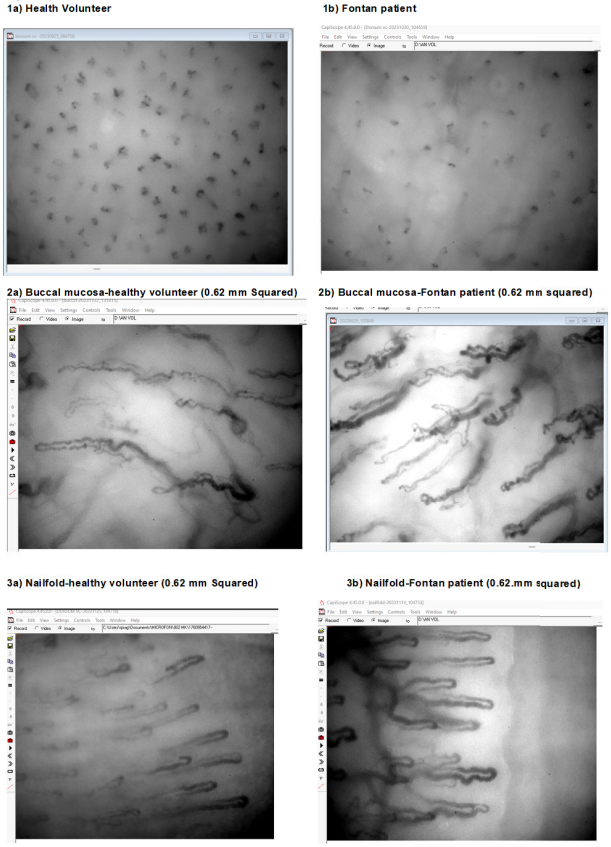
Background and Aim: Fontan palliation for functionally univentricular CHD carries long-term multiple organ sequelae and uncertain longevity in adulthood. The mechanisms behind these complications are incompletely understood but may not solely relate to systemic venous hypertension. We hypothesized that differences in the capillary microcirculation may play a role.
Method: We have so far evaluated the microcirculation in 10 adults (≥16 years) who underwent early childhood Fontan palliation and compared them with 11 healthy controls (≥16 years). Handheld Capillary Microscopy (HVCS, Exeter, UK) was used to assess the microcirculation in the hands, feet, nailfolds, and oral cavity. Basal and Maximal Capillary Density (BCD & MCD) were recorded & measured on the dorsum of the hand and feet after venous congestion according to a well-established protocol. Sitting and standing blood pressures were recorded as per British Hypertension Society guidelines. Central blood pressure, augmentation index, and pulse wave velocity were measured using Mobil-O-Graph® device (Stolberg, Germany). The independent 't' test was used for comparison of means among the groups using IBM-SPSS v26.
Results: The Fontan adults had statistically significantly lower basal (BCD-mean difference -18 capillaries/field, p = 0.001) and maximal capillary density (MCD-mean difference -23 capillaries/field, p = 0.001) on dorsum of the finger and of the feet (BCD - mean difference - 6 capillaries/field, p = 0.087; MCD-mean difference - 12 capillaries/field, p = 0.012). Other findings include thickening of the capillaries, increased looping, distortion, and widening of venous limb of the capillary loop in the upper and lower extremities and buccal mucosa. Our Fontan patients also had significantly lower systolic BP (mean difference -12 mm Hg, p < 0.011), diastolic BP (mean difference -8 mm Hg, p < 0.001), and pulse wave velocity (mean difference -.73 milliseconds, p <0.016) compared to the healthy controls. There was no statistically significant difference in central blood pressure between both groups.
Conclusions: Our Fontan adults have significant structural capillary rarefaction and likely adverse remodeling of the microcirculation in their extremities compared to the healthy controls. Studies to examine this in other repaired CHD phenotypes are ongoing,

Keywords: Fontan Procedure, Microcirculation, Capillary Rarefaction, Central Blood Pressure

MICROFON Study Image

Figure 1-Maximal capillary density after venous congestion in dorsum of the middle finger in hand (0.62 mm area squared)



The maximal capillary density in the dorsum of the finger in the hand, buccal mucosa capillaries, and nailfold capillaries in the Fontan adults and healthy volunteers

OP-045

Pregnancy outcomes in adult congenital heart disease women: a single centre experience

Rosalinda Palmieri[1], Emanuela Concetta D'angelo[1], Veronica Lisignoli[1], Giulia Di Già[1], Marco Della Porta[1], Rossella Iuzzolino[1] and Gianfranco Butera[2]
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Background and Aim: Pregnancy is associated with significant hemodynamic changes in ACHD women, increasing the risk of cardiac, obstetric and fetal complications. The aim of this study is an overview of a tertiary center experience.
Method: We collected data of ACHD pregnant women evaluated at "ACHD program" between January 2018 and October 2023. Cardiological risk of pregnancy was estimated according to WHO classification. Primary outcomes included maternal cardiological complications.
Results: We collected data of 73 consecutive patients and 78 pregnancies (5 women had 2 pregnancies). Mean age at pregnancy was 33 years old (+/-2.6 years). Eight patients had an history of previous spontaneous miscarriages. Sixty-four patients had a left systemic ventricle physiology and the most frequent primary diagnosis was Tetralogy of Fallot in 24 patients (33%). Five patient had systemic right ventricle physiology and 4 patient Fontan circulation. According to WHO classes: WHO I: 11 patients (15 %); WHO II: 31 patients (42 %); WHO II-III: 15 patients (21 %); WHO III: 16 patients (22%). None patient was in class WHO IV. Ten pregnancy are still ongoing, and 2 miscarriages occurred in first trimester. Mean gestational week at delivery was 38 weeks (SD +/- 1.9 weeks). Fourty-one patients (62%) underwent to Cesarean Section (CS). Three patients underwent to urgent CS for fetal distress. Preterm births (birth < 37 weeks) were 5 while

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4 low birth weights (birth weight <2500g) occurred. One Fontan patients had a twins-birth. During pregnancy and puerperium rate of cardiovascular events was 10%. During pregnancy functional class worsening occurred in 2 patients: in one persisted after delivery due to incremental gradient through aortic valve which led to surgery in follow-up. Arrhythmias occurred in 3 patients during pregnancy and in one presented after delivery. All arrhythmias were supraventricular arrhythmias. During pregnancy 1 patient developed hypertensive disorder. During the puerperium one Fontan patient developed deep venous thrombosis and pulmonary embolism. All complications occurred in III WHO class patients. No maternal or neonatal mortality occurred.

Conclusions: In the current era, pregnancy can be carried out successfully also in selected ACHD women. Correct counseling and strict follow-up during and after pregnancy in a tertiary center is recommended.

Keywords: ACHD, Adult Congenital Heart disease, Pregnancy

OP-046

exercise capacity is preserved, despite severe right ventricle dilation after repair of tetralogy of fallot

Benedetta Leonardi[1], Lucilla Ravà[1], Federica Gentili[1], Gianfranco Butera[1], Adriano Carotti[1], Gabriele Rinelli[1], Giulio Calcagni[1], Giovanni Antonelli[1], Michele Lioncino[1], Fabrizio Drago[1], Ugo Giordano[1], Giulia Cafiero[1], Fabrizio Drago[1] and Rachel Wald[2]

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Background and Aim: Pulmonary regurgitation (PR), can lead to right ventricular (RV) volume overload and cardiac adverse events in patients with repaired Tetralogy of Fallot (rTOF). Pulmonary valve replacement (PVR) can resolve the incompetence of valve, but its results on the outcome are still unclear. To this day, PVR indications are based on cardiac resonance imaging (CMR) thresholds of RV/left ventricle (LV), also in asymptomatic patients. Cardiopulmonary test (CPET) has been shown to evaluated the functional capacity in this population. Therefore, we sought if values above or below of right end-diastolic volume indexed for BSA (RVEDVi) of 160 ml/m2 and/or RV EF ≤47% and LV EF ≤50%, used for PVR indication, relate to exercise capacity in rTOF patients.

Method: All rTOF with moderate or greater pulmonary regurgitation (PR) enrolled in the CORRELATE registry (following in Rome or Toronto) were enrolled if they had contemporary CMR and CPET studies. The analysis was performed on three subgroups on the basis of CMR parameters [1) RVEDVi <160 ml/m2 vs RVEDVi ≥160 ml/m2 2) RVEDVi <160 ml/m2 + RVEF≥47% vs RVEDVi ≥160 ml/m2 + RV EF <47% 3) RVEF <47% + LVEF <50% vs RVEF≥47% + LVEF≥50%)]. The following CPET data were collected: peak VO2 absolute, percent-predicted (pred) peak VO2, peak O2 pulse absolute, peak O2 pulse percent pred, ventilatory efficiency (VE/VCO2) anaerobic threshold, and VE/VCO2 slope.

Results: In total, 587 tests were completed in 342 patients (55% males; mean age at CPET 31.9 ±12.8 years). The multivariate analysis adjusted for age at CPET, sex and BMI revealed no significantly statistical relationship between CMR thresholds and CPET parameters (all p>0.05). CPET data subdivided on the basis of subgroups mentioned are shown in Table 1.

Conclusions: Exercise capacity is maintained with RVEDVi above compared with below used cut-off for PVR referral

Keywords: Cardiopulmonary exercise stress test, Tetralogy of Fallot, peak VO2 absolute

Table 1

	RVEDVi			*RVEDVi+ RV EF*				*RV EF + LV EF*		
	RVEDVi <160 ml/m2	RVEDVi ≥160 ml/m2	P	RVEDVi <160 ml/m2 + RV EF>=47	RVEDVi ≥16 0 ml/m2 + RV EF <47	P	RVEDVi <160 ml/m2 + RV EF<47	P	RV EF<47% + LV EF < 50%	RV EF≥47% + LV EF≥50%	P
Peak VO2 [mL/min/kg]	22.5 (16-29.4)	23.8 (19.6 –28.9)	0.650	25.9 (20.3-33.8)	23.0 (19.6-27.2)	0.491	26.0 (19.1-31.0)	0.864	22.0 (18.0-27.0)	25.8 (19.0-31.1)	0.115
Peak VO2 percent max pred [%]	68.5 (61-77)	69 (58-128)	0.814	68.0 (63.0-69.2)	69.0 (57.0-69.0)	0.812	70.0 (61.0-81.8)	0.665	65.5 (56.0-79.0)	70.0 (61.0-81.7)	0.307
Peak O2 pulse absolute [mL/beat]	8.6 (6.2-10.1)	10.7 (8.6-13.1)	0.051	9.6 (8.6-13.0)	10.8 (8.8-12.9)	0.311	10.6 (8.5-13.6)	0.391	12.1 (9.6-14.4)	10.4 (8.6-13.6)	0.044
Peak O2 pulse percent pred [%]	83.5 (74.0-89.0)	84 (72-95.5)	0.723	86.0 (82.0-89.0)	82.0 (68.5-94.0)	0.418	87.0 (76.5-99.0)	1.000	85.0 (71.0-93.0	87.0 (76.0-99.0)	0.647
VE/VCO2 anaerobic threshold	36.7 (31.1-39)	33 (30-37)	0.256	32.0 (26.3-36.0)	33.0 (30.0-38.0)	0.802	33.0 (29.0-36.0)	0.806	32.0 (30.0-37.0)	33.0 (29.0-36.0)	0.497
VE/VCO2 slope	29.6 (24.9-35.4)	26.5 (23.4-31)	0.357	29.6 24.9-35.4)	30.0 (26.5-32.0)	0.792	23.6 (20.8-27.8)	0.247	32 (28.25-40)	23.6 (20.8-27.1)	0.086

Median values were compared through clustered quantile regression

CPET data subdivided on the basis of three subgroups: 1) RVEDVi <160 ml/m2 vs RVEDVi ≥160 ml/m2 2) RVEDVi <160 ml/m2 + RVEF≥47% vs RVEDVi ≥160 ml/m2 + RV EF <47% 3) RVEF <47% + LVEF <50% vs RVEF≥47% + LVEF≥50%

Fetal Cardiology

OP-048

Improving fetal heart care in under-resourced areas: task-switching, telemedicine, and collaboration

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Background and Aim: Prenatal diagnosis of congenital heart disease allows for delivery planning and optimization of newborn outcomes. Kentucky is largely rural and under-resourced, with the fourth highest poverty rate in the US, and 20% of children living below the poverty line. In 2011, because the prenatal diagnosis rate around Louisville, Kentucky was 28%, a regional fetal tele-echo program was started. Local sonographers with primary roles in adult cardiology or obstetrics received focused fetal echo training. Fetal echocardiograms were performed in local hospitals and pediatric cardiologists in Louisville provided results and delivery recommendations via telemedicine. The aim of this study was to evaluate the effectiveness and potential healthcare cost-savings of this novel program.

Method: The study population included all expecting mothers who had a fetal tele-echo performed between February, 2011 and June, 2018 at 3 outreach sites in Kentucky (Owensboro, Paducah, and Lexington). Retrospective chart review was performed on maternal and newborn records at 4 hospitals; the tertiary hospital and 3 outreach hospitals. Fetal tele-echo results and recommendations were compared to maternal records, newborn echocardiogram results, and newborn clinical course. Potential transport costs were calculated using the US government reimbursement formula.

Results: A total of 410 expecting mothers had a fetal tele-echo during the study period, with an average of 1.04 fetal echocardiograms per patient. 118 mothers (29%) did not deliver at any study hospital, none of those infants received cardiac care. 292 mothers (71%) delivered at a study hospital with median follow-up of 4 days. Delivery was recommended at a local hospital for 359 normal fetal tele-echos (87%), and for 21 minor abnormalities (5%), with potential cost-savings of $124,752 by avoiding unnecessary transport. Delivery was recommended at the tertiary hospital for 18 moderate/severe cardiac abnormalities (4.4%), with potential cost-savings of $89,351.85 by avoiding emergent transport.

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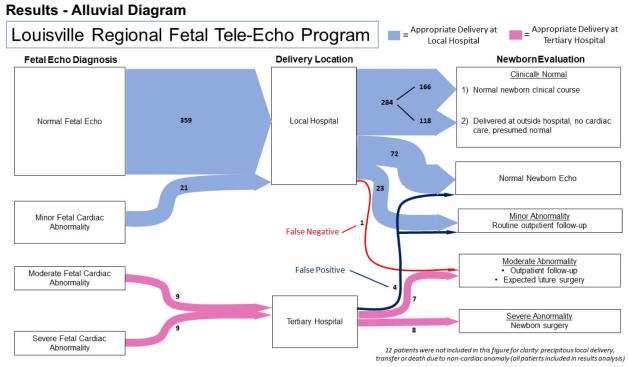
Comparing fetal tele-echo findings to newborn findings, the false negative rate was 0.34%, and the false positive rate was 1.4%.
Conclusions: A regional fetal tele-echo program using focused specialty training for local sonographers and collaboration with regional hospitals can accurately and efficiently identify congenital heart defects during pregnancy, inform decisions about appropriate delivery location, and provide substantial potential healthcare cost-savings.

Keywords: fetal echocardiography, telemedicine, prenatal diagnosis, congenital heart disease, healthcare costs, resource-limited settings



Alluvial diagram providing visualization and explanation of study results

OP-049

The role of the non-invasive abdominal fetal ECG in the detection and monitoring of fetal tachycardia

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Background and Aim: Fetal tachycardias have the potential to cause adverse fetal outcome including ventricular dysfunction, hydrops and fetal demise. In postnatal life the ECG is the gold standard, but in fetal practice, echocardiography is the most frequently used technique to diagnose and monitor fetal arrhythmia. Non-invasive extraction of the fetal ECG (fECG) has the potential to provide additional information on tachycardia mechanism and to monitor intermittent arrhythmias. New signal processing techniques have been developed to provide improved data quality and clinical translation. The aim of this study is to assess the fetus with known or suspected tachycardia using non-invasive abdominal fetal ECG and correlate results with fetal echocardiography findings.
Method: Prospective recruitment of pregnant participants where the fetus had known or suspected tachycardia in a tertiary fetal cardiology unit. Overnight fECG recording at home using the portable Monica AN24 monitor was performed. Data processing using bespoke Matlab scripts to provide information on fetal heart rate (FHR) and cardiac rhythm. Fetal ECG data compared with clinical data obtained using echocardiography and postnatal findings.
Results: 12 participants were recruited and undertook between 1-4 fECG recordings during the pregnancy. A total of 20 fECG recordings were obtained. Gestational age was 28 (23.5-32.9) weeks [median(IQR)].Duration of recording was 512(415-607) minutes [median(IQR)]. 12/12(100%) recordings were successful, and 20/20(100%) recordings had FHR graphs extracted. Intermittent tachycardia was demonstrated on FHR graphs. Rhythm strips correctly identified short-VA tachycardia, long-VA tachycardia, atrial flutter and atrial ectopic beats with findings correlating with echocardiography. Postnatal recurrence in one case (long-VA tachycardia) was consistent with the prenatal fECG findings.
Conclusions: Our initial results show that rhythm strips of fECG signal can be extracted and correctly identify the electrical mechanism of arrhythmia in cases of fetal tachycardia. The potential to monitor FHR over a prolonged period is an additional advantage over current monitoring strategies which facilitates recording of intermittent arrhythmias and gauging the response to medical therapy.

Keywords: fetal, tachycardia, ECG

Figure

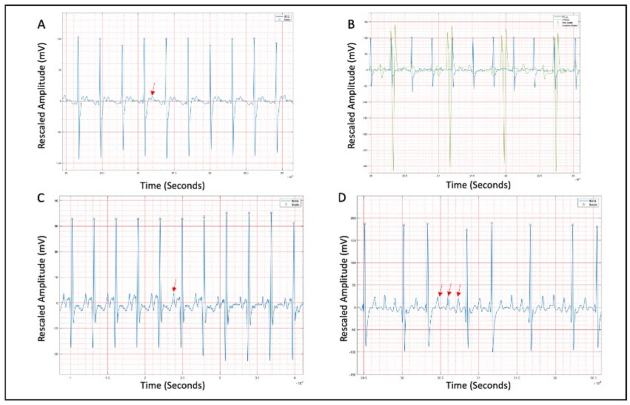

Figure 1: A&B short-RP fetal tachycardia shown as A the fetal ECG trace (blue) with fetal P waves (red arrow) and B the fetal ECG trace (blue) with the maternal ECG trace (green). C long-RP fetal tachycardia with fetal P waves (red arrow). D Atrial flutter with variable conduction with atrial rate of 360 beats per minute. fetal P waves (red arrow)

Rhythm strip of cases with fetal tachycardia (see figure legend)

OP-050

Survival and long-term outcomes of antenatally diagnosed transposition of the great arteries

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Background and Aim: Transposition of the great arteries (TGA) is commonly diagnosed prenatally. We sought to describe survival outcomes and morbidity for prenatally-diagnosed patients with TGA to inform prenatal counselling for expectant patients.
Method: Fetuses with TGA or TGA with ventricular-septal-defect (VSD) prenatally diagnosed between 1995-2022 at Evelina London Children's Hospital (United Kingdom) were identified and electronic records retrospectively reviewed. Those with

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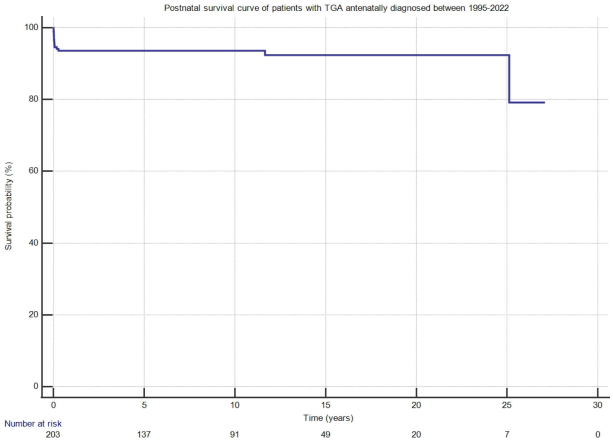
complex TGA, including co-existing coarctation and pulmonary stenosis, were excluded.
Results: There were 214 fetuses diagnosed resulting in 203 live-births, 1 intrauterine-demise and 10 underwent termination of pregnancy. 137/203 (67.5%) were simple TGA and 66/203 (32.5%) additionally had a VSD. Median birthweight 3.2kg (IQR 2.95-3.5kg), median birth gestation 38-weeks (IQR 38-39-weeks). 57/203 (28%) were female. Pre-operative balloon-atrial-septostomy was performed in 130/203 (64%) with 65 (32%) in the first 24 hours. Arterial-switch-operation (ASO) was performed in 201/203 (99%) with 47/201 (23.4%) having concomitant VSD closure. The 2 patients not undergoing ASO were born prematurely and died prior to surgery. Survival of live births was 99% at 30-days, 93.6% at 1-year/5-years/10-years, 92.6% at 20-years and 79.2% at 25 years of age (figure-1). 5-year mortality, significantly reduced from the era of 1995-2005 (6/41, 14.6%) compared to 2006 to 2015 (3/87, 3.4%) and 2016-2022 (4/75, 5.3%), p=0.02. There was no significant difference in mortality between latter two eras (p=0.7). Reintervention following ASO (including in early postoperative period) occurred in 17/201 (8.5%): coronaries (n=6), aortic valve (n=2), main pulmonary/branch pulmonary artery stenosis (n=7), VSD reintervention (n=1), diaphragm plication (n=1). Of the 201 having ASO, 182 (90.5%) were followed in our institution for a median 7.8-years (IQR 3.9-13.8-years). Recorded morbidity included: mild myocardial dysfunction in 3/182 (1.6%), pulmonary hypertension 1/182 (0.6%), arrhythmia 3/182 (1.6%), neurological disorders 19/182 (10%), autism 11/182 (6%) and attention deficit hyperactivity disorder 2/182 (1%).
Conclusions: Survival for prenatally-diagnosed patients TGA has improved in recent eras. Overall, most survive into the 3rd decade of life following prenatal diagnosis. Reintervention was needed in 8.5% by 25 years. Neurological and behavioural morbidity is reported in up to 10%. This data will be helpful when counselling parents faced with a diagnosis of TGA in pregnancy, informing decision making and expectations.

Keywords: Transposition of the great arteries, survival, fetal cardiology, outcomes

Figure-1. Postnatal survival curve of patients with TGA antenatally diagnosed between 1995-2022



Postnatal survival curve of patients with TGA antenatally diagnosed between 1995-2022

OP-051

Fetal right to left ventricular discrepancy as a marker of extracardiac anomalies with and without coarctation

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Background and Aim: Right heart enlargement suspecting coarctation of the aorta (CoA) is a frequent referral reason but appears to be an unspecific sign of a possible other anomaly in numerous cases. The objective was to illustrate the incidence and the spectrum of other possible pathologies from unconfirmed cases.
Method: For this retrospective observational study, we assessed all cases with right to left heart discrepancy referred to our institution from 2010-2023, where CoA was suspected or could not be dismissed with the high false negative diagnosis rate in mind. Cases with complex congenital heart disease (CHD) were excluded except for those with atrioventricular septal defect and double outlet right ventricle with normal relation of the great arteries both resulting in biventricular physiology. Prenatal cases were compared to neonatal outcome. Common institutional policy is to follow all cases until 6 months after birth.
Results: From 140 cases with a suspicion of CoA, 6 pregnancies were terminated (TOP), 6 were lost in utero (IUFD) or were supported palliatively after birth, both for other reasons than the suspected CoA; and 2 were lost to follow-up. Of 126 livebirths, n=50 had CoA surgery (40%), whereas in n=76 (60%) no CoA was diagnosed. 20/50 (40%) of the confirmed CoA cases had a genetic defect, syndrome or extracardiac anomaly. Of the patients without CoA 31/76 (41%) were found to have other pathological findings; 7 had a genetic defect, 8 had a genetic defect with at least one minor congenital heart abnormality, 9 had isolated minor congenital heart defects other than CoA and 7 had extracardiac anomalies of other organs without a specific genetic defect. Intrauterine growth retardation at birth was found in 13/50 (26%) cases with coarctation and in 14/76 (18%) cases without CoA.
Conclusions: Fetal right heart enlargement is associated with high percentage of genetic defects, extracardiac anomalies and minor cardiac anomalies even in cases without CoA. In 40% of cases without CoA an extracardiac, genetic or minor cardiac abnormality other than CoA was found in our population.

Keywords: Coarctation, Fetal right to left discrepancy of the ventricles

OP-052

Multidisciplinary approach to vascular rings with prenatal diagnosis: symptoms are never enough

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Background and Aim: Vascular rings are a rare group of congenital abnormalities that can cause tracheal or esophageal compression. Their approach is under *Discussion:* there is no clear correlation

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between clinical and radiological findings. The aim of our study is to describe prenatal and postnatal findings, management and outcome of vascular rings in two tertiary centers.
Method: Retrospective, descriptive study including all cases diagnosed with vascular ring (double aortic arch (DAA) or right aortic arch (RAA) with left ductus arteriosus and/or aberrant left subclavian artery (ALSA), both without associated cardiac abnormalities) in two fetal cardiac units from 2006 to 2023. Clinical records were reviewed for prenatal/postnatal sonographic features, genetic studies, symptoms, imaging studies performed, surgery and follow-up.
Results: 87 fetuses with vascular rings were identified (median gestational age 23+2). 78 cases were RAA (53 with ALSA), 8 were DAA and 1 had left aortic arch with right ductus and ARSA. Genetic screening was performed on 57% of patients (4 cases of 22q11 syndrome). In 11 cases postnatal follow-up was not possible. Only 21% of patients (17) had symptoms before 6 months of age, but during the follow-up (median of 6.8 years), up to 57% (45) of them reported respiratory and/or digestive mild symptoms. AngioTAC was performed in 57 patients (73%), being normal in only 15.7%. Tracheal compression <50% was identified in 38/57 patients and 10/57 had >50% of compression. Regarding fibrobronchoscopy, it was performed in 23 patients (29.4%), finding compression in most of the cases: <50% of compression in 12/23 cases and >50% of compression in 7/23 cases. In both tests there was no correlation between symptoms and severity of tracheal compression. Surgery was performed in 29 patients (33%) with symptoms and/or tracheal compression superior to 40%, at a median age of 4 years. In all cases except from 1, symptoms were resolved after the procedure.
Conclusions: Multidisciplinary approach of vascular rings is mandatory. Beyond symptoms and clinical follow-up, imaging and/or respiratory studies must be performed the sooner the better in order to identify those cases with compression that can benefit from surgery.

Keywords: Vascular rings, angioTAC, congenital heart disease, prenatal diagnosis

OP-053
Fetal brain oxygenation remains constant in maternal hyperoxygenation despite marked placental response at the expense of blood supply to the brain

Johannes K. Steinweg[1], Alena Uus[2], Milou Van Poppel[1], Reza Razavi[1], Mary Rutherford[2], David Lloyd[1], Kuberan Pushparajah[1] and Jana Hutter[2]
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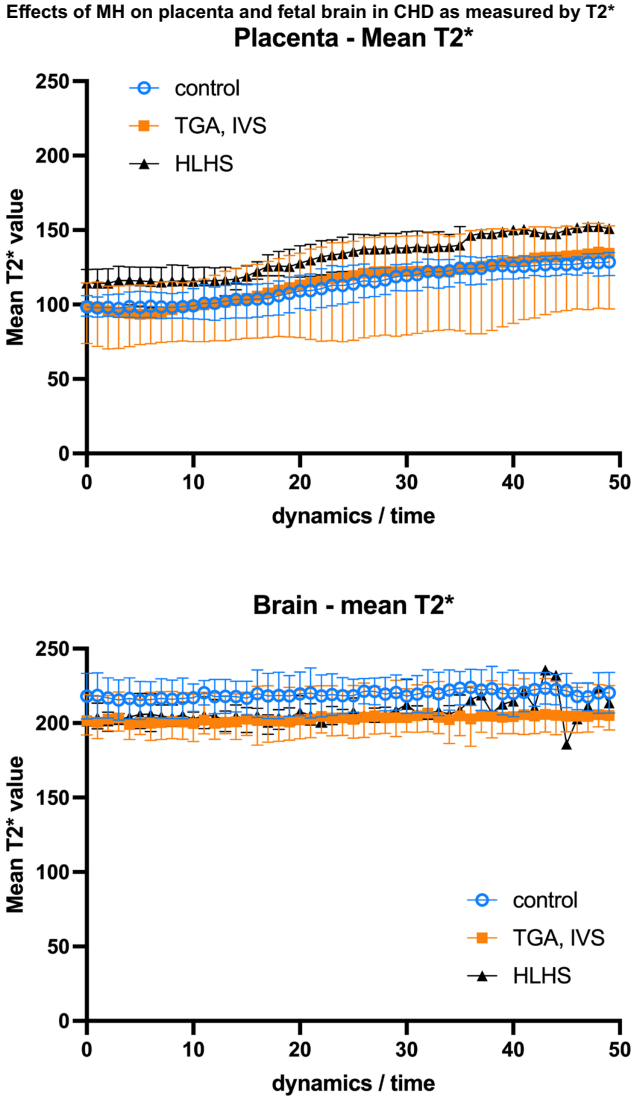
Background and Aim: Maternal hyperoxygenation (MH) has been used temporary as a diagnostic marker, but also as therapeutic option, following the theory that persistent pulmonary vasodilation leads to increased left heart throughput, thereby stimulating growth. Impaired fetal brain growth was associated with reduced oxygen delivery to the fetal brain in CHD, but reduced cerebral blood flow and reduced head growth has been shown in previous studies. We hypothesised that MH changes placental and brain oxygenation and fetal cerebral blood flow in CHD and controls.
Method: In this prospective cross-sectional interventional study, pregnant women carrying a fetus diagnosed with congenital heart disease (CHD) alongside a control group of women with uncomplicated pregnancies were scanned using a 1.5 Tesla Philips Ingenia scanner at similar gestational age with a free breathing multi-echo gradient-echo (MEGE) echo planar imaging pulse sequence (Resolution 1.5x1.5x2.5mm, typical FOV 256x256x40, TE= [7.3,57.8,108.3,154.6ms], TR=10–12s, up to 50 timepoints, resulting scan time 8–10min.) MH was started 2min into the scan and T2* maps were calculated for placenta and brain to estimate changes in oxygenation. SVC flow was measured with phase contrast sequences using metric optimized gating. Cases with contractions during the scan were excluded from further analysis.
Results: From 21 scanned cases 10 were excluded for contractions. 3 controls, 5 TGA (IVS) and 3 HLHS allowed mixed model analysis for repeated measurements. All showed a marked increase of placental mean T2* values with an increase by 21% in HLHS, 30% in control and 39 % in TGA ($p<0.001$). Brain mean T2* value increase with MH was small (1.2–1.9%) and only significant in TGA ($p<0.001$), while changes in control and HLHS did not reach statistical significance ($p=0.108$ and 0.269, respectively). Blood flow to the fetal brain decreased in almost all cases during MH.
Conclusions: While placental oxygenation increases markedly with MH, effects on brain oxygenation are at least 10 times smaller in



Placenta and fetal brain mean T2 values during continuous scan of 8-10 minutes. Oxygen supply via face mask started at timepoint 10. Values depicted with mean & error per colour-coded diagnosis groups control, TGA (IVS), HLHS. Mean T2* values over the entire organ per timepoint.*

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uncomplicated pregnancies and those with fetal CHD. Furthermore, reduced SVC flow during MH suggests intact autoregulatory processes responding to O2 delivery even with antenatal CHD and may impair nutrient supply. MH effects the complex interplay of placenta, fetal heart and brain beyond reduced PVR.

Keywords: Maternal hyperoxygenation, Fetal MRI, Brain, Placenta, Blood Flow

General Cardiology

OP-054
Effects of anticoagulant treatment options on embryopathy in pregnant women

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Background and Aim: Pregnancy is associated with a hypercoagulable state, which makes pregnant women with prosthetic heart valves at high risk of thromboembolism. However, no consensus on the present anticoagulation treatment for those women has been reached yet, in pregnancy. Oral anticoagulants (warfarin), standard heparin and low molecular weight heparins (LMWH) are the anticoagulant agents we have available. Warfarin appears to offer the best protection against thromboembolic complications, but the risk of its teratogenic effect is the concern of the clinicians. The use of heparin derivatives is also controversial. Although it has better fetal outcomes, it may be insufficient to protect the mother with mechanical heart valves from thromboembolic complications. Large controlled studies cannot be performed in this patient group for ethical reasons.
In this study; Children of patients with mechanical heart valves who were followed up by the cardiology department of Kosuyolu Yuksek Ihtisas Training and Research Hospital and who received anticoagulant treatment during pregnancy were evaluated. Our aim is to investigate the long-term effects of fetal anticoagulant treatment exposure.
Method: Detailed physical examinations of the children were performed; hearing and vision test, developmental screening with Denver II, cardiac evaluation with echocardiography and electrocardiography, whole abdominal ultrasonography and thyroid ultrasonography were performed. The results obtained were compared with the anticoagulant treatment used during pregnancy.
Results: In our study, 32 live-born babies of 30 mothers who had mechanical prosthetic valves and received anticoagulant treatment throughout pregnancy were evaluated in the late period. The patients included in the study were divided into 3 groups: those exposed to enoxaparin during the fetal period (n:12), those exposed to 2.5 mg/day warafarin together with enoxaparin (n:8), and those exposed to 4 mg warfarin together with enoxaparin (n12). It has been observed that there is no difference in terms of teratogenicity in the long term when using LMWH, LMWH + 2.5 mg/day warfarin and LMWH +4 mg/day warfarin during the fetal period.
Conclusions: Our retrospective results indicate that low-dose warfarin use during pregnancy is safe in terms of long-term teratogenicity, but prospective randomized controlled studies are needed to support this.

Keywords: warfarin, embryopathy

Genetics, Basic Science and Myocardial Disease

OP-055
Pioglitazone and its R-enantiomer prevent disease in a hypertrophic cardiomyopathy mouse model

Anna Theresa Pfaller[1], Sarala Raj Murthi[2], Andreas Petry[3], Jan B. Stöckl[4], Bachuki Shashikadze[4], Florian Flenkenthaler[4], Manuel Schmid[5], Gianluca Santamaria[6], Joachim P. Schmitt[7], Josh Gorham[8], Christopher N. Toepfer[5], Nadine Spielmann[9], Agnes Görlach[10], Alessandra Moretti[11], Peter Ewert[12], Thomas Fröhlich[4], Karin Klingel[13], Jonathan G. Seidman[8], Christine E. Seidman[8] and Cordula M. Wolf[12]
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Background and Aim: Hypertrophic cardiomyopathy (HCM) is a genetic disorder marked by progressive myocardial hypertrophy, fibrosis, ventricular dysfunction, arrhythmias, and an increased risk of sudden cardiac death, particularly in the young. It primarily

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results from autosomal-dominant mutations in sarcomere protein-encoding genes, leading to altered biophysical properties, increased energy demand, and activation of pro-hypertrophic and pro-fibrotic signaling pathways. Research underscores disruptions in metabolic signaling, mitochondrial dysfunction, inflammation, and oxidative stress as secondary perturbations exacerbating HCM progression. This highlights potential drug targets to improve metabolic function and reduce mitochondrial damage in HCM. Pioglitazone (pio), a PPARY agonist and anti-diabetic agent, emerges as a promising candidate, recognized for ameliorating the aforementioned disruptions in various diseases. Pio comprises S- and R-enantiomer (R-pio), with R-pio lacking PPARY activity and having fewer side effects. To date, no studies have investigated pio, especially R-pio, in HCM. Thus, this study examines the potential of the thiazolidinediones pio and R-pio as disease modifiers in HCM, utilizing their anti-inflammatory properties and their capacity to enhance mitochondrial function and metabolic homeostasis.

Method: Drug effect was studied in the established α-MHC719/+ HCM mouse model, which mimics the characteristic features of human HCM. Disease phenotype, assessed by transthoracic echocardiography for myocardial hypertrophy and histopathology for myocardial fibrosis, was compared between vehicle-treated α-MHC719/+ (α-MHC719/+,n=21) and wildtype (WT,n=22) mice, as well as pio- (n=7) and R-pio-treated (n=9) α-MHC719/+ mice. Ex vivo analyses included transcriptomics, proteomics, and pathway exploration.

Results: Compared to WT, there was marked left ventricular hypertrophy and myocardial fibrosis in α-MHC719/+, but not in pio- and R-pio-treated α-MHC719/+ mice (Figure). Whole transcriptome mRNA sequencing and proteomics analyses revealed robust upregulation in pro-hypertrophic, pro-fibrotic, oxidative stress, mitochondrial dysfunction, disturbed metabolism, and inflammation pathways in left ventricular myocardial samples from α-MHC719/+ mice compared to WT. Pio- and R-pio-treated α-MHC719/+ mice, on the other hand, presented significant improvement up to normalization in these analyses.

Conclusions: Pio and R-pio effectively prevent pathological myocardial remodeling in an HCM mouse model by limiting processes associated with hypertrophy and fibrosis, together with a reduction in oxidative stress, mitochondrial dysfunction, perturbed metabolic signaling and inflammation.The data suggest promise for pio and R-pio in the treatment of HCM.

Keywords: Hypertrophic cardiomyopathy, pioglitazone, myocardial fibrosis and hypertrophy, mitochondrial dysfunction, inflammation, oxidative stress

Figure

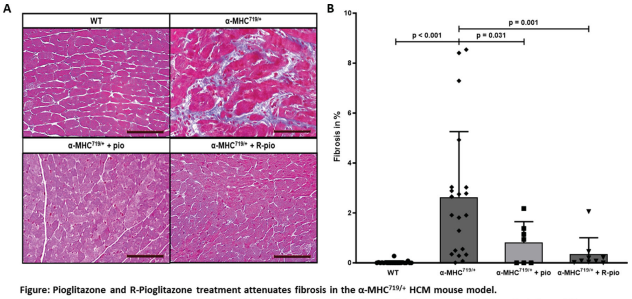

Figure: Pioglitazone and R-Pioglitazone treatment attenuates fibrosis in the α-MHC$^{719/+}$ HCM mouse model.
Panel A: Representative Masson trichrome stained histopathologic sections of left ventricular myocardial tissue. Scale bars: 100 µm.
Panel B: Quantification of fibrosis area (%) stained in blue relative to the total myocardial area in vehicle-treated wildtype (WT, n=22) and α-MHC$^{719/+}$ (α-MHC$^{719/+}$, n=21) mice, as well as pio- (α-MHC$^{719/+}$+pio, n=7) and R-pio-treated (α-MHC$^{719/+}$+R-pio, n=9) α-MHC$^{719/+}$ mice. The scatter plot depicts the mean + SD, with significances indicated by p-values calculated using the Wilson Mann-Whitney U test.

OP-056

Performance of the primacy sudden death risk prediction model for childhood hypertrophic cardiomyopathy

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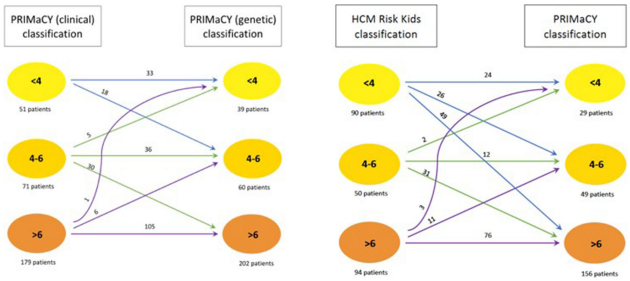
Background and Aim: The validated HCM Risk-Kids model provides accurate individualised estimates of sudden cardiac death risk in children with hypertrophic cardiomyopathy (HCM). A second validated model, PRIMaCY, also provides individualised estimates of risk, but its performance and clinical impact has not been independently investigated. The aim of this study was to investigate the clinical impact of using the PRIMaCY SCD risk model in childhood HCM.

Method: The estimated 5-year SCD risk was calculated for children meeting diagnostic criteria for HCM in a large single-centre cohort using PRIMaCY (clinical and genetic) and HCM Risk-Kids model and model performance was assessed.

Results: 301 patients [median age 10 (IQR 4-14)] were followed up for an average of 4.9 (+/- 3.8) years, during which 30 (10.0%) reached the SCD or equivalent event end-point. Harrell's C-statistic for the clinical and genetic models was 0.66 (95% CI 0.52 – 0.8) and 0.66 (95% CI 0.54 – 0.80) with a calibration slope of 0.19 (95% CI 0.04-0.54) and 0.26 (95% CI -0.03 – 0.62), respectively. For patients who had undergone genetic testing (n=207), forty-eight patients (23.2%) were assigned to a higher risk group by the PRIMaCY model following inclusion of genetic testing results. When comparing PRIMaCY (clinical) and HCM Risk-Kids, there was between-model agreement in the calculated risk group for 118 patients (50.4%). Of the remaining 116 patients, 101 (87.1%) were assigned to a higher estimated risk group using the PRIMaCY model. The proportion of patients above the threshold ≥6% for ICD implantation was 158 (67.5%), 135 (57.7%) and 94 (40.2%) for the PRIMaCY genetic, PRIMaCY clinical and HCM Risk-Kids model respectively. The number needed to treat to potentially treat one life-threatening arrhythmia for the PRIMaCY genetic, PRIMaCY clinical and HCM Risk-Kids models was 14.5, 13.7 and 9.4 respectively.

Comparison of 5-year estimated risk for patients using a) PRIMaCy genetic and PRIMaCy clinical model b) PRIMaCY clinical and HCM-Risk Kids



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Conclusions: Although PRIMaCY has a similar discriminatory ability to that reported for HCM Risk-Kids, estimated risk estimates did not correlate well with observed risk. A higher proportion of patients met ICD thresholds using PRIMaCY model compared to HCM Risk-Kids. This has important clinical implications as these patients will be exposed to a life-time risk of complications and inappropriate therapies.

Keywords: Sudden death, childhood, risk, prediction, cardiomyopathy, implantable cardioverter defibrillator

OP-058
High prevalence of myocardial inflammation and genetic predispositions in children with severe forms of cardiomyopathies

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Background and Aim: Pediatric patients with acute or chronic heart failure may need mechanical circulatory support or orthotopic heart transplantation, respectively. The availability of donor hearts is limited, especially in this age group. Cardiomyopathy or myocarditis are the leading cause for heart failure therapy, but differentiation of genetical or/and inflammatory cause is crucial. It remains unclear, if pathogenic genetic variants may be associated or detectable also in inflammatory cardiomyopathic presentation.
Method: We included pediatric patients, who underwent heart transplantation (HTx), mechanical circulatory support (MCS) or in hospital treatment for acute heart failure between 1990-2021. We excluded patients with congenital heart disease. DNA was extracted from blood or myocardial sampling during or after treatment. Patients were sequenced by Illumina technology and SNPs were classified according to current ACMG guidelines. Histological examination was performed to investigate inflammation and cardiotropic agents to prove myocarditis by an experienced pathologist in all patients. Where possible familial co-segregation was examined.
Results: We included 104 genotyped individuals (53% males). 83% had a dilated cardiomyopathy and 47% carried a (likely) pathogenic (PLP) variant with TNNI3 as the dominantly affected gene. 86% received HTx or MCS, 15 received in hospital treatment. In 83/104 myocarditis was examined. 66/83 had histological signs of myocarditis with chronic lymphocytic myocarditis in 52%. Data from genotyping and myocarditis were available: In 44% with and 59% without myocarditis PLP variants were found. 57 received MCS, 7 cases were were weaned. 4/7 cases, all lacking PLP variants, were successfully weaned from MCS. In 37 cases familial disease was examined revealing in 30% de novo PLP variants.
Conclusions: In severe forms of pediatric cardiomyopathies genetic etiologies are frequent. Etiological conclusions based on histological biopsy examination without genetic information may underestimate the leading cause of pediatric heart failure. Genotyping of pediatric patients with severe heart failure should always be considered and may have consequences for families and MCS treatment. Genetic counseling by a multidisciplinary team is mandatory and has also the potential to support families in a difficult psychosocial situation.

Keywords: Myocarditis, genetic disease, cardiomyopathy, mechanical circulatory support, heart transplantation

OP-059
Effects of troponin-mutations in pediatric myocardium on alternative splicing and gene expression

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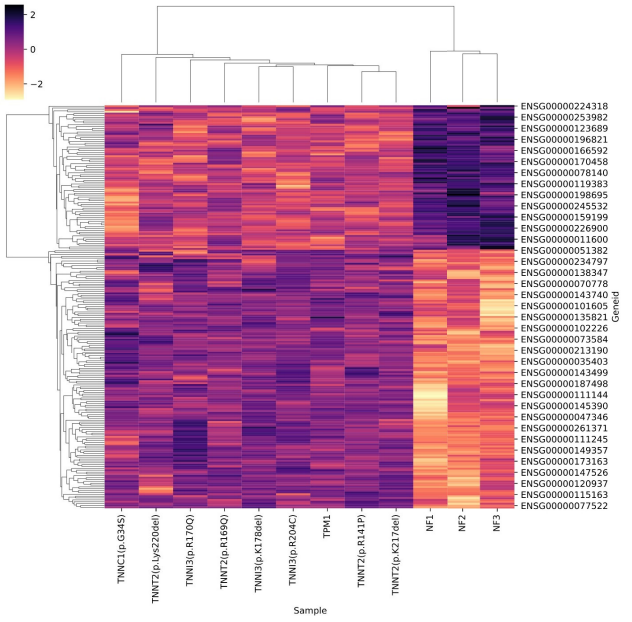
Background and Aim: In the last years, several mutations in the Troponin complex encoded by the genes TNNI3, TNNT2 and TNNC1 including TPM1 have been found to be one of the leading causes of pediatric cardiomyopathies. We were able to identify mutations in this complex leading to dilated cardiomyopathy (DCM) or restrictive (RCM) in pediatric cases of HTx. Alternative splicing and gene expression profiles in genotyped pediatric myocardium was analysed and compared to non-failing controls. Alternative splicing of the troponin complex is not completely understood especially in TNNT2 where 18 different protein-coding transcripts have been reported in databases.

Figure 1: Heatmap for differentially expressed genes for 9 patients bearing mutations in the troponin complex and 3 non-failing patients



Purple indicates increased expression; Yellow indicates decreased expression. Depicted are the differentially expressed genes with their gene IDs

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Method: We isolated total RNA from the left ventricle (LV) of 60 pediatric patients aged 2 months to 18 years, with cardiomyopathies and 3 non-failing controls. The pediatric patients were stratified by genetic variants in TNNT2, TNNC1, TPM1 or TNNI3, with miscellaneous genetics and cardiomyopathies lacking genetic evidence. After RNA isolation, myocardial samples were sequenced by Nanopore-technology and bioinformatically evaluated on differential gene expression and alternative splicing.
Results: We found significant gene expression changes in the failing pediatric myocardium in comparison to controls. Apart from typical cardiomyopathy-associated genes like NPPA and NPPB, we identified numerous genes to be differentially expressed in our RNA-seq data including METTL12A and TIMM8B (Figure 1) which have not been described in genomics of adult hearts so far. Additionally, our study sheds a light on the relevance of troponin transcripts in the context of cardiomyopathies.
Conclusions: Our findings provide evidence in molecular differences of early childhood cardiomyopathies caused by troponin variants. The genotype specific transcriptome and alternative splicing will be subject of future research aiming to improve treatment of these patients.

Keywords: pediatric cardiomyopathy, troponin, alternative splicing, gene expression, mutation

OP-060
RNA-sequencing unveils novel *FLT4* splice site variants in isolated CHD

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Background and Aim: Congenital heart disease (CHD) affects more than 8 in 1000 live-born babies and may occur isolated or in the context of a syndromic constellation. The pathophysiology of CHD is complex with both genetic and environmental contributions, and the etiology remains unknown in the majority of the patients. Heterozygous loss-of-function (LOF) pathogenic variants (PVs) in *FLT4* have recently been associated with Tetralogy of Fallot (TOF). Here we report on two novel families with LOF PVs in *FLT4*, further contributing to the molecular etiology of isolated CHD.
Method: In 318 probands with isolated CHD and normal copy number variant analysis, we performed trio exome sequencing (ES) on genomic DNA followed by analysis of a virtual congenital heart disease gene panel including 471 genes. *FLT4* (NM_182925.5) variants were classified according to the American College of Genetic and Genomic Medicine (ACMG) guidelines. We applied RNA-sequencing on lymphocytes to evaluate the consequences of variants predicted to affect splicing of *FLT4*.
Results: We identified novel PVs in *FLT4* in two families with non-syndromic CHD. In family one, two siblings diagnosed with TOF and major aortopulmonary collateral arteries (MAPCAs) harbored a heterozygous c.985+1G>A *FLT4* variant, inherited from their asymptomatic father. In family two, the proband presented with bicuspid aortic valve and aortic coarctation and harbored a heterozygous c.1657+6T>C variant in *FLT4*. Of note, the variant was inherited form her asymptomatic mother, but occurred de novo in the maternal grandmother who was diagnosed with TOF. For both variants RNA-sequencing revealed skipping of exon 7 and 12 in the c.985+1G>A and c.1657+6T>C variant respectively, resulting in a premature termination codon, reclassifying both variants as class 5 (pathogenic). Notably, previous targeted RT-PCR analysis for the c.1657+6T>C variant revealed normally.
Conclusions: Our report widens the clinical presentation of heterozygous splice site variants in *FLT4*, that further show reduced penetrance. Reanalysis of patients with recent diagnostic techniques such RNA-sequencing are powerful to elucidate some of the missing heritability in isolated CHD.

Keywords: Isolated CHD, Genetics, RNA-sequencing, FLT4

OP-061
A new truncating MYBPC3 variant for hypertrophic cardiomyopathy detected in the north of spain

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Background and Aim: Hypertrophic cardiomyopathy (HCM) is a disease prevalent in our setting that is genetic based and inherited through families. Genotype-phenotype association studies have shown a degree of variable penetrance for many variants, while the influence of other endogenous or environmental factors has not been well established. Most pathogenic variants are limited to a small number of families or individuals, so the study of recently identified pathogenic variants that affect homogenous populations represents a great opportunity for the description of clinical phenotypes. Our main objective is to describe the clinical phenotype, penetrance, and prognosis of a new MYBPC3 variant, p.Val931Glyfs*120, present in several families from northern Spain, to adapt the monitoring and advice given to these patients.
Method: We describe the clinical characteristics of a new truncating MYBPC3 variant, p.Val931Glyfs*120, in 75 subjects from 18 different families from northern Spain with the p.Val931Glyfs*120 variant. Index cases were recruited from the Cardiology Units of our center; a pedigree was designed for each case and a genetic study was offered to first-to-third-degree relatives. Probands were studied by Next generation sequencing (NGS), and family members were studied by Sanger sequencing of the variants detected. A medical history was produced for all the carriers, and they were all given a physical examination, ECG, echocardiogram (and magnetic resonance imaging (MRI) if indicated) according to the latest HCM guidelines.
Results: Our cohort allows us to estimate the penetrance and prognosis of this variant. The penetrance of the disease increases with

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age, whereas 50% of males in our sample developed HCM by the age of 36 years old, and 50% of women developed the disease by the time they reached 48 years of age (p = 0.104). Men have more documented arrhythmias with potential risk of sudden death (p = 0.018), requiring implantation of cardioverter defibrillators (p = 0.024). Semi-professional/competitive sport among males is related to earlier onset of HCM (p = 0.004).
Conclusions: The p.Val931Glyfs*120 truncating variant in MYBPC3 is associated with a moderate phenotype of HCM, with a high penetrance, onset in middle age, and a worse outcome in males due to higher risk of sudden death due to arrhythmias.

Keywords: MYBPC3, genotype-phenotype, hypertrophic cardiomyopathy, sarcomeric gene variant

Table 1

	Carrying Patients with HCM			
	All	Men	Women	p
N	44	28	16	
Current age *	55 ± 17	56 ± 16	55 ± 20	0.846
Diagnosis age	41 ± 17	39 ± 15	46 ± 19	0.206
Reason for diagnosis:				
Symptoms	20 (45.5%)	14 (50%)	6 (37.5%)	0.382
Family history of disease	19 (43.2%)	10 (35.7%)	9 (56.3%)	
Irregular ECG	5 (11.4%)	4 (14.3%)	1 (6.3%)	
Thickness of left ventricle wall (IVS)	20.5 ± 5.8 (Range 13.0–43.0)	21.4 ± 6.3 (Range 13.0–43.0)	18.9 ± 4.6 (Range 13.0–26.0)	0.208
Thickness of posterior wall (LVPW)	12.2 ± 4.5 (Range 6.6–30.0)	12.3 ± 4.8 (Range 7.6–30.0)	11.8 ± 3.9 (Range 6.6–20.0)	0.759
Hypertrophy type:				
Apical	1 (2.3%)	0 (0%)	1 (6.3%)	0.346
Asymmetric septal	31 (70.5%)	21 (75%)	10 (62.5%)	
Concentric	12 (27.3%)	7 (25%)	5 (31.3%)	

Table 1. Characteristics of the 44 affected MCH carriers of MYBPC3 p.Val931Gly*120. Data are expressed as: Nº (%) or Yes/No (% Yes) or mean ± standard deviation. Decreased patients were excluded from the calculation of average age.

*Characteristics of the 44 affected hypertrophic cardiomyopathy carriers of MYBPC3 p.Val931Glyfs*120.*

OP-062
Genotype and cardiac outcomes in pediatric dilated cardiomyopathy: A single center cohort

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Background and Aim: Dilated cardiomyopathy (DCM) is the most common phenotypic pattern of cardiomyopathy in childhood. Pediatric dilated cardiomyopathy (DCM) progress to heart failure, death, or transplantation within 2 years of diagnosis. Clinical correlation between genetic testing and outcomes in pediatric DCM remains unclear. Our objective is to provide genotype associations with life-threatening cardiac outcomes in our pediatric DCM population
Method: We perform a retrospective review of children with DCM followed at our outpatient clinic at a pediatric tertiary referral center during period 2018–2023, excluding syndromic, chemotherapy-induced, congenital heart disease causes and other types of cardiomyopathies. Genetic variants were adjudicated by whole exome test or panel DCM.
Results: We identify 79 children with DCM during this period. Of those, 46 patients (58.2%) were tested with a cardiomyopathy genetic test (31 patients with +ve genetic study and 13 patients were -ve and 2 patients have pending result) and genetic testing were not done for 33 patients (26%). Patients with +ve genetic test had a more severe cardiac outcome 42% serious outcomes (5 died, 5 transplanted, 2 listed for transplant, one patient underwent LVAD, waiting for Donner) while39% improving and 19% have static condition, comparing with the patients with negative gene study, 31% had severe cardiac outcome (3 patients have been transplanted and one listed for heart transplant, 46% are improving and 23% has static condition. 19% of genotype +ve patients has arrhythmia (67% ventricular and 33 supraventricular tachycardia) while 36% genotype -ve patients have arrhythmias inform of isolated premature ventricular contraction.
Conclusions: Genotype were associated with significant cardiac outcome. Some DCM genotype is associated with high risk or arrhythmia and rapid progression of the disease so an early extensive genetic and clinical evaluation is very important for better outcome

Keywords: Dilated cardiomyopathy (DCM), genotype

OP-063
Genetic testing provides insight on precocity and severity of pediatric cardiomyopathies

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Background and Aim: The genetics of cardiomyopathies (CM) have been studied for decades in adults, but only recently in children, and no association with childhood mortality has been reported. To describe the genetic spectrum of pediatric CM (pCM) and compare it with that of adults to assess penetrance. Then, to assess whether genotype in children are associated with increased risk of death or heart transplantation.
Method: We performed a retrospective cohort study of patients diagnosed with primary CM who underwent genetic analysis between 2014-2022 in 32 French tertiary centers. Genetic results from the pediatric cohort were compared to an index CM case with an adult-onset cohort sequenced on the same genes panel. We defined a variant of interest (VOI) as a certainly/likely pathogenic variant and variant of uncertain significance with additional proof of pathogenicity according to the ACMG-AMP classification. Survival analysis was performed to avoid left truncation. A Cox proportional hazards model was used to investigate risk factors.

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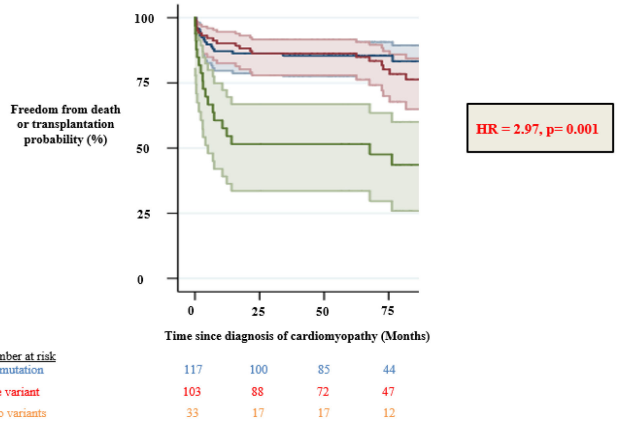
Results: 1719 patients with CM were included, including 256 (14.9%) pCM and 1463 (85.1%) adult CM (aCM). Compared to aCM, pCM had higher rates of patients with one VOI (53.7% vs. 47%, p=0.001), two VOI (23% vs. 15.6%, p=0.02), and of de novo variants (6.3% vs. 0.1%, p <0.001). Additionnaly, we observed different genes and allelic spectrum between aCM and pCM. The median follow-up for pCM was 5.7 years (IQR, 2.8 to 7.0 years) and 64 patients (25.3%) had a cardiovascular event. Survival at one and six years was 84.1% CI95% [79.0% - 88.1%] and 78.1% CI95% [72.2% - 82.9%], respectively. At six years, freedom from death or heart transplantation was worse in patients with two mutations (43.6%CI95% [25.9% - 60.3%], p < 0.001) compared to those without mutations (85.4% CI95% [77.6% - 90.7%]) (Figure 1). Age at diagnosis less than 1 year (HR = 2.07, p= 0.03), restrictive CM (HR = 2.87, p= 0.03) and two VOI mutations (HR = 2.97, p= 0.001) were independent risk factors for death or heart transplantation.
Conclusions: Genetic testing for pCM has identified a high proportion of VOI that have a significant association with survival in these children.

Keywords: genetic, pediatric cardiomyopathies, mortality

Figure 1: Time to survive or transplant according to genetic status



Sports Cardiology, Physical Activity and Prevention

OP-064
Use of new paediatric VO2MAX reference equations to evaluate aerobic fitness in overweight or OBESE children with congenital heart disease

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Background and Aim: Overweight and obesity in children with congenital heart disease (CHD) represent an alarming cardiovascular risk. Promotion of physical activity and cardiac rehabilitation in this population requires assessing the level of aerobic fitness (VO2max) by a cardiopulmonary exercise test (CPET). Nevertheless, the interpretation of CPET in overweight/obese children with CHD remains challenging as VO2max is affected by both the cardiac condition and the body mass index (BMI). The new paediatric VO2max Z-score reference equations, based on a logarithmic function of VO2max, height and BMI, were applied to overweight/obese children with a CHD, and compared to overweight/obese children without any other chronic condition.
Method: This cross-sectional study was carried out from November 2010 to March 2020 in two paediatric CPET laboratories in France. All children aged 5–18 years, with a BMI>85th percentile, and who underwent a CPET in one of the two centres, were screened. They were recruited after a regular paediatric cardiology outpatient visit. A BMI>85th percentile defined overweight, and a BMI>95th percentile defined obesity, Two groups were identified: overweight or obese children with CHD ("CHD group") and overweight or obese children without any other chronic condition ("control group").
Results: In this cross-sectional controlled study, 344 children with a BMI>85th percentile underwent a CPET (54% boys; mean age 11.5±3.1 years; 100 CHD; 244 controls). Using the VO2max Z-score equations, aerobic fitness was significantly lower in obese/overweight CHD children than in matched obese/overweight control children (-0.43±1.27 vs. -0.01±1.09; p=0.02, respectively) and the proportion of children with impaired aerobic fitness was significantly more important in obese/overweight CHD children than in matched controls (17% vs.6%, p=0.02, respectively). The paediatric VO2max Z-score reference equations also identified specific complex CHD at risk of aerobic fitness impairment (univentricular heart, right outflow tract anomalies). Using Cooper's weight and height-based linear equations, similar matched-comparisons analyses found no significant group differences.
Conclusions: As opposed to the existing linear models, the new paediatric VO2max Z-score equations can discriminate the aerobic fitness of obese/overweight children with CHD from that of obese/overweight children without any chronic disease.

Keywords: cardiopulmonary exercise test, obesity, congenital heart disease, children, Z-score

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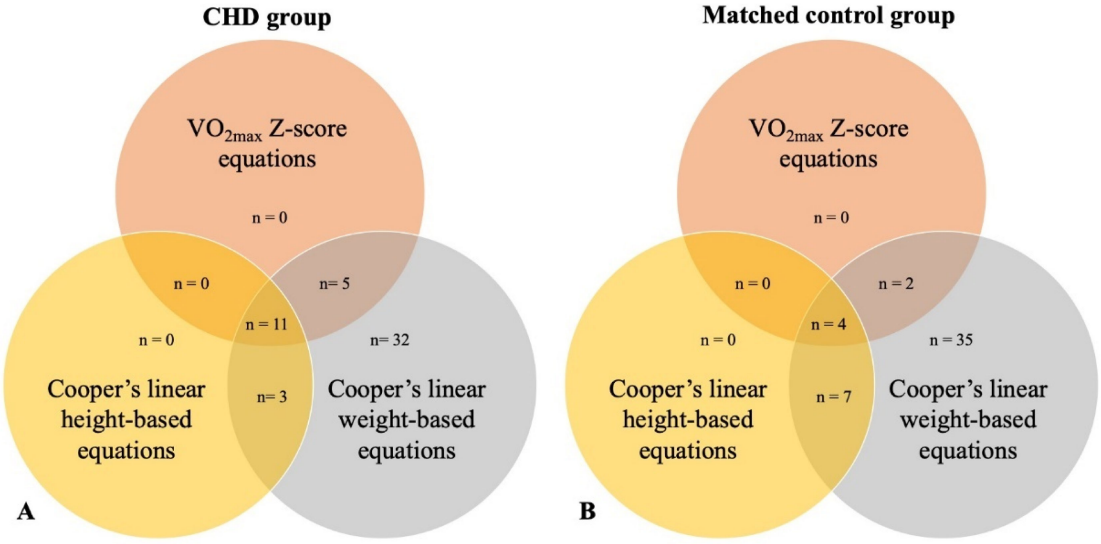
Distribution of impaired aerobic fitness according to three different VO2max reference equations for CHD and matched controls.



Patients with impaired aerobic fitness in CHD (A) and matched control (B) groups, according to the three VO2max equations, using Venn diagrams. This figure shows which subjects are identified by the same equations (intersection of the circles).

OP-065

Early hybrid cardiac rehabilitation in congenital heart disease: The qualirehab multicentre randomized controlled trial

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Background and Aim: Cardiopulmonary fitness in youth with congenital heart disease CHD decreases faster than in the general population, resulting in impaired health-related quality of life (HRQoL) and increased long-term cardiovascular risk. Cardiac rehabilitation reduces cardiac morbidity and improves HRQoL in adult heart failure, but in youth with CHD, the level of evidence for physical activity interventions remains limited and is not part of the standard of care. The aim is to assess the impact of a combined centre and home-based cardiac rehabilitation program on HRQoL in youth with CHD.

Method: The QUALIREHAB multicentre, randomized, controlled trial evaluated and implemented a 12-week center and home-based hybrid cardiac rehabilitation program, including multidisciplinary care and physical activity sessions. Adolescent and young adult CHD patients with impaired cardiopulmonary fitness were randomly assigned to either the intervention or the standard of care. The primary outcome was the change from baseline to 12-month follow-up in HRQoL using the PedsQL total score in an intention-to-treat analysis. The secondary outcomes

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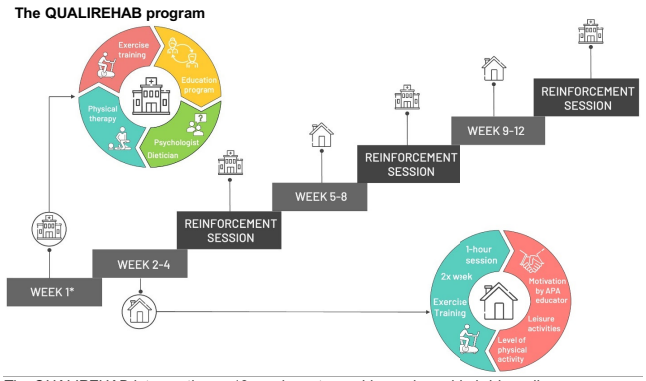
were the change in cardiovascular parameters, cardiopulmonary fitness, and mental health.
Results: A total of 142 patients were enrolled in the study (mean age 17.4±3.4 years, 52% female) and randomly assigned to the rehabilitation intervention group or to the control group. Patients assigned to the intervention had a significant positive change in HRQoL total score (mean difference = 3.8;95% CI = [0.2;7.3]; P=0.038; effect size = 0.34), body mass index (BMI) (mean difference = -0.7 kg/m2; 95%CI = [-1.3; -0.1]; P = 0.022; effect size = 0.41), level of physical activity (mean difference = 2.5; 95% CI = [0.1;5]; P = 0.044; effect size = 0.39), and disease knowledge (mean difference = 2.7; 95% CI = [0.8;4.6];P = 0.007; effect size = 0.51). The per-protocol analysis confirmed these results with a higher magnitude of differences. Acceptability, safety, and short-time effect of the intervention were good to excellent.
Conclusions: This hybrid cardiac rehabilitation program improved HRQoL, BMI, physical activity, and disease knowledge, in youth with CHD, opening the field to implement prevention programs for other childhood diseases, and also for adults with CHD.

Keywords: congenital heart defect, physical activity, physical fitness, patient education, quality of life, exercise therapies



The QUALIREHAB intervention: a 12-week center and home-based hybrid cardiac rehabilitation program.

Figure 1

OP-066
Nutritional status in school-aged children and adolescents with congenital heart defects in Germany - comparison with data from the KIGGS study

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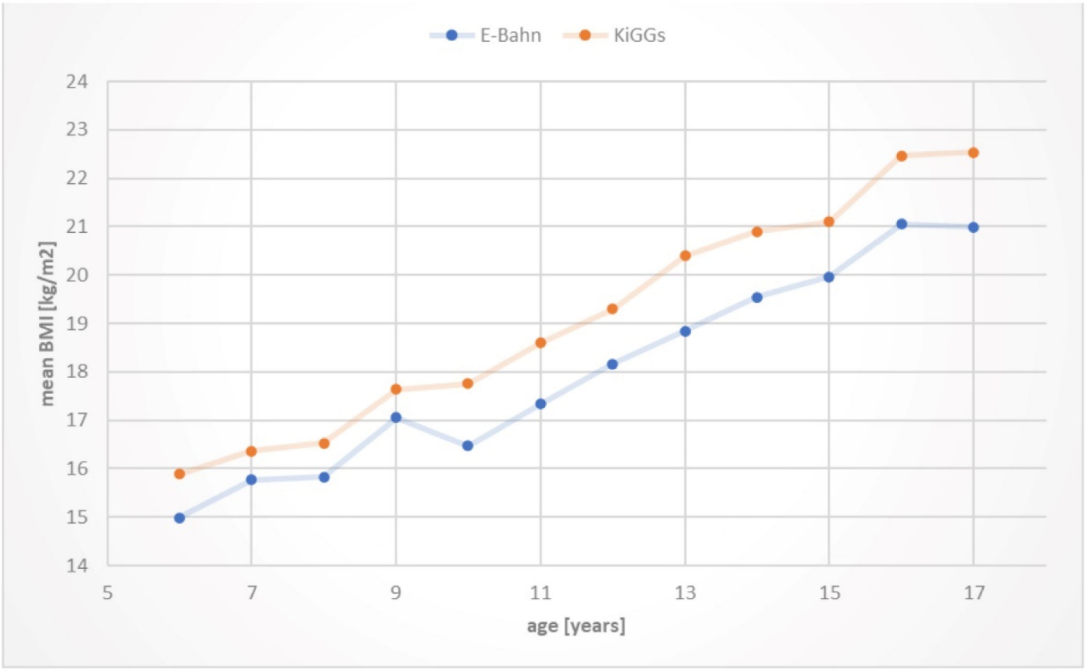
Background and Aim: Malnutrition and poor weight gain are frequent features in congenital heart disease (CHD). In order to



Age-related mean BMI of children with CHD (E-Bahn) compared to an age-matched healthy reference group (KiGGS). Children with CHD showed significantly lower BMI values (p<0.001).

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obtain representative data on the nutritional status and potential influencing factors in school-aged children and adolescents with CHD, we performed the nationwide survey "E-Bahn" (Ernährung bei angeborenen Herzfehlern).
Method: All patients aged 6 to 17 years registered in the German National Register for Congenital Heart Defects (NRCHD) were contacted by email and asked to participate in the survey using the comprehensive questionnaire of the "Eating study as a KiGGS (German Health Interview and Examination Survey for Children and Adolescents) Module", which assessed the self-reported dietary habits of children and adolescents in Germany. This allowed the comparison with a representative age-matched subset of 4,569 participants of the KiGGS study.
Results: Complete datasets of 894 patients (mean age of 12.5 ± 3.0 years; 47.2% female) were available for evaluation. The study participants were allocated according to anatomic complexity into simple (23.8%), moderate (37.8%) and complex CHD (38.4%).
The consumption of sugar-containing food ($p<0.001$) and fast food ($p<0.05$) was significantly lower among CHD-patients as in healthy children. Consequently, overweight and obesity, increasingly seen in the general population, was only rarely detected in CHD-patients (5.0%, and 0.9%, respectively).
More common was underweight: Compared to their healthy peers, children with CHD showed significantly lower body mass index (BMI) ($p<0.001$) (Fig. 1), while children with complex and moderate CHD had the lowest BMI.
Multiple regression analyses revealed age, physical activity, sugar-containing food, CHD severity and number of previous interventions as potential contributing factors for malnutrition in CHD patients, but in contrast to KiGGS, no correlation was found between BMI and consumption of fast food.
Conclusions: According to this nationwide survey, school-aged children with CHD living in Germany showed significantly lower BMI than their healthy peers. Children with complex and moderate CHD had the lowest BMI. BMI was related to age, CHD severity, and number of interventions, but also to physical activity and sugar-containing food. Preventative measures including promotion of exercise and nutritional counseling should therefore be considered as important components of multidisciplinary care for children with CHD.

Keywords: prevention, nutrition, exercise

OP-067

Longitudinal health related physical fitness in children with congenital heart disease - Narrowing the gap

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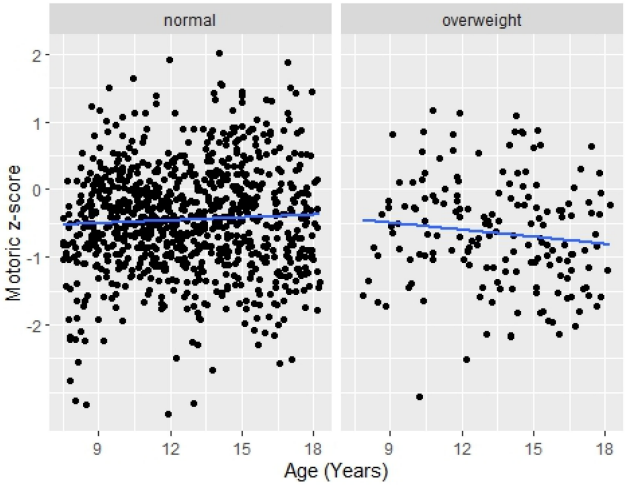
Background and Aim: Children with congenital heart diseases (CHD) have impaired health related physical fitness (HRPF). However, its development over time has not yet been investigated. The aim of this study was to evaluate the natural course of HRPF over time from longitudinal assessment in children with CHD.
Method: In this longitudinal study, 370 children and adolescents (11.7 ± 2.6 years, 37 % girls) with various CHD were prospectively examined from July 2014 to October 2023. Over a mean follow-up length of 2.6 ± 1.6 years, 747 follow-up assessments have been conducted. HRPF was assessed by FintessGram test battery. The battery consists of five tasks, namely push-ups, curl-ups, trunk lift, sit and reach and shoulder stretch. Results were transformed into standard deviation scores (z-score) and converted to an overall motoric z-score. A linear mixed effect model was performed to examine the course of motoric z-scores over time.
Results: Baseline motoric z-score of children with CHD was -0.64 ± 0.78 and therewith significantly reduced to healthy peers ($p<0.001$). Fortunately, motoric z-score increased significantly over time by 0.07 per year ($p<0.001$) which means that the deficits decrease over time. The longitudinal increase of motoric z-scores over time remained significant when including sex ($b=0.08$, $p<0.001$), BMI ($b=0.07$, $p<0.001$), disease severity ($b=0.08$, $p<0.001$) and CHD type ($b=0.05$, $p=0.035$) into the model. Remarkably, there was a significant decrease over time in motoric z-scores in overweight CHD children ($b=-0.04$, $p=0.034$) and girls ($b=-0.04$, $p=0.031$).
Conclusions: Overall, this study shows a longitudinal increase in motor z-scores, which can be seen as a good sign that children with CHD compensate for their initial deficits as they grow up. Monitoring of the HRPF from childhood to adulthood and beyond is indicated with a special focus on patients with overweight and girls.

Keywords: physical fitness, motoric fitness, congenital heart disease

Longitudinal development of motoric z-score by sex



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General Cardiology

OP-068

Association between pre-existing maternal cardiovascular disease and cardiovascular disease in children – cohort study of 2.6. million children

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Background and Aim: The question if maternal cardiovascular conditions are associated with increased risk of longer-term cardiovascular disease in children is understudied. We aimed to investigate the association between pre-existing maternal cardiovascular disease (CVD) and CVD in children occurring from infancy to early adulthood, using paternal CVD as negative control comparison.
Method: We performed a population-based cohort study on national registers in Sweden, including singletons live-born without major malformations or congenital heart disease between 1992 and 2019. Children were followed from age 1 until CVD diagnosis, death, emigration, or December 2020, whichever came first. As exposure, we defined any pre-existing maternal CVD (e.g., cerebrovascular, ischemic heart disease, arrythmia, hypertension) diagnosed before conception. The primary outcome was the first incidence of any CVD recorded after the first year of life in the offspring (e.g., cerebrovascular disease, arrythmia and hypertension). Cox proportional hazards models were fitted to estimate Hazard Ratios (HRs), adjusted for important maternal characteristics. Using paternal CVD as negative control, we compared maternal-offspring and paternal-offspring associations to assess shared genetic and environmental confounding.
Results: The study population included 2,597,786 children. Among them, 26,471 (1.0%) were born to mothers with pre-existing CVD. During a median follow-up of 14 years from age of 1 year, 17,382 children were diagnosed with CVD (incidence rate 5.0 per 10,000 child-years). After adjusting for potential confounders, children of mothers with CVD had 2.1 times higher HR of CVD (95%CI: 1.83-2.40), 2.62 times higher HR of arrythmia (95% CI: 2.17-3.17) and 1.63 times higher HR of hypertensive disease (95% CI: 1.18-2.25), compared with offspring of mothers without CVD. Stratification by maternal CVD subtypes showed increased hazard of CVD in children for maternal arrythmia, vascular heart disease and structural heart disease. Paternal CVD was also associated increased HR of CVD, but these associations were weaker than those with maternal CVD.
Conclusions: In this cohort study, maternal CVD was associated with increased risk of CVDs in children during childhood and young adulthood. The parental-comparison results suggest that the observed association was unlikely to be fully explained by shared familial factors, such as genetic liability.

Keywords: Maternal cardiovascular disease, paternal cardiovascular disease, offspring cardiovascular disease, developmental programming, negative control exposure

OP-069

Transcriptomic profiling and commonly-affected genes in both mothers and neonates of preeclampsia: A case-control study

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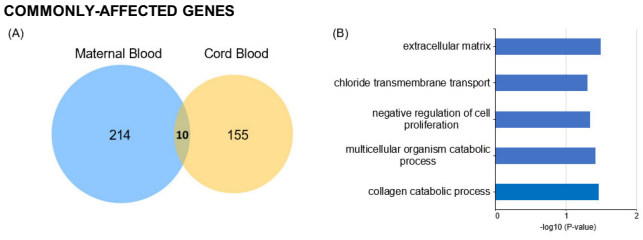
Background and Aim: Both women with the pregnant history of preeclampsia (PE) and the offspring of maternal PE are at high risk of premature cardiometabolic disorder. However, the underlying mechanism remains undiscovered. Thus, we proposed and aimed to explore whether there were certain genes which should be commonly affected in both during perinatal period–the timing of simultaneous exposure to PE.
Method: This prospective case-control study was conducted in the Departments of Pediatrics and Obstetrics at Kaohsiung Chang Gung Memorial Hospital in Taiwan. In addition to the clinical parameters, the gene-expression profiles of antepartum maternal (MBLs) and cord blood leukocytes (CBLs) were performed by using RNA-seq analysis and further confirmed by real-time RT-PCR.
Results: Forty pairs of mother-neonate of PE and of healthy normotension controls were recruited. Using RNA-seq analysis, the transcriptomic profiles of MBLs and CBLs were investigated, revealing that 10 genes were commonly affected in both mothers and neonates. Further validation showed that lamin A/C (LMNA) expression levels in both MBLs and CBLs were significantly higher in PE. In CBLs, the mRNA expressions of ADAM metallopeptidase with thrombospondin type 1 motif 2, gamma-aminobutyric acid A receptor epsilon (GABRE), microtubule associated serine/threonine kinase family member 4 (MAST4), oncostatin M, and solute carrier family member 1 were significantly higher, and matrix metallopeptidase 8 (MMP8) was significantly lower in PE. In the adjustment model of gestational age, LMNA expression remained higher (adjusted odds ratio, 1.68) in PE MBLs. The MAST4 (2.55) and GABRE expressions (1.66) remained higher, and MMP8 (0.54) remained lower in PE CBLs.
Conclusions: In this study, we demonstrated the transcriptomic profiles individually in mothers and neonatal offspring of PE and identified ten genes commonly affected in both, which are involved in the cardiovascular and metabolism-related pathways. Therefore, the results of our study may provide a basis for future research in exploring the pathogenetic mechanism of cardiometabolic disorders or screening those at risk following the exposure to PE.

Keywords: cardiometabolic, offspring, preeclampsia, RNA-seq analysis, transcriptomic profiling



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Sports Cardiology, Physical Activity and Prevention

OP-070

A randomized trial of home-based high-intensity interval training to improve exercise capacity in patients with tetralogy of fallot

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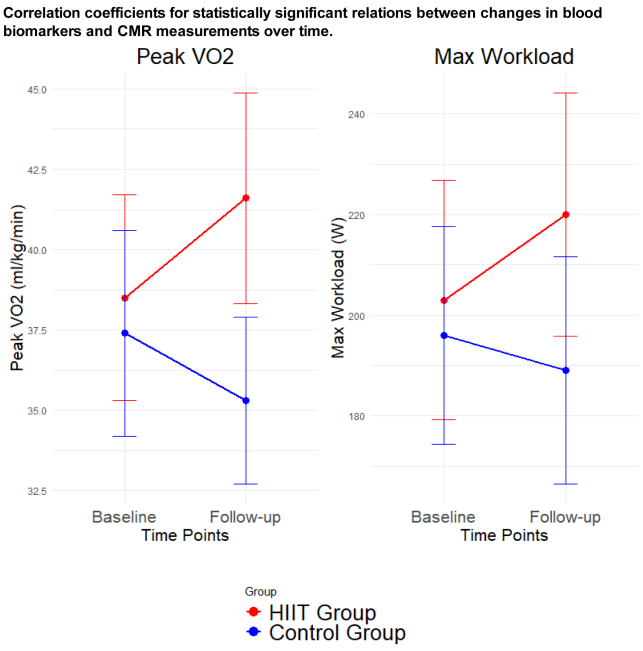
Background and Aim: Reduced exercise performance is a common finding in Tetralogy of Fallot(ToF) patients, which relates to adverse (long term) outcomes. There is increasing evidence on the beneficial effects of on-site supervised-based exercise training in ToF patients, but the effects of home-based exercise training are unknown. We aimed to examine the effect of a home-based, online monitored exercise program in patients with repaired TOF on exercise capacity.

Method: Patients aged 12-30 (median 16 (IQR 14-23), 65 % male) years with repaired ToF were randomized to a 12-week home-based high-intensity interval training program (HIIT) or standard of care (SoC). HIIT was prescribed 3 times 30 minutes a week. Patients were monitored by heart rate telemonitoring (Polar M9, Polar Electro, Kempele, Finland) and MS Teams video calls (1 in 3 HIIT sessions). Patients randomized to SoC were instructed to exercise as usual. The primary endpoints were increase in peak VO2 and maximum workload. Secondary endpoints were measurements of cardiac structure and function (biventricular volumes and ejection fraction) as assessed with magnetic imaging.

Results: A total of 34 patients were included in this study. No serious adverse events were recorded. After HIIT, peak VO2 increased from 38.5 ml/kg/min ± 8.3 to 41.6 ml/kg/min ± 8.3 (mean increase 3.1 ml/kg/min ± 3.1, $p < 0.001$). Workload increased 203 watt ± 62 to 220 watt ± 63 (median increase 20 watt IQR (12 – 26, $p < 0.001$), compared to a mean decrease control group -2.1 ml/kg/min ± 2.4 ($p = 0.001$) and median change in workload of 0 watt IQR (-15 - 0) ($p = 0.06$) (See figure 1). Changes in peak VO2 were independent of baseline fitness levels, BMI and biventricular cardiac size and function. No changes in cardiac volumes and ejection fraction were observed after HIIT.

Conclusions: In patients with ToF aged 12-30 years, a 12 week home-based, online monitored HIIT-program appeared safe and feasible and improved exercise capacity with ± 10%. The effect of HITT intervention was not related to cardiac status and baseline exercise.

Keywords: Tetralogy of Fallot, exercise training, HIIT, peakVO2, cMRI



Pulmonary hypertension, heart failure and transplantation

OP-071

ABO-incompatible heart transplants in older children: A new paradigm in transplant?

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Background and Aim: Paediatric heart transplantation is hampered by the limited availability of donor organs. ABO-incompatible heart transplantation is well established for patients <2 years of age, however limited data is available for older children and the practice is not widely adopted. Our first ABO-incompatible transplant was performed in 2001, and over the past decade we have considered ABO-incompatible transplant in patients aged 2-9 years if isohaemagluttinin titres were 1:32 or less. Herein we report the outcomes of those that received an ABO-incompatible transplant. Further, as we routinely measured isohaemagluttinin titres in all listed children in this age group, we also explore the potential for ABO-incompatible transplant.

Method: Data were retrospectively analysed on all paediatric heart transplants undertaken at our centre between 1st January 2013 and 1st June 2023. Primary outcomes were isohaemagluttinin titres at listing and whether an ABO-incompatible transplant was performed. Secondary outcomes were survival and incidence of rejection over the follow-up period, up to October 2023.

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Results: In this 10-year period there were 61 heart transplants for children aged 2-9 years, and 48 of these had isohaemagluttinins measured at the time of listing. 42 patients (88%) had isohaemagluttinins titres to Anti-A or Anti-B of 1:32 or less and were listed as being suitable for ABO-incompatible transplant.
Of the 42 patients identified as being suitable for ABO-incompatible transplant, 13 patients received ABO-incompatible organs. All 13 are alive at the time of follow-up, including 6 patients that have survived >5 years. 3 patients have received treatment for rejection, 2 were asymptomatic, and there were concerns of immunosuppression compliance in the patient with symptomatic rejection.
Conclusions: • ABO-incompatible heart transplant is safe in selected patients up to 9 years of age. Our oldest ABO-incompatible heart transplant was in an 8.6-year-old recipient. • In our cohort, most listed children 2-9 years of age had isohaemagluttinin titres in the range that ABO-incompatible transplant could be considered. • These findings may have a substantial impact on donor availability world-wide. • Further work is needed to explore whether the age range for ABO-incompatible heart transplant can be extended. • Interestingly, our data shows no correlation between age and isohaemagluttinins.

Keywords: Heart Transplant, ABO-incompatible, isohaemagluttinin titres

OP-072

Inflammasomme in experimental pulmonary arterial hypertension, a natural history study

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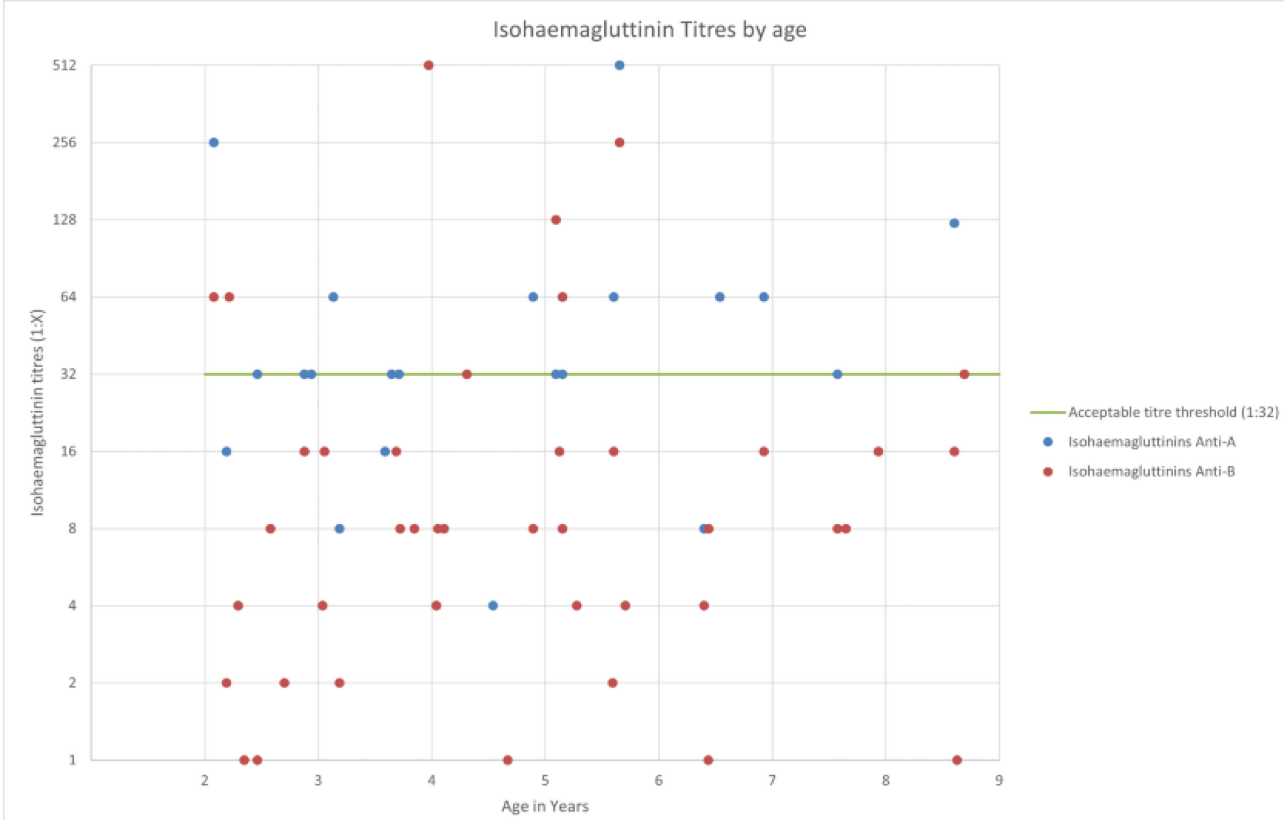
Background and Aim: We have shown previously that the NLRP3 inflammasome is activated in lungs of the rat model of monocrotaline and flow induced neo-intimal end-stage experimental pulmonary arterial hypertension (PAH), and that this aggravates PAH and was accompanied by increased macrophage accumulation. Additionally, suppressing inflammasome activation by pirfenidon led to amelioration of PAH. We now studied inflammasome activation at earlier stages of PAH to evaluate when these inflammatory pathways are activated.
Method: We used the well characterized rat model of PAH that is treated with monocrotaline or NaCl injection for controls on day 1 and undergoes aortocaval/shunt construction or sham operation on day 8, respectively. This model shows a gradual increase in

Isohaemagluttinin titres by age



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vascular occlusion of intra-acinar pulmonary arterioles up to end stage neointimal disease on day 28 and a dramatic worsening in pulmonary arterial pressure between day 14 and 28. To evaluate location of inflammasome activation, we stained for NLRP3 and α-smooth muscle actin on lung sections, and assessed caspase-1, IL-1β and IL-18 cleavage by Western blot on lung homogenates on day 8, 14, and 28. We concomitantly stained for CD68 to assess macrophage accumulation.

Results: NLRP3 staining in perivascular CD68+ macrophages increased at day 8 with further increases at day 14, and 28, compared to controls, accompanied by an increase in perivascular macrophage accumulation. Moreover, we observed gradual increase of NLRP3 positive cells located in the intima of intra-acinar arterioles (diameter ~50μm) as well as of pre-acinar PAs (diameter100-250μm). PAH rats showed increased cleavage of caspase-1, a hallmark of inflammasome activation on lung homogenates at day 8, 14, and 28 compared to controls. IL-1β and IL-18 cleavage were increased in a similar manner. (figure)

Conclusions: The NLRP3 inflammasome is activated early in PAH, and activation increases further during development of the disease,

figure

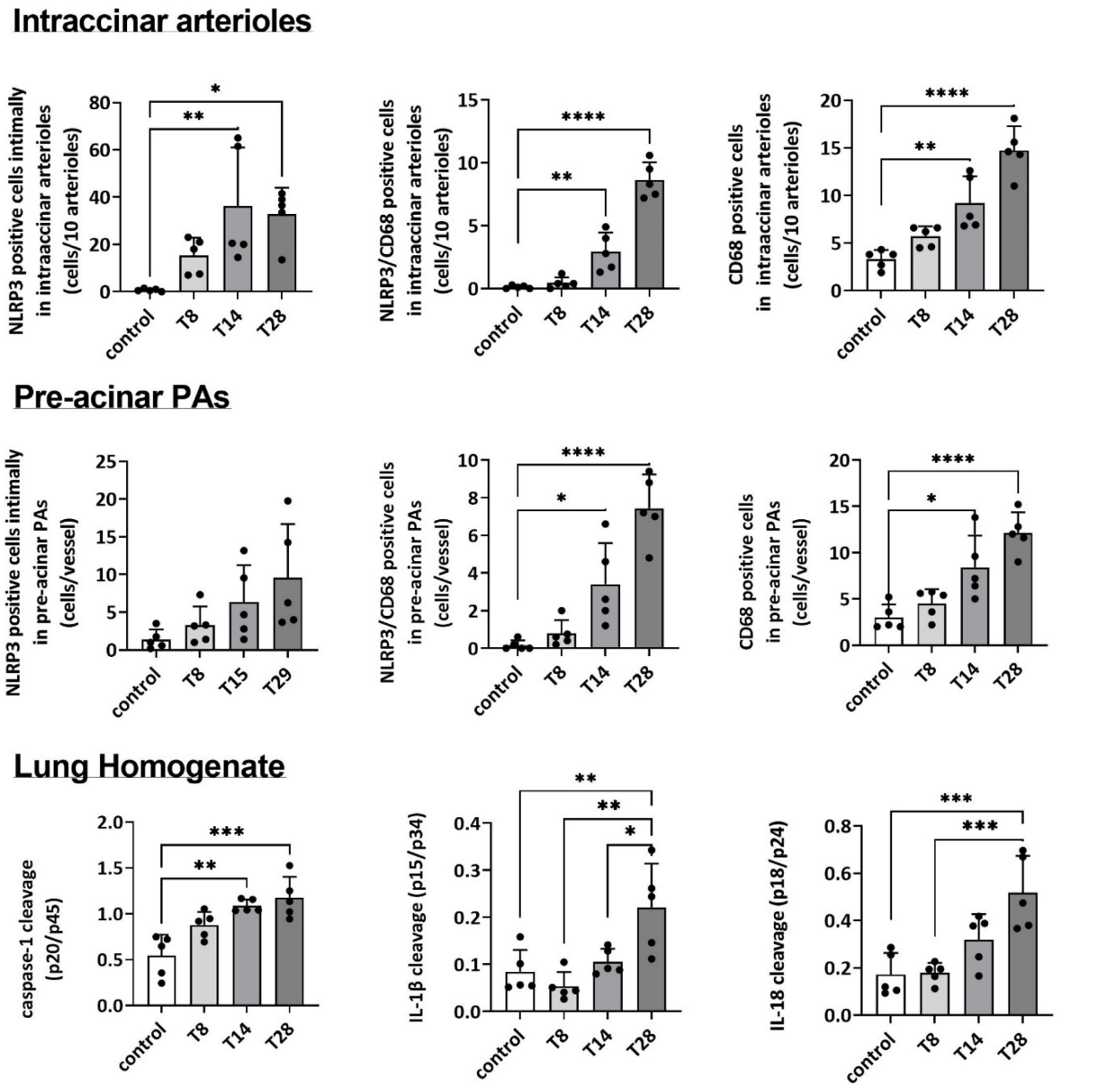

Figure: Pulmonary arterial hypertension was induced by monocrotaline injection on day 1 followed by shunt operation on day 8 Groups control (NaCl and sham operation), T8 (sacrificed on day 8 after sham operation), T14 (sacrificed on day 14) and T28 (sacrificed on day 28). Intraacinar arterioles: diameter <50μm; pre-acinar PAs: diameter 100-250μm; *p<0.05, **p<0.005, ***p<0.0005, ****p<0.00005

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accompanied by macrophage accumulation. Inhibitors of NLRP3 inflammasomes may prevent deterioration of PAH when used at early disease stages, which we will evaluate in future studies.

Keywords: PH, PAH, NLRP3, inflammasome, inflammation, vascular remodelling

OP-073

Fontan-associated liver disease (FALD) in the eurofontan experience. An insight into european awareness

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Background and Aim: The diagnosis, impact and implications of Fontan-Associated Liver Disease (FALD) in the long-term follow-up of Fontan patients are subjects of intense debate within the field of cardiology.

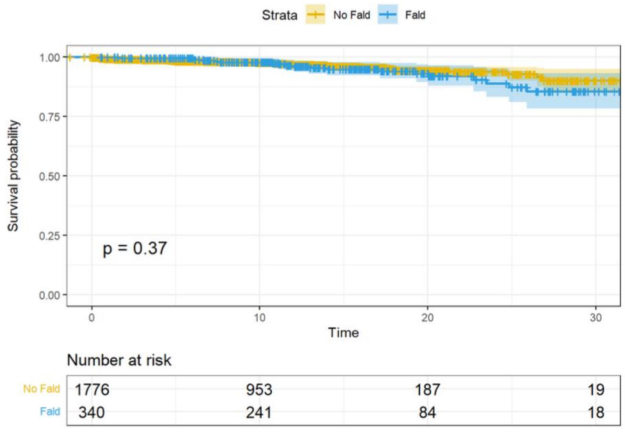
Method: Conducted as a retrospective multicenter analysis, this study encompasses patients undergoing Fontan completion between 1990 and 2022, exhibiting FALD during the long term follow-up. FALD was defined as a spectrum of time-related structural and functional liver modifications due to congestive hepatopathy, ranging from mild liver fibrosis to liver cirrhosis and hepatocellular carcinoma. Patients were included according to a multiparametric evaluation incorporating clinical (symptoms and signs), laboratoristic (liver function-synthesis, cholestasis and viral hepatitis markers, INR, alpha-fetoprotein) and instrumental (abdomen ultrasound, transient elastography, MRI, CT, biopsy) data.

Results: Among 21 centers involved in the EUROFONTAN experience, only 14 routinely assessed liver function during long-term follow-up after Fontan completion. This study includes 342 patients (16,1% of 2132) diagnosed with FALD at a median follow-up of 14 years. Of these patients, 198 (58%) were males and FALD showed no significant association with gender (p=0.5). The most frequent anatomical diagnosis was tricuspid atresia or pulmonary atresia with intact ventricular septum (30%). Among the 19 (5,6%) patients with FALD that died during the long-term follow-up, 5 (1,5%) died for advanced liver disease/cancer. The remaining patients exhibited NYHA classes as I (46%), II (40%), III (14%) and IV (1%). Statistical analysis revealed significant associations between FALD and absence of fenestration (p<0,001), systemic ventricular and atrio-ventricular valve dysfunction (p<0,001), tachyarrhythmias (p<0,001) and liver stiffness ≥22kPa in transient elastography (p<0,001) during the long-term follow-up. FALD was also significantly associated with late death due to liver disease/cancer (p<0.001) but not with abnormal liver function tests (p=0.037). The analysis demonstrated growing FALD-prevalence with increasing time from Fontan completion, although no significant association with worse survival was observed (p=0.37).

Conclusions: This analysis, derived from the first European multicenter study, underlines the significant incidence of FALD occurring late after Fontan completion. Moreover, this study highlights the urgent need for universally shared diagnostic criteria and clinical surveillance protocols to rigorously monitor and promptly diagnose FALD during the long-term follow-up.

Keywords: Fontan, follow-up, FALD, congestive hepatopathy

FALD survival



OP-074

European cooperation: evaluation of the MYKKE risk prediction score in pediatric patients with myocarditis data from the swedish swedcon registry

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Background and Aim: Pediatric myocarditis is heterogeneous and may lead to severe heart failure. The Swedish prospective national register for pediatric heart disorders (SWEDCON) includes >50,000 pediatric patients, with high internal and external validity and population-level coverage. The German prospective multicenter registry for pediatric myocarditis "MYKKE" with >750 enrolled patients has developed a risk model – the MYKKE-Score – to predict severe disease courses. We aimed to validate the MYKKE-Score with the data from the Swedish SWEDCON registry.
Method: Clinical characteristics of children enrolled in SWEDCON were analyzed between 2009 and 2022. Descriptive epidemiology was investigated according to STROBE guidelines. The MYKKE risk prediction model is based on the analysis of 20 baseline variables of 414 enrolled patients according to the primary combined outpoint of major adverse cardiovascular event (MACE: inotropic support, need for mechanical circulatory support, heart transplantation and death), using logistic regression models for final variable selection. Initial validation was performed on a second MYKKE cohort of 108 patients. In this study, the SWEDCON cohort is to be used for further validation.
Results: In the SWEDCON registry 35 centers included a total of 306 cases, representing a cumulative incidence of 13.9 cases per 100,000 children. The data showed a significant predominance of male cases (77.5%) with a bimodal age distribution at diagnosis: 15% of cases were found in children under the age of 3, while 70% occurred in children over 13 years old. The median age was 15.7 years. The mortality rate was 1.3%. Demographic results were similar to the data from the MYKKE registry. For the final risk model, age and left ventricular ejection fraction (LVEF) could be selected. Their regression coefficients allow the calculation of the predicted probability for a MACE using following sum: $n = -0.111 * LVEF\ (in\ \%) - 0.150 * age\ at\ admission\ (in\ years)$. The AUC for the validation cohort was 0.936 (95% CI: 0.89-0.99). The validation of the SWEDCON cohort is in progress.
Conclusions: In this nationwide, population-level study on pediatric myocarditis, we showed the results of the MYKKE registry to be applicable in a different European population identifying age and LVEF as main risk factors.

Keywords: myocarditis, heart failure, outcome, risk prediction, risk score

OP-075

Eosinophilic gastrointestinal disease in children with heart transplantation: Case series

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Background and Aim: Paediatric heart transplantation (HT) is a life-saving therapy, but with considerable morbidity.

Table 1. Dates of diagnosis and treatments

	BIRTHDATE	DIAGNOSIS	HT DATE	INDUCTION THERAPY	MAINTENANCE IMMUNOSUPPRESSION	DIAGNOSIS EoE	EoE TREATMENT
CASE 1	07/02/2012	Severe aortic stenosis Left ventricle fibroelastosis	16/07/2012	Basiliximab Mycophenolate Corticosteroids Tacrolimus	Tacrolimus	12/2020	PPIs Fluticasone Exclusion diet
CASE 2	16/01/2006	Unbalanced atrioventricular septal defect	15/06/2006	Basiliximab Azathioprine Corticosteroids	Tacrolimus Sirolimus Mycophenolate	04/2018	PPIs Budesonide Fluticasone
CASE 3	03/12/2007	Hypoplastic left heart syndrome	14/04/2008	Basiliximab Mycophenolate Corticosteroids Tacrolimus	Tacrolimus	04/2016	PPIs Fluticasone Exclusion diet
CASE 4	27/05/2004	Transposition of the great arteries	15/06/2005	Corticosteroids Tacrolimus Azathioprine	Tacrolimus Mycophenolate	06/2015	PPIs Fluticasone Exclusion diet

EoE: Eosinophilic esophagitis; PPIs: Proton-pump inhibitors

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Autoimmune and allergic diseases are significant because of the selective suppression of TH1 immune response and promotion of the TH2 lymphocytes, resulting in an imbalance with eosinophilic activation. However, scarce data is available about eosinophilic esophagitis (EoE) in HT. We aim to describe our paediatric patients with diagnosis of EoE after HT.
Method: Descriptive case series including all children who underwent HT between 2005 and 2021 in a single centre. We reviewed the immunosuppressive treatment, digestive symptoms, allergy history, endoscopic evaluation and outcome.
Results: There were 137 HTs. Of them, 29 died in the first year after HT, leaving 108 in follow-up; 47 patients presented gastrointestinal symptoms and an endoscopic study was performed in 25 of them. Only 4 were diagnosed with eosinophilic gastrointestinal disease, all of them had EoE and one also had eosinophilic gastritis. The four patients were transplanted in the first 12 months of life because of congenital heart disease, and EoE was diagnosed 8 to 11 years after HT. The induction and maintenance immunosuppressive therapy is described in Table 1. Only one patient developed humoral graft rejection 16 years after transplantation. In the follow-up, all of them presented food allergies, atopic dermatitis and cheilitis, and 3/4 bronchospasm history and eosinophilia. Initial gastrointestinal symptoms were choking and esophageal impaction in 2/4, dysphagia in 3/4, and one referred nonspecific presentation as chronic abdominal pain and anaemia. Upper endoscopies showed whitish exudates, esophageal trachealization and histological eosinophil count >15Eo/hpf. Treatment was started with proton-pump inhibitors, but they were switched to topic steroids in all patients for bad tolerance, treatment failure or relapse. All of them reached clinical and histological remission with this treatment 5 to 15 months later. None of the patients had EBV disease nor developed post-transplant lymphoproliferative disease.
Conclusions: EoE is one of the more significant manifestations of HT immunologic disbalance. It seems to affect patients who were transplanted earlier and who have atopy history. This disease is usually responsive to treatment, but a high index of suspicion is important due to the nonspecific clinical presentation.

Keywords: Heart transplantation, eosinophilic esophagitis

Cardiac Imaging

OP-076
Complete isolated vascular rings: More than 20 years experience at a single tertiary referral center

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Background and Aim: BACKGROUND: the term Complete Vascular Ring (CVR) encompasses various malformations of the aortic arch that can compress trachea and/or esophagus, posing challenges in postnatal diagnosis due to subtle symptoms. Prenatal diagnosis, however, can alter clinical approaches and surgical timing.
Objectives: to outline our experience managing pre and postnatally diagnosed CVR, highlighting the evolving diagnostic and treatment strategies at our Center over the past two decades
Method: We included all patients with prenatal or postnatal diagnosis of CVR from January 2000 to December 2022. Demographic data (preoperative clinical status, timing of diagnosis, cross-sectional imaging, operative and perioperative details, and clinical outcomes) were collected for all CVR patients.
Study population was divided in two groups, fetal (Group A) versus postnatally diagnosed (Group B) and among each group we identified those diagnosed between 2000 to 2010 and those between from 2011 to 2022. A multidisciplinary approach determined surgery based on respiratory or swallowing issues, confirmed by imaging and bronchoscopy
Results: Our study includes 125 CVR patients (Table 1), 57% diagnosed prenatally. Fetal diagnosis frequency increased in the last decade, with three terminations, two associated with 22q11 microdeletion. Fetal diagnoses correlated with younger age at initial imaging and surgery. Surgical techniques varied by CVR type, with no tracheal reconstructions. No early or late deaths occurred. Long-term symptoms persisted in two prenatal cases versus 23% postnatally.
Conclusions: Prenatal detection rate improved over time, prompting earlier interventions without increased morbidity or mortality. Early CVR diagnosis and referral to specialized centers can prevent long-term complications, especially tracheal compression-related issues, improving respiratory outcomes. Our current policy advocates early treatment for symptomatic patients, elective repair at 6-9 months for double aortic arch (DAA), and at 12-18 months for right aortic arch and aberrant left subclavian artery (RAA ALSA), even if asymptomatic, considering significant tracheal compression or Kommerell's diverticulum aneurysm.

Keywords: Complete vascular ring, Fetal Echocardiography, Surgery, Tracheomalacia, Congenital heart disease

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Table 1

Table 1. Characteristics of study population

	Group A	Group B	Tot Cohort
All Patients	71	53	124
Diagnosis			
- *DAA n(%)*	18(25)	25 (47)	
- *RAA n(%)*	51(72)	28 (53)	
ALSA	*46*	21	
C-RAA	5	7	
RDA	2	-	
RDA and LDA	4	-	
- *C-LAA n(%)*	2(3)	0	
RDA and ARSA	1		
RDA,LDA and ARSA	1		
Time Period at Diagnosis			
- *2000-2010*	15	14	
- *2011-2023*	56	39	
TOP *n(%)*	3(4)	-	
Chromosomal anomaly *n(%)*	10(14)	7(13)	
22q11del	7	3	
Other	3	4	
Age at symptoms(months), median(range)	7 (2-24)	28 (2-112)	
Age at first imaging (months), median(range)	13 (1-132)	55(1-240)	
Age at surgery (months)			
- *2000-2010*	*24* (3-92)	73 (2-180)	
- *2011-2023*	*10* (2-35)	51 (1-174)	
Post-operative complication			
- *2000-2010*	2	1	
- *2011-2023*	*5*	8	
Maximum FU (months)	68 (5-288)	93 (3-210)	
Lost at long term FU n(%)	3/68 (4)	3/53 (5)	
Asymptomatic patients at last FU n(%)	63/65 (97)	34/50(68)	

DAA double aortic arch; RAA right aortic arch; C-RAA circonflex right aortic arch; C-LAA circonflex left aortic arch; RDA right ductal arch; LDA left ductal arch; ARSA aberrant right subclavian artery; ALSA aberrant left subclavian artery; FU follow up.

Characteristics of study population

General Cardiology

OP-077

Fontan associated liver disease – impact of cardiac function on disease progression

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Background and Aim: Fontan associated liver disease (FALD) is a long-term sequela of the Fontan circulation. Chronic venous congestion is thought to be the main driver of fibrosis. FALD screening with abdominal ultrasound and laboratory testing is part of the routine follow-up in our centre and is performed every 2 to 3 years. Serial screening data was evaluated to identify potential risk factors for FALD progression.

Method: Patients living with a Fontan circulation for a minimum of 5 years who underwent serial FALD screening were eligible for

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inclusion. A "liver disease score (LDS)" incorporating items from abdominal ultrasound and blood testing was calculated to grade FALD severity (5 items each, maximum score 10 points). Data from echocardiography, cardiac MRI and cardiac catheterization were included to evaluate potential hemodynamic risk factors for FALD progression.
Results: Serial data from FALD screening was available in 115 patients (male: n=65, female: n=50, HLHS: n=63). Data from the first screening visit was obtained 11.2 (IQR 8.0-14.4) years after surgery. Median LDS increased from 3 (2-4) to 4 (3-5) points over a period of 5.6 (4.3-6.6) years ($p<0.001$). There was no difference in FALD progression between HLHS and non-HLHS patients (1.0 (0.0-1.6) vs. 1.1 (0.0-2.0) points/5 years, $p=0.489$) or between patients with a systemic right and left ventricle (1.1 (0.0-1.8) vs. 0.9 (0.0-1.7) points/5 years, $p=0.637$), respectively. Indexed end-diastolic and end-systolic ventricular volumes assessed by MRI correlated with LDS difference (EDVi: $r=0.197$, $p=0.037$; ESVi: $r=0.293$, $p=0.002$). Central venous pressure (13 ±3 vs. 10 ±2 mmHg, $p<0.001$) and end-diastolic pressure of the systemic ventricle (9 ±4 vs. 7 ±2 mmHg, $p=0.004$) were higher in patients with LDS score > 4 points at the second visit.
Conclusions: Abdominal ultrasound and laboratory abnormalities suggestive of FALD are common during routine follow-up in adolescent and young adults with Fontan circulation. Progression can be seen within a relatively short period of follow-up irrespective of the underlying ventricular morphology. Indexed systemic ventricular volumes were weakly correlated to the rate of progression. More advanced liver disease at the most recent visit was related to higher central venous pressure and diastolic dysfunction.

Keywords: Fontan associated liver disease, Fontan circulation, single ventricle palliation

Pulmonary hypertension, heart failure and transplantation

OP-078
4D Flow cardiac magnetic resonance imaging to assess pulmonary blood flow in patients with pulmonary arterial hypertension associated with congenital

Estibaliz Valdeolmillos[1], Hichem Sakhi[2], Marine Tortigue[1], Marion Audie[1], Marc Antoine Isorni[3], Emmanuelle Fournier[1], Olivier Sitbon[4], David Montani[4], Laurent Savale[4], Xavier Jais[4], Marc Humbert[4], Arshid Azarine[2] and Sebastien Hascoet[1]
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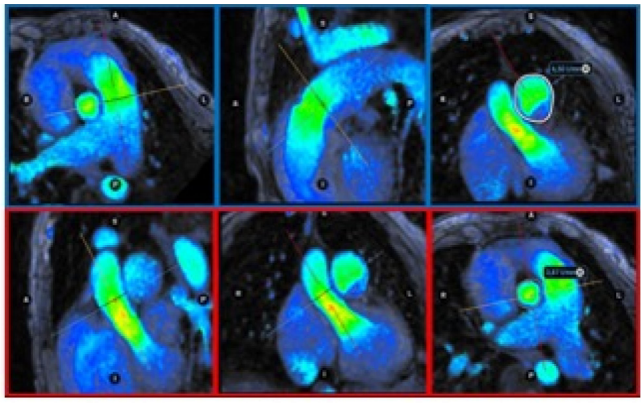
Background and Aim: Right heart catheterization (RHC) is the gold standard to assess hemodynamics in pulmonary arterial hypertension associated with congenital heart diseases (PAH-CHD). Four-dimensional flow cardiac magnetic resonance (4D flow CMR) has emerged as a promising non-invasive imaging technique for comprehensive assessment of cardiovascular hemodynamics. This study aims to evaluate the accuracy of 4D flow CMR, compared to RHC, in measuring the pulmonary flow (Qp), the systemic flow (Qs) and the ratio of pulmonary to systemic flow (Qp/Qs) in PAH-CHD.
Method: In this single centre cohort study (4DflowHTAPCC) 64 patients (mean age was 45.3 (±13.7) years old) were prospectively included with PAH-CHD predominantly presenting with pre-tricuspid shunt (n=50, 78%). Qp, Qs and Qp/Qs were measured invasively using direct Fick principle during RHC and compared with measurements assessed by 4D flow CMR within a 24-48-hour window.
Results: The average mean pulmonary artery pressure was 51 (±17) mmHg with median pulmonary vascular resistance of 8.8 Wood Unit [5.3-11.7]. A strong linear correlation was observed between Qp measurements obtained from 4D flow CMR and the Fick method ($r = 0.962$, $p <0.001$). The Bland-Altman analysis indicated a mean difference of 0.152 l± 0.481 L/min. The Qs and Qp/Qs also exhibited excellent correlation ($r=0.851$ and $r=0.923$, both $p<0.001$)
Conclusions: Qp as measured by 4D flow CMR shows a strong correlation with measurements derived from the direct Fick method. Further investigation is needed to develop less complex and standardized methods for measuring essential PAH parameters, such as pulmonary arterial pressures and pulmonary vascular resistance.

Keywords: Pulmonary arterial hypertension, congenital heart disease, atrial septal defect, cardiac magnetic resonance.

Placement of regions of interest (ROIs) at the level of the pulmonary artery (above in blue) and the aorta (below in red).



General Cardiology

OP-079
Risk factors for reintervention following treatment for aortic coarctation in sweden: From simple coarctation to complex congenital heart disease

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Background and Aim: Repaired coarctation of the aorta (CoA) – either simple or as part of complex congenital heart disease (CHD) - can be associated with long-term sequalae such as recoarctation (reCoA) and/or hypertension. The aim of this study was to determine the prevalence and risk factors for requiring reintervention for reCoA in patients with simple CoA as well as in the setting of complex bi- and univentricular CHD.
Method: Data on patients with a diagnosis of CoA was retrieved from the Swedish National Registry of Congenital Heart Disease (SWEDCON) from 1973 – 2020. Surgically and catheter-based repaired CoA were analysed separately.
Results: 1609 children and adults with surgically repaired CoA and 170 with catheter-based CoA intervention were included. 249 (15%) of the patients after surgically repaired CoA and 59 (35%) after catheter-based repaired CoA received interventions for reCoA. The risk of reintervention following primary surgical CoA repair at the CoA site was increased in patients with complex biventricular and univentricular CHD. Use of patch augmentation and aortic arch reconstruction through Norwood procedure were associated with increased risk of reCoA, while end-to-end or end-to-side anastomosis were associated with a decreased risk of reCoA. Younger age at primary surgical CoA repair was associated with increased risk of reCoA after primary surgical repair.
Conclusions: Our study shows for the first time that patients with univentricular and biventricular complex CHD and those following patch aortoplasty or single ventricle palliation with Norwood surgery are at a higher risk for reinterventions at the CoA site after surgical repair than patients with uncomplicated CoA.

Keywords: coarctation, aorta, re-coarctation, aortic arch hypoplasia, risk factor

Neurodevelopment and psychosocial care

OP-080

The whole is greater than the sum of its parts: Sustainable, low-cost, and impactful inpatient cardiac neurodevelopmental care

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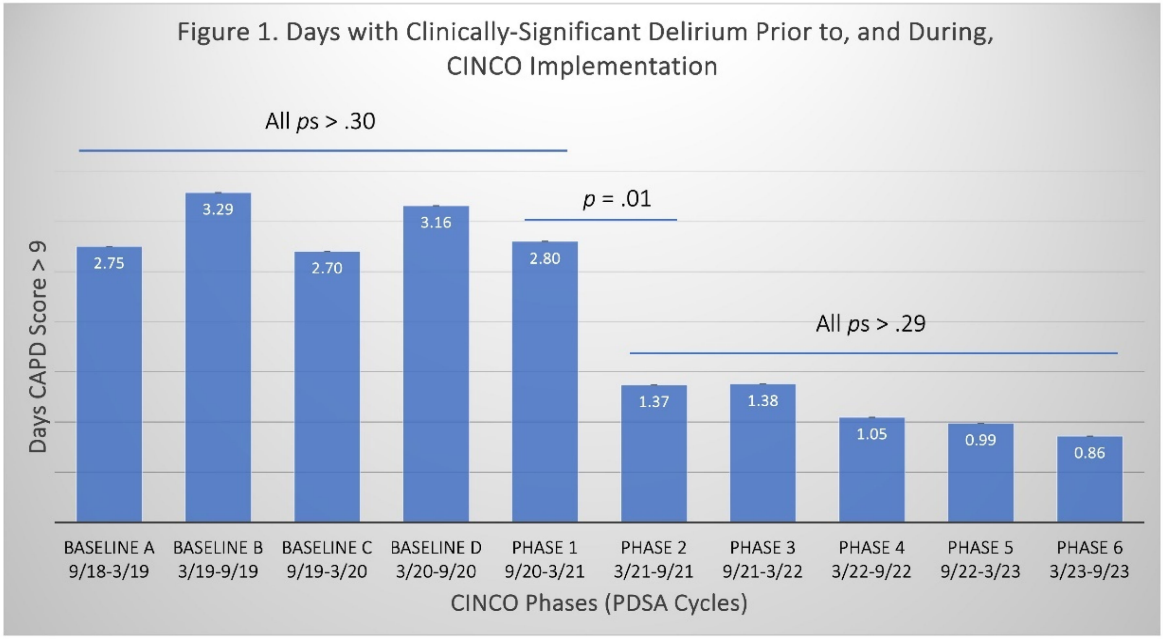
Background and Aim: Neurodevelopmental (ND) impairment is the most common long-term morbidity associated with congenital heart disease (CHD). Recent statements from the American Heart Association and the Cardiac Neurodevelopmental Outcome Collaborative emphasize the importance of high-quality inpatient cardiac ND care, which may improve outcomes. We aimed to examine the feasibility, sustainability, and impact of a low-cost inpatient ND program over a 3-year period.
Method: The Cardiac Inpatient Neurodevelopmental Care Optimization (CINCO) program was implemented at a children's hospital in the United States, starting in 2020, using a quality improvement framework. CINCO included five pillars: 1) Nursing orders for ND care; 2) Bedside ND plans and toy kits; 3) Caregiver mental health support; 4) Developmental care rounds; and 5) Specialized volunteers to provide developmental stimulation. Inclusion criteria were all patients aged 0-2 years admitted for > 7 days to the cardiac intensive or acute care units, from 9/1/20 - 9/1/23. Baseline data were obtained from 9/1/18 - 8/31/20. CINCO's impact on delirium was based on Cornell

Figure 1



Note. CAPD = Cornell Assessment of Pediatric Delirium. Data shown are average number of days with CAPD score > 9 per patient, across six-month periods. Baselines A through D were prior to the launch of the CINCO program. Phases 1-6 represent the six Plan-Do-Study-Act quality improvement cycles of the CINCO program to date.

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Assessment of Pediatric Delirium scores and examined using linear modeling.
Results: There were 847 admissions during the CINCO implementation period and 508 admissions in the pre-CINCO baseline period. Over three years, 563 patients received ND nursing orders, 331 patients received developmental kits, 509 patients received bedside developmental plans, 354 caregivers received mental health support, and 206 patients and families were seen in developmental care rounds. There were 519 CINCO volunteer shifts (1,382 patient encounters). During these, 653 patients were read books, 980 patients were held, and 950 patients were played with. The average number of days of delirium per patient was stable for 2 years before CINCO. Between Phases 1 and 2 of CINCO, delirium dropped by 51% (p=.01) and this improvement was sustained over the next 2.5 years (Figure 1).
Conclusions: By organizing a pre-existing interdisciplinary team to provide systematic, standardized inpatient cardiac ND care, with minor associated costs, the CINCO program has demonstrated feasibility and sustainability over 3 years. Beyond the intuitive impact of promoting sleep, developmental stimulation, and parent and child psychosocial health, sustained reductions in delirium scores for children age 0-2 with CHD were achieved.

Keywords: cardiac neurodevelopment, inpatient neurodevelopmental care, Cardiac Neurodevelopmental Outcome Collaborative, cardiac psychosocial care, delirium

Nursing and Allied Health Professionals

OP-081
Adolescents and young adults with congenital heart disease, down syndrome and both conditions talk about their quality of life

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Background and Aim: Advances in care have resulted in individuals with Congenital Heart Disease (CHD), Down syndrome (DS) and both conditions living longer, more productive lives. While there have been many studies focused on quality of life in families affected by CHD and DS, few have explored the perspectives of individuals living with these conditions. The aim of this study was to gain a better understanding of life with CHD, DS and both conditions from the perspective of adolescents and young adults who are living with these conditions.
Method: Semi-structured interviews were conducted with 17 individuals (6 with CHD, 7 with DS and 4 with both conditions). Prior to the interviews, participants were asked to share 15-20 pictures showing them with their favorite people and/or doing their favorite things. The pictures were used during the interview to prompt discussion, reflection and recollection. Interviews were transcribed verbatim; then they were coded using coding categories based on the Resiliency Model of Family Stress, Adjustment and Adaptation. For this presentation, the focus will be on responses to the following questions: Overall, do you think you have a good life? If so, what makes it a good life? Tell me about your strengths. Now, tell me about any challenges you face.
Results: All of the participants reported that they had a good life. In fact, many said they had a great life. Participants described a wide variety of strengths. The main challenges reported by participants were physical limitations, interactions with peers, and mental health issues.
Conclusions: Findings from this study indicate that even though life can be challenging for adolescents and young adults living with CHD, DS, or both conditions, some individuals adapt well to these challenges and others may even thrive. To gain a better understanding of their lives, we need to give them the opportunity to share their own stories and pictures of their life with one or more chronic conditions. Then, we need to acknowledge the role family and friends play. Finally, we need to tailor our interventions so that they build on strengths, rather than focus solely on problems or limitations.

Keywords: Congenital Heart Disease, Down syndrome, Quality of Life

Task Force on Clinical Drug Trials

OP-082
Theophylline effects in the fontan circulation (THIEF). Results from a phase 2 pilot study

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Background and Aim: The pharmacodynamics of theophylline match remarkably with limitations in the Fontan circulation. Our phase 2 pilot study aimed to investigate safety/feasibility and to indicate short-term effects of oral treatment with theophylline in clinically stable adolescent Fontan patients (European Medicines Agency No. EU CT 2022-500301-41-00, ClinicalTrials.gov ID NCT05717049).
Method: Eligible patients (Fontan circulation, 16-25 years of age, home address within 1 hour travel distance of nearest hospital) were identified in a nation-wide patient registry at Oslo University Hospital. Exclusion criteria were tachyarrhythmia, current use of pulmonary vasodilators, severe liver disease, pregnancy and known hypersensitivity to theophylline. Assessment before and after 12-15 weeks of oral theophylline treatment included cardiopulmonary exercise test (CPET), echocardiography, ECG, pulmonary function test, quality-of-life questionnaires (SF-36, EQ-5D-5L), and home-based polygraphy. Primary endpoints were safety/feasibility and change in cardiorespiratory fitness. Secondary endpoints were changes in health-related quality of life (HRQoL), cardiac or pulmonary function, and apnoea/hypopnoea index.
Results: From 16 screening visits, ten patients were enrolled. Three participants were excluded during the trial (non-compliance N=2, arrhythmia N=1 [extra beats/non-sustained atrial tachycardia]). There were no serious adverse events. Participants achieved therapeutic theophylline concentration levels (30-80 μmol/Liter 4-6 hours after intake) with low doses (3 females on 100 mg BID, all others on 200 mg BID). There were variable changes after theophylline treatment compared to baseline in almost every physiological and HRQoL parameter measured for primary and secondary endpoints. Individual results are given in Table 1.

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Table 1

Table 1: Results table (positive changes marked in green colour, negative changes marked in red colour)

Cardio-pulmonary exercise test																				
Participant-ID	Maximum oxygen uptake [ml/min]	Baseline	Treatment	Change (total)	Change (%)	VO_2 at VT (ml/min)	Baseline	Treatment	Change (total)	Change (%)	Maximum heart rate [bpm]	Baseline	Treatment	Change (total)	Change (%)	Heart rate reserve [bpm]	Baseline	Treatment	Change (total)	Change (%)
NO-01-001		1376	1536	160	11,6		903	892	-11	-1,2		157	172	15	9,6		70,0	85,0	15	21,4
NO-01-006		988	818	-170	-17,2		651	546	-105	-16,1		141	159	18	12,8		48,0	48,0	0	0,0
NO-01-010		1805	2092	287	15,9		1284	1227	-57	-4,4		180	174	-6	-3,3		88,0	94,0	6	6,8
NO-01-011		1734	1690	-44	-2,5		1150	1088	-62	-5,4		145	146	1	0,7		64,0	70,0	6	9,4
NO-01-012		2114	2024	-90	-4,3							173	184	11	6,4		86,0	97,0	11	12,8
NO-01-013		1655	1679	24	1,5		1374	1345	-29	-2,1		169	175	6	3,6		108,0	112,0	4	3,7
NO-01-015		1058	1395	337	31,8		943	1236	293	31,1		167	169	2	1,2		82,0	85,0	3	3,7

Cardiac function (ECG, echocardiography)															
Participant-ID	Heart rate at rest (ECG)	Baseline	Treatment	Change (total)	Change (%)	Fractional area change [%]	Baseline	Treatment	Change (total)	Change (%)	Longitudinal strain [%]	Baseline	Treatment	Change (total)	Change (%)
NO-01-001		66	74	8	12,1		42	44	2	4,8		-15,6	-11	4,6	-29,5
NO-01-006		89	81	-8	-9,0		38	39	1	2,6		-14,9	-13,2	1,7	-11,4
NO-01-010		58	62	4	6,9		43	32	-11	-25,6		-16,4	-16,6	-0,2	1,2
NO-01-011		51	77	26	51,0		28	21	-7	-25,0		-13,3	-9,4	3,9	-29,3
NO-01-012		67	76	9	13,4		41	34	-7	-17,1		-16,5	-13,7	2,8	-17,0
NO-01-013		54	54	0	0,0		54	44	-10	-18,5		-22,2	-16,6	5,6	-25,2
NO-01-015		74	89	15	20,3		30	32	2	6,7		-11,9	-13,2	-1,3	10,9

Pulmonary function											Home-based sleep study (polygraphy)		
Participant-ID	Diffusion capacity for carbon monoxide [% predicted]	Baseline	Treatment	Change (total)	Change (%)	FEV1 [L]	Baseline	Treatment	Change (total)	Change (%)	Apnea Hypopnea Index	Baseline	Treatment
NO-01-001		82	81	-1	-1,2		3,02	3,19	0,17	5,6		0 - 4	
NO-01-006		70	59	-11	-15,7		2,87	2,77	-0,1	-3,5			0 - 4
NO-01-010		66	62	-4	-6,1		4,37	4,4	0,03	0,7		0 - 4	0 - 4
NO-01-011		55	68	13	23,6		4,32	4,26	-0,06	-1,4		0 - 4	0 - 4
NO-01-012		66	65	-1	-1,5		3,97	4,03	0,06	1,5			
NO-01-013		85	72	-13	-15,3		3,33	3,29	-0,04	-1,2		0 - 4	
NO-01-015		77	79	2	2,6		2,06	2,03	-0,03	-1,5		0 - 4	0 - 4

Health-related Quality of Life (EQ-5D-5L and Rand SF-36 questionnaires)										
Participant-ID	EQ-5D-5L Self-scoring of health state [0-100]	Baseline	Treatment	Change (total)	Change (%)	SF-36 Physical sum score [10 topics, each 0-100]	Baseline	Treatment	Change (total)	Change (%)
NO-01-001		98	80	-13	-14,0		80	85	5	6,3
NO-01-006		80	89	9	11,3		80	95	15	18,8
NO-01-010		85	80	-5	-5,9		100	90	-10	-10,0
NO-01-011		80	90	10	12,5		85	100	15	17,6
NO-01-012		80	85	5	6,3		95	100	5	5,3
NO-01-013		96	80	-16	-16,7		100	100	0	0,0
NO-01-015		60	56	-4	-6,7		80	95	15	18,8

Results table (positive changes marked in green colour, negative changes marked in red colour)

There seemed to be no obvious consistent beneficial effect of treatment for any subgroup. However, several of our study participants reported to be more energetic, and they requested to continue with off label use of theophylline. We hypothesize that increased heart rate at rest and increased heart rate reserve might be responsible for subjectively positive drug effects.

Conclusions: Under close monitoring for arrhythmia and correct dosage, the use of oral theophylline seems safe in adolescents with Fontan circulation, and it has subjective positive effects in a subset of patients. The positive effect might be related to increased heart rate at rest and during activity.

Keywords: Fontan circulation, theophylline, cardiorespiratory fitness, clinical trial

Cardiovascular Morphology

OP-083
Clarifying morphology of congenitally corrected transposition of great arteries

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Background and Aim: Morphologic variations in congenitally corrected transposition of great arteries (CCTGA) determine the clinical presentation and management. The description, however, is mostly based on surgical series or autopsy studies both of which are skewed towards unfavourable morphology. There is a paucity of clinical studies providing the exact details of various malformations encountered in ccTGA. Hence, in this prospective study, we sought to clarify the clinical and morphological features of ccTGA using computed tomographic (CT) angiography.

Method: We included 140 consecutive patients of ccTGA visiting our institution between January 2020 to June 2023. Patients with heterotaxy syndrome, decompensated heart failure, and those with contraindications to CT angiography were excluded. Following clinical assessment, chest X-ray, electrocardiography and echocardiography all patients underwent CT angiography.

Results: Median age at diagnosis was 1 year with patients ranging from prenatal to 53 years. The majority (73%) were males with a male-to-female ratio of 2.7:1. Among these 118 (84.2%) patients had situs solitus while the remaining 22 (15.7%) had situs inversus. Ventricular septal defect (VSD) was present in 116 (82.8%) patients, the most common type being mal-aligned inlet and non-committed VSD. In 43 (30.7%) patients pulmonary outflow was unobstructed while among the remaining 70 (50%) and 27 (19.3%) had pulmonary stenosis and pulmonary atresia, respectively. Twenty-eight (20%) patients had tricuspid valve abnormality with hemodynamically significant tricuspid regurgitation in 9 (6.4%) patients. Any form of atrioventricular (AV) block was detected in 43 (30%) of patients while complete heart block was seen in 18 (13%) patients. Compared to patients with situs solitus, patients with situs inversus had less frequent complete heart block (15% versus 0; $p = 0.05$) and tricuspid valve abnormalities (25% versus 0; $p = 0.032$).

Conclusions: Patients with situs inversus have lesser complete heart block and tricuspid valve abnormalities. These findings and other morphologic details obtained during the study may be helpful in the optimal management of patients with ccTGA.

Keywords: congenitally corrected transposition, malaligned ventricular septal defect, situs inversus, atrioventricular block

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Nursing and Allied Health Professionals

OP-084
Improving adolescents' knowledge about their CHD through a transition program: A mixed-methods evaluation of the stepstones-CHD trial

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Background and Aim: Adolescents with congenital heart disease (CHD) need to acquire knowledge about their condition and treatment. Transition programs aim to equip adolescents with this capacity. However, evidence on the effectiveness of transition programs is limited. The aim was to investigate the effectiveness of the transition program in improving disease-related knowledge of adolescents with CHD and how the adolescents acquired this knowledge.

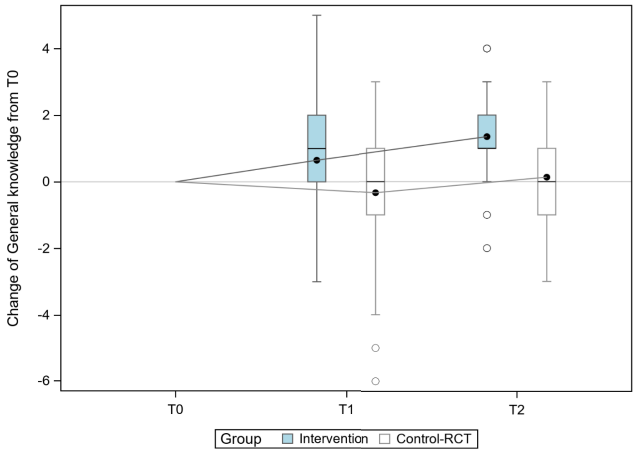
Method: The STEPSTONES transition program is an 8 components intervention. The central component is a transition coordinator who has three consultations with the participants (age 16-18.5). A randomized controlled trial was conducted in two CHD centers in Sweden. Participants were randomized to the intervention (IG; n=70) or control group (CG; n=69). Knowledge was measured at the age of 16 y (T0; baseline), 17y (T1) and 18.5y (T2) using the Knowledge Scale for Adults with Congenitally Malformed Hearts questionnaire. The total score ranges from 0-7, with higher scores denoting a higher CHD-related knowledge. Change in score between T0 and T2 was analyzed using Fisher's non-parametric permutation test unadjusted between the groups. In addition, qualitative data on participants (n=14) and parents (n=12) experiences of knowledge acquisition were collected through semi-structured interviews after participation in the intervention and analyzed with content analysis.

Results: The quantitative evaluation included 114 participants (IG; n=54, CG; n=60). Knowledge scores differed at baseline between groups (IG: 3.7±1.6; CG: 4.4±1.5; p=0.03). At T2, the knowledge score was significantly higher in the IG (5.0 ±1.3) than in the CG (4.5±1.5) (p=0.045). The change over time (T0-T2; 2.5y) was significantly different between the groups (Mean difference=1.22; 95%CI=0.60-1.82; p=0.0002). The effect size was 0.74 (moderately large effect). Qualitative data revealed that the knowledge process was person-centered, tailored to the adolescents learning style, which empowered them to take charge of their health and care. Provision of information and knowledge exchange individually and in group enhanced this process.

Conclusions: By tailoring patient education to the adolescents' learning style, this transition program was effective in increasing disease related knowledge, which is a key aspect in preparing them to assume charge of their health in adulthood and adult care.

Keywords: Transition to Adult care, Congenital heart disease, Mixed-methods, Randomized controlled trial, Adolescent

Figure 1. Change in knowledge from T0-T2 in the intervention and control group



Line in box is median and the marker shows the mean.

Neurodevelopment and psychosocial care

OP-085
Pediatric delirium outside the PICU: Insights from a pediatric cardiology ward with nurse and parent-driven detection

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Background and Aim: Pediatric delirium, a severe acute brain dysfunction, increases mortality, morbidity, and hospitalization duration. Despite its substantial impact, it often remains unrecognized, especially outside pediatric intensive care units (PICUs). This study aimed to determine delirium incidence in a pediatric cardiology ward of a university hospital. It compared patients with delirium or withdrawal to those without, exploring the feasibility of a nurse and parent-driven detection using the Sophia Observation Scale – Pediatric Delirium (SOS-PD).

Method: In a prospective study, conducted from January 1, 2022, to December 31, 2022, 596 patients were screened in the pediatric cardiology ward. Using SOS-PD, patients were assessed thrice daily during nursing routines. In a subset we additionally investigated parent-driven detection, involving 56 families from May 1, 2022, to October 31, 2022. Parent-driven SOS-PD scores were compared to nursing staff assessments. Patients with <24 hours inpatient stay or >18 years were excluded.

Results: Among 596 patients, 94 (15.8%) experienced delirium, withdrawal, or both. Patients with a positive Score (SOS-PD > 3 items) were younger than those negatively scored (mean age 1.4 vs. 5.1 years, $p < 0.01$) and received more non-opioid and opioid analgetic treatment. The majority of affected patients (89.4%) received pediatric intensive care. While PICU length of stay showed no significant difference, affected patients had a significantly longer hospital stay (mean 19.8 days vs. 6.9 days, $p < 0.01$).

Results from 990 parent-driven SOS-PD scores matched 1187 nursing staff scores, with 80% agreement on positive screening result for delirium or withdrawal. Parents selected a total of 456

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items compared to the nursing staff's 193, showing a distinct distribution pattern. Inconsolable crying stood out as the most frequently reported item by parents, at a rate of 23.2%, contrasting with the nursing team's lower rate of 4%.
Conclusions: This study confirms the significance of delirium and withdrawal in pediatric patients within a cardiology ward. It underscores the effectiveness and feasibility of nurse-driven detection using the SOS-PD score. Additionally, we demonstrated the potential of parents in detecting these conditions. These findings emphasize the need for routine screening outside the PICU and encourages the active involvement of parents in detection strategies.

Keywords: Delirium, Withdrawal, Sedation, Parents, SOS-PD

Cardiovascular Morphology

OP-086
Pulmonary atresia with "intact ventricular septum" associated with a ventricular septal defect: An apparent paradox?

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Background and Aim: Pulmonary atresia with intact ventricular septum (PA-IVS) means pulmonary valvar atresia in the absence of a ventricular septal defect (VSD). In PA-IVS, the right ventricle (RV) appears externally normal in size, but the internal cavity is hypoplastic to various degrees with major wall hypertrophy. The leaflets of the pulmonary valve are present but fused. Histology of this abnormal ventricle reveals myocardial disarray, abnormal capillary distribution, myocyte hypertrophy, and endocardial fibroelastosis. This suggests that in PA-IVS, pulmonary atresia is an acquired lesion, evolutive during fetal life.
Method: The hospital files of all children diagnosed with PA-IVS in our institution between 1981 and 2023 were reviewed, using a data warehouse (Dr Warehouse) to look for associated VSD. Patients with Ebstein anomaly and pulmonary atresia were excluded. When available, echocardiographic examinations of patients with an associated VSD were reviewed. One of these patients had severe coronary arterial anomaly leading to death and cardiac autopsy.
Results: The total cohort included 390 consecutive patients. Among those, we found 8 children with an associated VSD (2% of the cohort). According to the IPCCC-ICD11 nomenclature, the VSD was central perimembranous in 3, muscular in 4, outlet in 1. In this last patient, cardiac autopsy revealed a hypoplastic but not malaligned outlet septum, without aortic overriding, which discarded the diagnosis of tetralogy of Fallot.
Conclusions: Paradoxically, the presence of a VSD is compatible with the diagnosis of PA-IVS. This rare association cannot be classified as "PA-VSD", because this last term should be reserved to extreme forms of tetralogy of Fallot, associating outlet VSD with anteriorly malaligned outlet septum, overriding aorta and PA. Conversely, PA-IVS may be due to a specific RV myocardial disease, leading to a diminutive RV cavity, tricuspid valve dysplasia and hypoplasia, and progressive fusion of the pulmonary leaflets leading to atresia. The anatomic type of VSD, muscular or central perimembranous, and their rarity, are in favor of an incidental association. The term "PA-IVS" is too imprecise, does not reflect the anatomy and the pathogenesis of the defect. It should therefore be modified, as it is crucial to use an accurate terminology to better describe congenital heart defects.

Keywords: Congenital cardiac defect, Anatomy, Pulmonary atresia with intact ventricular septum

Neurodevelopment and psychosocial care

P-087
Results from an audit of the past decade of the ages and stages questionnaire used in our paediatric cardiac in-patient population in New Zealand

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Background and Aim: On setting up the Neurodevelopmental Follow up Program (NDFUP) in our institution in 2006 we focused on the high risk patient population, who we assessed at 2 and 4 years of age. In time we realized there were missed opportunities especially for single ventricle patients, who would normally have 2 surgeries and potentially multiple other procedures by 2 years of age. We also wanted to capture the patients consider low risk, from a surgical perspective, who had not been included in our NDFUP previously. One of our aims in introducing routine in-patient developmental screening, using the Ages and Stages Questionnaire (ASQ), was to provide an opportunity for earlier intervention.
Method: Since May 2014 on admission to the paediatric cardiac inpatient ward, in the country's only cardiac surgical centre, all New Zealand cardiac patients were given an ASQ unless they had a previously identified developmental delay or genetic condition known to effect development.
Results: Over the nearly 10 years 1,194 patients completed 1,926 ASQs with 15% of the ASQs meeting criteria for formal developmental assessment, a further 15% requiring ongoing monitoring and 70% in the normal range. The percentage (14-24%) of total ASQs that met referral criteria remained fairly static over the decade. There was a 27% referral rate in those ≤ 2 year of age compared to the total ASQs in all age groups of 15% referred.
Conclusions: We showed a consistent percentage of children needing monitoring or formal assessment over the past 10 years despite an increase in acuity over the same timeframe. The domain most often affected in our population was gross motor. With the ASQ introduction we were able to provide an opportunity for earlier intervention as 27% of our less than 2 year olds were referred for a formal assessment. We also provided, after 30% of the ASQs, age appropriate practical activity sheets for parents to support their child's specific areas of weakness so the parents and children could benefit from the developmental screening.

Keywords: ages and stages questionnaire, pediatric cardiology, neurodevelopmental outcomes

General Cardiology

OP-089

Lung function in fontan patients over a ten-year period; is the fontan circulation impairing lung development?

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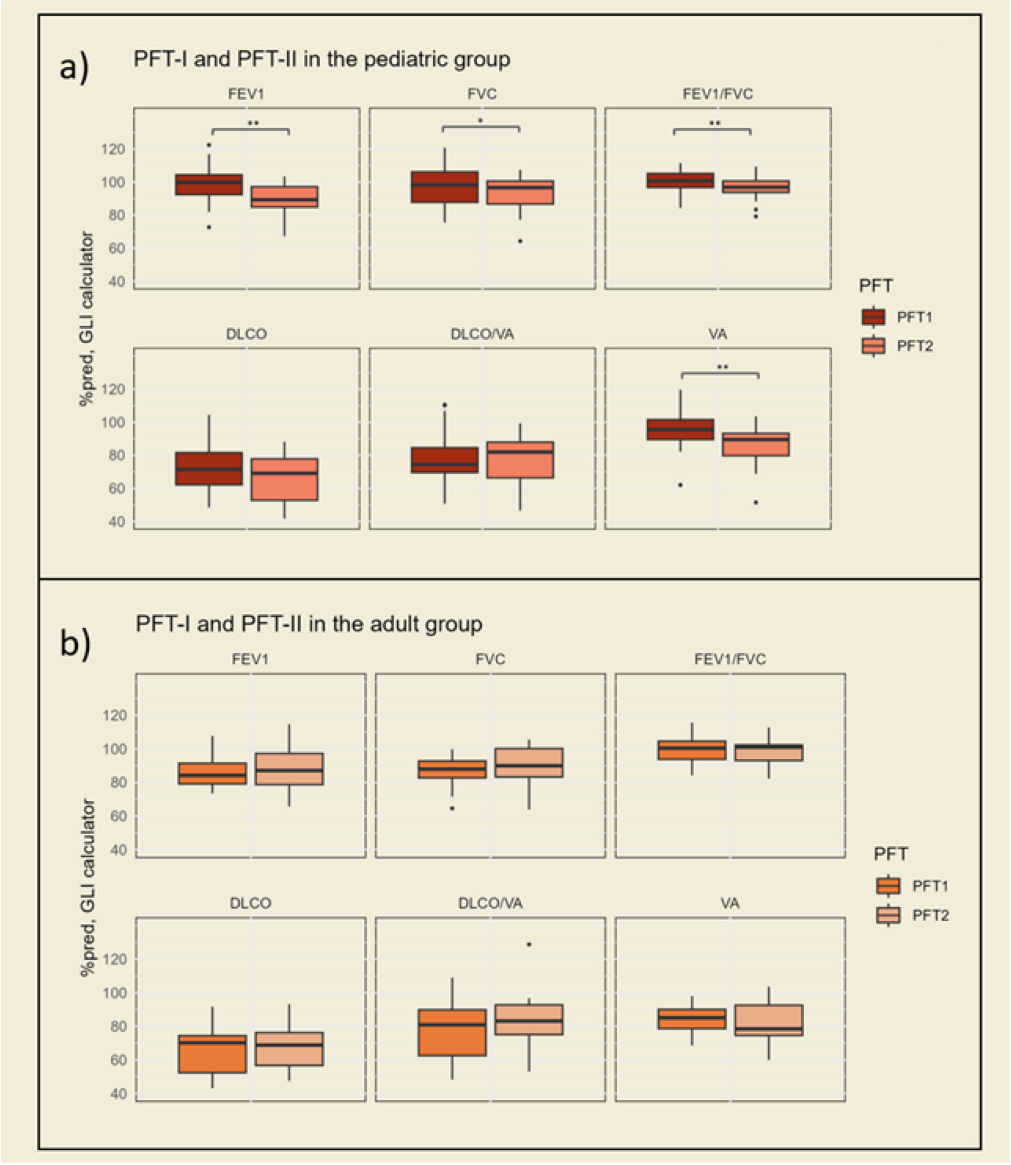
Background and Aim: Few studies have investigated how the Fontan circulation affects lung function and no studies have investigated the evolution of lung function over time in these patients. We aimed to describe the evolution of lung function in Fontan patients over a ten-year period.

Figure 1



*Results from spirometry, lung volume, and DLCO10S in PFT-I and PFT-II, presented as percent predicted (%pred). Patients are categorized into a pediatric group (under 18 years at PFT-I), results presented in, and an adult group (18 years or older at PFT-I). All %pred values are calculated with the GLI calculator. Wilcoxon signed-rank test for paired data was conducted to check for significant differences from PFT-I to PFT-II within each group (** p <0.001,* p < 0.05) a) Results from PFT-I and PFT-II in the pediatric group. b) Results from PFT-I and PFT-II in the adult group. Abbreviations: FEV1 (forced expiratory volume in 1 second; FVC (forced vital capacity); DLCO (diffusing Capacity for Carbon Monoxide); DLCO/VA (coefficient of the lung for carbon monoxide); VA (alveolar volume)*

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Method: Pulmonary function tests (PFT), including spirometry and diffusion capacity for Carbon Monoxide (DLCO) and Nitric Oxide (DLNO), were conducted in a Danish Fontan cohort in 2011 (PFT-I). In 2021, re-investigations were performed (PFT-II). We investigated changes in percent predicted (%pred) lung function from PFT-I to PFT-II. Patients were categorized into a pediatric group (age under 18 at PFT-I) and an adult group (age 18 or older at PFT-I). In addition, cross-sectional PFT-I results from the pediatric group were compared with a Danish reference material from healthy children.
Results: Out of the 81 patients completing PFT-I, 48 completed PFT-II. In the pediatric group, (32 patients), there were significant declines in %pred forced expiratory volume in 1 second (FEV1) (99.7 (92.4, 104.4) - 89.3 (84.9,97.2), p<0,001), forced vital capacity (FVC) (98.3 (87.8,106.1) - 96.7 (86.7,100.6), p=0.008), and alveolar volume (VA) (95.5 (89.5,101.6) - 89.5 (79.7,93.2), p<0.001), Figure 1a. The corresponding measurements remained stable in the adult group, Figure 1b. However, the median %pred DLNO significantly declined in the adult group (58.4 (53.3, 63.5) – 53.7 (44.1, 57.3), p=0.005). Longitudinal DLNO results were not analyzed in the pediatric group due to the lack of suitable reference material. Interaction analyses using linear regression found that DLCO10s, DLNO, and the membrane diffusing capacity of DLNO (Dm), increased significantly less with age in the pediatric Fontan group compared to healthy children (Figure 2). FEV1, FVC, and VA showed a similar trend, although not significant.
Conclusions: Over a ten-year period, several lung function parameters declined significantly in the younger Fontan patients, suggesting possible impairments in lung development during growth. The decline in %pred DLNO in the adult patient group indicates deterioration of the membrane component of diffusion capacity, implying that the Fontan circulation might negatively affect the alveolar membrane over time.

Keywords: Fontan circulation, Lung function, Pulmonary function test, Lung development, Complications

OP-090

Utilizing machine learning and ensemble techniques to construct a prediction model for kawasaki disease identification in febrile children

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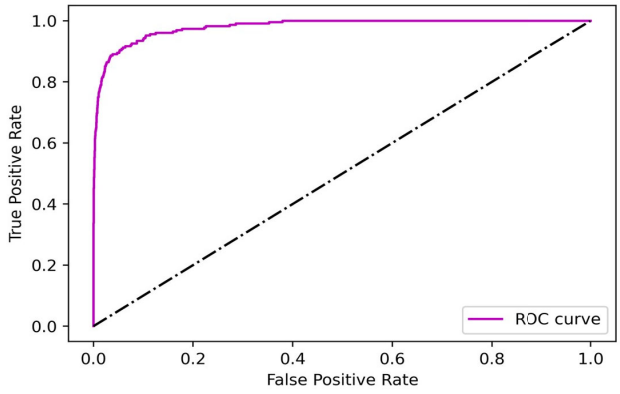
Background and Aim: The early detection of Kawasaki disease (KD) in children is essential for ensuring timely treatment and averting the potential development of acquired heart disease. However, diagnosing KD is a complex task primarily dependent on subjective diagnostic criteria. Our aim is to create a machine learning prediction model that utilizes objective parameters to distinguish children with KD from those with fever.
Method: We conducted a retrospective study involving 74,641 febrile children under the age of 5, who presented at four pediatric emergency departments in Taiwan between January 2010 to December 2019. These children were categorized as either having Kawasaki disease (KD) or serving as febrile controls (FC). We gathered demographic information and objective laboratory data from electronic medical records, including complete blood cell counts with differential count, urinalysis, and biochemistry, to consider them as potential predictive factors. We employed boosting and stacking techniques in the development of our prediction model to effectively address the imbalance in the dataset. For evaluating the model's performance, we utilized metrics like the confusion matrix and likelihood ratio.
Results: This study encompassed a total of 1,142 cases of Kawasaki disease (KD) and 73,499 febrile controls (FC). The KD group exhibited a male predominance (60.2% vs. 56.4%, p=0.011) and a younger age (1.08 ± 0.84 vs. 1.55 ± 1.36, p<0.001) compared to the FC group. The top five important features in predicting KD in the model were pyuria, white blood cell counts in urine, alanine aminotransferase level, C-reactive protein level, and eosinophil percentage. The performance of the prediction model on the testing set exhibited excellent results with a sensitivity of 95.2%, specificity of 96.7%, a positive predictive value of 30.7%, a negative predictive value of 99.9%, a positive likelihood ratio of 28.55, and a negative likelihood ratio of 0.05. These metrics collectively highlight the model's exceptional performance.
Conclusions: This cohort study indicates that the results of objective laboratory tests have the potential to serve as predictors of KD. Moreover, these findings imply that machine learning techniques can assist physicians in effectively distinguishing between children with KD and other febrile children in pediatric emergency departments.

Keywords: Kawasaki disease, machine learning, artificial intelligence, fever, children, pediatric emergency department

The ROC curve of the prediction model



The area under the receiver operating characteristic curve (AUC-ROC) in our prediction model is 0.988.

Interventional Cardiology

OP-092

Erosion and embolization after trans-catheter atrial septal defect closure: An european survey

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Background and Aim: Patients with device atrial septal defect (ASD) closure have an excellent prognosis with low mortality. Delayed

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erosion of the aortic or atrial wall and embolizations are infrequent but worrisome complications with a yet poorly understood epidemiology. The aim of this European survey is to find the real incidence of cardiac erosion and device embolization in a cohort of pediatric and adult patients undergoing ASD closure.
Method: We retrospectively analyzed data of ASD closure performed in 28 European centers from 2011 to 2021. We collected data about total numbers of treated patients, of device characteristics with detailed informations about erosion and embolization patients.
Results: During the study period transcatheter ASD closure was performed in 10.039 patients, 5183 were pediatric patients (66%). Sizing balloon was performed in every case in 13 centers, in 9 centers only in specific situations and was never performed in 1 center. Most common used devices were Amplatzer (46%), Occlutech (29%), Gore septal occluder (8%) and others (17%).
Cardiac erosion was registered in 12 patients (0.12%), 58% in pediatric patients. In all patients an Amplatzer device was implanted except one treated with a Cardia device. Median device dimension was 20 mm (IQR 17-23 mm). Median time of erosion from ASD closure was 5 days (IQR 1.5-93.5 days). Absence of the aortic rim was present in 58% of patients. All patients were surgically treated with good results and only one death was reported.
Device embolization was described in 71 patients (0.7%), 57% in pediatric patients. Embolized devices were Amplazter (44%), Occlutech (43%) and Memopart (7%). Site of embolization was the left heart in 68% of patients and right heart in 32%. Median time of embolization from ASD closure was 8 hours (IQR 0.8-22 hours) with one after 2 years. Absence of the anterior rim was described in the majority of patients (65%). Devices were retrieved percutaneously in 75% of patients, others underwent surgical removal. No deaths were described.
Conclusions: Percutaneous ASD closure is a safe procedure. In our survey, risk of erosion is 0.12 % and is more frequent in patients implanted with an Amplatzer device. Embolization incidence is 0.7% and is associated to margins deficiency. Mortality associated to these complications is low.

Keywords: ASD, erosion, embolization

Interventional Cardiology

OP-093
Early and midterm experience with novel konar TM multifunctional occluder for closure of ventricular septal defects in children less than 3 years

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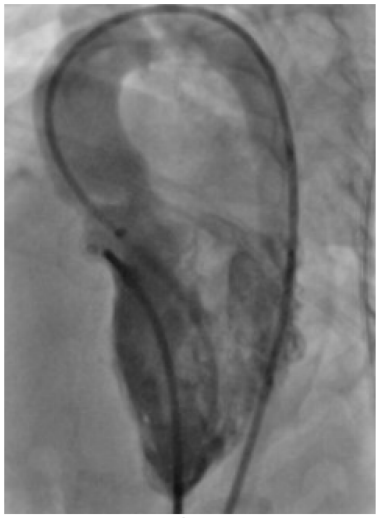
Background and Aim: One of commonest congenital heart defects is Ventricular septal defect
[VSD] in children. Incidence of VSD is around 1.5 to 3.5 per 1000 live births. Percutaneous closure of ventricular septal defect (VSD) in children is an established procedure. Various devices are being used however, every occluder has got its own advantages and disadvantages.KONAR multifunctional occluder (MFO) is a novel medium profile occluder developed to close VSD's. This study is to assess the feasibility and efficacy of the MFO in children with VSD with midterm follow up for any complications.
Method: We present here a total of 68 children less than 3 years of age, who underwent closure of VSD's from December 2019 to March 2023 using MFO. VSDs were closed under fluoroscopy and transthoracic echocardiography (TTE) guidance. All patients were followed - up at 1,3,6, 12 and 18 months with clinical, ECG and echocardiography checks.
Results: VSD closure was successful in all 68 cases (100%). The mean age of patient was 16 ± SD (5.7- 26.3) months and mean weight was 8.8 kgs ± SD (7.1- 10.5). Mean VSD diameter was 5.4 mm± SD (4 - 6.8). Forty two (61.7%) children had muscular VSD (mVSD), 24 (35.4%) perimembranous VSD (pmVSD) and 2 patients (2.9%) had subpulmonic VSD's. Only one child (1.5%) developed first degree heart block with no hemodynamic compromise and remained so on follow-up. Three patients (4.4%) had mild tricuspid regurgitation showed no progression on follow-up. Both antegrade (27%) and retrograde (73%) methods were used to deploy device for VSD closure. Small residual shunts < 2mm was seen in 7patients (10.2%) and significant shunt > 2mm was seen in only 3 (4.4%) with larger devices. Small shunts disappeared within 48 hrs, however significant shunts persisted for 2-4 weeks but with no hemolysis. All patients were followed for mean of 16 months ± SD (12.8 - 19.2months). The mean fluoroscopy time for VSD closure was 10.45mins ± SD (8.4 – 12mins).
Conclusions: MFO device can be safely and effectively used to close VSD's in smaller children. No serious adverse events were noticed in midterm follow-up.

Keywords: Ventricular septal defect, transcatheter closure, KONAR-MFO

Post MFO device closure of VSD



Angiogram in LAO view showing MFO device occlusion of VSD

Congenital Heart Surgery

OP-094
Association of pulmonary hemodynamics at fontan with long-term outcomes after the fontan palliation

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Background and Aim: To determine the association of pulmonary hemodynamics at time of Fontan with in-hospital and long-term outcomes after the Fontan operation.

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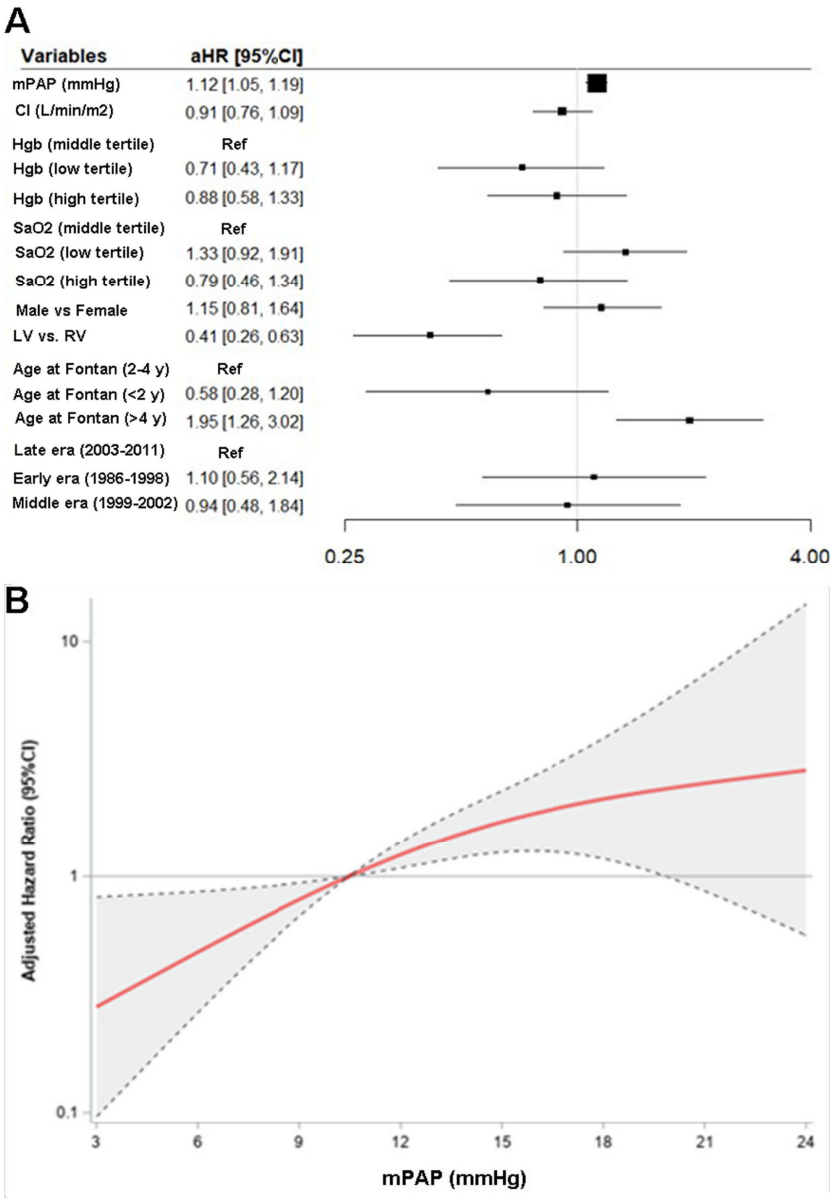
Method: This is a cohort study of patients undergoing Fontan in the Pediatric Cardiac Care Consortium, a US-based registry of congenital heart surgeries. Post-discharge deaths were captured through 2022 by linkage with the National Death Index. Regression analysis and Cox proportional hazard models were used to assess the association between pulmonary hemodynamics [mPAP=mean pulmonary arterial pressure, PVR=pulmonary vascular resistance, TPG=transpulmonary gradient] and in-hospital Fontan failure (death or takedown) and post-discharge risk of death respectively. *Results:* Between 1986 and 2011, 1461 patients were enrolled in the PCCC who underwent pre-Fontan cardiac catheterization. A total of 95 patients had in-hospital Fontan failure (deaths: 78, Fontan take-down 17) and 1366 patients were discharged alive with Fontan physiology. Over 21.2 years of median follow-up (IQR: 18.4-24.5) 184 deaths occurred. On multivariable analysis for in-hospital Fontan failure, mPAP was found to have borderline significance (aOR 1.09 for each mmHg increase in mPAP; 95%CI: 0.99-1.20, p=0.08); no other variable reached statistical significance. Risk for post-discharge death increased by 10% at any time during the follow-up period for each mmHg increase in the mPAP (aHR 1.10, 95%CI 1.0-1.17, p<0.01) (Figure A). mPAP had an almost linear relationship with the risk of post discharge death

Figure: Association of mPAP at Fontan with long-term survival



(A) Adjusted long-term hazard of death (aHR) by mean pulmonary arterial pressure (mPAP) increase by 1 mmHg at time of Fontan after adjustment for cardiac index (CI), hemoglobin (Hgb), systemic arterial oxygen saturation (SaO2), Fontan age and surgical era with CI used as continuous variable, Hgb and SaO2 as categorical variables by tertiles (middle group being the reference in both cases) and more recent era (2003-2011) used as reference. (B) Adjusted dose response curve of the association between the mPAP at Fontan and longitudinal hazard of death by using as reference the middle mPAP (11 mmHg)

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along the range of values obtained at time of pre-Fontan evaluation (Figure B). mPAP also had a strong interaction with the presence of systemic left ventricle (aHR 1.15, 95%CI:1.06-1.26, p<0.02). TPG and PVR did not reach significant association neither with in-hospital nor with post-discharge risk of death. Systemic RV and age >4 years at Fontan were both independently associated with higher risk of post-discharge mortality.
Conclusions: Our data demonstrate that low mPAP and age of 2-4 years at time of Fontan are strong predictors of long-term success in the Fontan pathway. The findings, also, suggest that maintaining low mPAP may prolong the longevity of Fontan physiology for many years after the procedure, especially for patients with systemic left ventricle.

Keywords: single ventricle, Fontan operation, pulmonary arterial pressure, outcomes

Congenital Heart Surgery

OP-095
Adverse neurological events following total cavopulmonary connection surgery

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Background and Aim: Patients undergoing total cavopulmonary connection (TCPC) surgery are at risk of suffering adverse neurological events such as stroke, intracranial haemorrhage, seizures, hemiparesis, and hypoxic ischaemic encephalopathy. Most of the current literature is approximately two decades old, contains small sample sizes, and solely focuses on stroke. We investigated a variety of adverse neurological events, in order to provide a more comprehensive understanding of neurological outcomes for these patients. Our aims included assessment of incidence rates, time-to-event, clinicopathological characteristics, and risk factors for adverse neurological events following TCPC surgery.
Method: Paediatric patients that underwent TCPC at Birmingham Children's Hospital between 2012-2022 were included in a single-centre retrospective review. Demographic, surgical, and haemodynamic variables were assessed. Kaplan-Meier and Cox proportional hazard methods were used to analyse timing and associated risk factors.
Results: TCPC was performed in 406 patients. Median follow-up time was 3.18 years (IQR, 0.22-7.14). Nineteen (4.7%) suffered adverse neurological events. Median time-to-event was 35 days (IQR, 6-340.5). Thirteen (68%) events occurred within the first 2 post-operative months. Seizures were the most common clinical presentation (N=6). Stroke was the most common radiological diagnosis (N=8). Following an event, 5 patients made a complete recovery; 10 experienced residual neurological deficits, and 4 died. Among the 8 stroke patients, 50% underwent ligation or transection of the main pulmonary artery, compared to 17% among the study cohort. Each additional minute of cardiopulmonary bypass was significantly associated with a 2% increase in the risk of a post-operative event (HR, 1.02; 95% CI, 1.00-1.03; p=0.01). While not statistically significant, the occurrence of adverse neurological events prior to TCPC increased post-operative event risk 2.47-fold (HR, 2.47; 95% CI, 0.81-7.47; p=0.11). Each additional year that passed before TCPC was protective (HR, 0.78, 95% CI, 0.54-1.14; p=0.20).
Conclusions: We present an up-to-date analysis, using a comparatively large sample size, of adverse neurological events following TCPC surgery. Most events occurred within the first 2-month post-operative period; ligation or transection of the main pulmonary artery may increase stroke risk, and increasing cardiopulmonary bypass time was found to be a statistically significant risk factor. Expansion into adult congenital heart disease databases are under discussion.

Keywords: TCPC, stroke, intracranial haemorrhage, seizure, hemiparesis, hypoxic ischaemic encephalopathy

Table 1. Table expressing time-to-event, clinical presentation, radiological findings, and outcomes for the nineteen patients who suffered neurological complications following TCPC surgery.

Patient No.	Time-to-event	Clinical Presentation	Radiological finding	Outcome
Stroke patients				
1	4 days	Post-op Fontan failure	CT: Bilateral infarcts	Death
2	7 days	Seizure & L.Hemiparesis	MRI: Bilateral infarctions	Ongoing hemiparesis
3	12 days	L.Hemiparesis & L. facial droop	CT: R. MCA infarct	Improving hemiparesis
4	25 days	Post-op Fontan failure	CT: L. occipital infarct; R. cerebral haemorrhage	Death
5	38 days	L.Hemiparesis		Improving hemiparesis
6	41 days	L. Hemiparesis	CT: R.MCA infarct	Improving hemiparesis
7	8 months	Fixed dilated L. pupil	CT: L. MCA haemorrhage	Death
8	4 years	Confusion & L. 6th, 7th CN palsy	MRI: Brainstem stroke	Ataxia
Non-stroke patients				
9	0 days	Seizures	CT: Normal	Commenced Keppra → Complete recovery
10	1 day	Abnormal extensor posturing		Complete recovery
11	4 days	Fixed dilated pupils	CT: HIE & uncal herniation of temporal lobe	Death
12	5 days	Absent L. movement and reflexes	CT: R. Posterior subdural bleed	Neurological referral → Improving movement
13	17 days	Encephalopathic changes on EEG	MRI: Normal	Improving hemiparesis
14	1 month	Seizure	CT: Subdural haematoma MRI: White matter atrophy	Ongoing seizures
15	2 months	L.Hemiparesis	CT: HIE R. frontal lobe	Neurophysiotherapy → Improving hemiparesis
16	1 year	R. Hemiplegia	MRI: White matter injury	Ongoing hemiplegia
17	6 years	Seizure		Complete recovery
18	9 years	Seizure		Complete recovery
19	9 years	Seizure	CT: Normal	Complete recovery

Patients are separated into a stroke and non-stroke group. Blank cells in the fourth column represent patients in which radiological imaging was not requested. L: left, R: right, CN: cranial nerve, EEG: electroencephalogram, CT: computed tomography, MRI: magnetic resonance imaging, MCA, middle cerebral artery; HIE, hypoxic ischaemic encephalopathy.

General Cardiology

OP-096
Pulmonary artery growth in fontan: What is the most effective strategy

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Background and Aim: The best strategy to achieve optimal growth of pulmonary artery branches (PAs) in FUH between partial cavopulmonary connection (PCPC) and total cavopulmonary connection (TCPC) is still controversial, especially the preservation of anterograde pulmonary blood flow (APBF). To model the growth of PA in FUH between PCPC and TCPC and to assess whether APBF promotes this growth. Then to determine whether the

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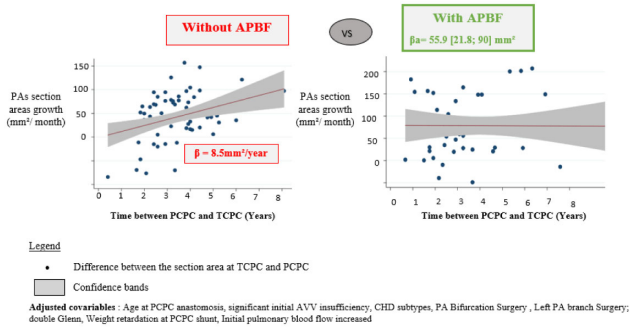
maintenance of an APBF is associated with higher morbidity and mortality.
Method: We retrospectively included all patients with FUH who underwent cardiac catheterization before PCPC and TCPC between 2004 and 2021. A linear regression model was used to model PA growth. We compared mortality and morbidity outcomes between the APBF group and no APBF group using the Kaplan-Meier method
Results: 118 children with FUH with a median follow-up of 8.8 years were included, of whom 49 (41.5%) had maintained APBF. PA branch growth can be considered continuous and linear over time (fractional polynomials (p=0.2)), estimated at β= 8.5 [0.7-16.2] mm^2/year. In multivariate analysis, maintaining an APBF was the only factor associated with increased PA branch growth (β= 55.9 [21.8; 90]mm^2 (p=0.001)), regardless of TCPC timing (Figure). Before PCPC, there was no difference in mean pulmonary artery pressure (mPAP) between groups with and without APBF. Between PCPC and TCPC, there was no significant increase in mPAP (13.2(12-16) vs. 14.1(12.4-15.8), p=0.3) or transpulmonary pressure gradient (7.2(5.2-9.3) vs. 6.9(4.6-9.1), p=0.6) in the APBF group. There was no difference in survival at 6 years after TCPC between the group with APBF (87.6 CI95% (65.6% - 95.9%)) and the group without APBF (82.3 CI95% (67.8 - 90.6)). No difference was also found when comparing morbidity characteristics between the two groups, such as length of hospital stay after TCPC (p= 0.7), chylothorax (p= 0.81), hemodynamic contraindications to fenestration closure (p= 0.9), failing Fontan rate (p=0.38).
Conclusions: PAs growth between PCPC and TCPC is significant and can be considered linear over time. Delaying TCPC as long as possible allows full growth potential to be achieved. By maintaining APBF, growth potential can be achieved much more quickly and without additional risk of morbidity or mortality.

Keywords: FUH, Fontan, pulmonary artery growth, mortality

Graph of PAs growth versus time between PCPC and TCPC



General Cardiology

OP-097
Early cardiovascular events in pediatric cancer survivors - insights from a retrospective observational studY

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Background and Aim: The increasing incidence of cardiotoxicity, particularly arising from conventional chemotherapy and radiation, presents challenges for childhood cancer survivors. Limited evidence exists on cardiotoxic damage in acute, subacute phases, and one-year post-chemotherapy. Our study aim is to describe early cardiotoxicity expression and identify associated risk factors.
Method: Data on demographics, clinical features, neoplastic diagnosis, therapeutic regimens (chemotherapy, targeted therapy, radiotherapy, and hematopoietic stem cell transplantation, HSCT), and cardiac evaluations were collected from childhood cancer survivors aged 0-18 yrs admitted at IRCCS University hospital of Bologna from January 2017 to September 2023. Statistical analyses included univariate logistic regression.
Results: 369 subjects were enrolled (51.49% males, 89.16% Caucasians). Hematologic neoplasms accounted for 53.93%, and solid organ tumors for 46.07%. During antineoplastic therapy 22% of patients presented at least one of the following: LV systolic (26%) and diastolic (4%) dysfunction, dilated cardiomyopathy (2%), thrombotic events (6%), pericardial diseases (4%), valvulopathies (2%), and systemic hypertension (43%). 62% of events occurred within the first 6 months of antineoplastic therapy (AT) after the diagnosis (mean time 310 days (IQR 29 – 331). The need for suspension and/or modification of AT and/or intensive care admission occurred in 23% of patients. The cardiovascular (CV) events persisted in 15% at the last cardiological evaluation (mean time 2.3 yrs, IQR 1.5– 3).
Cardioactive medications were administered in 71% of cases during the acute stage of CV event, and continued in 27% of them at the last clinical assessment. A trend of weight gain was observed during the follow-up after the end of antineoplastic therapy, with the percentage of overweight patients increasing from 10% at the time of tumor diagnosis to 36% at the last clinical evaluation. Risk factors for cardiotoxicity were age at diagnosis <8 years (p: 0.032), the exposure to a cumulative dose of anthracycline exceeding 240 mg/m2 (p<0.001), a diagnosis of hematological tumor (p<0.001), the administration of cyclophosphamide (p<0.001), 6-mercaptopurine (p<0.001) thioguanine (p<0.001) cytarabine (p<0.001), and methotrexate (p<0.013), and the HSCT (p: 0.018).
Conclusions: Our study emphasized the need for comprehensive pre-treatment risk assessments, personalized management based on clinical risk, and interventions to reduce modifiable CV risks, such as weight control.

Keywords: cardio-oncology, cardiovascular risk factors, childhood cancer survivors, left ventricular dysfunction, systemic hypertension

Adult Congenital Heart Disease

PP-003
The late complications of undiagnosed ebstein anomaly

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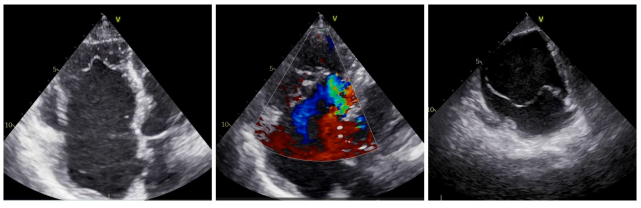
Background and Aim: Ebstein anomaly is a rare congenital disorder that involves dysplasia of the tricuspid valve with apical displacement of the septal or anterior leaflet, tethering of the leaflet tissue, right atrium enlargement, atrialized right ventricle (RV) and reduced RV systolic function. Symptoms and signs can occur at any age with a varied severity according to the degree of anatomic abnormality. Late presentation is usually associated with milder anatomic anomalies.
Method: An 83-year-old female was admitted to the Emergency Department with mild dyspnea (SatO2 80%, 22 breaths/min) and lower limbs edema. Clinical examination revealed a holosystolic murmur in the 4th intercostal space in the parasternal region and distended jugular veins.
Results: Laboratory findings included a sinus rhythm and a right bundle branch block on the electrocardiogram (ECG), an increased cardiothoracic ratio and signs of congestion on the chest X-ray and an elevated Brain Natriuretic Peptide value. The transthoracic echocardiogram (TTE) showed a preserved systolic function of the right and left ventricle (LV), increased dimensions of the right and left atrium, mild mitral regurgitation, moderate tricuspid regurgitation (TR), elevated LV filling pressures and increased dimensions of the inferior vena cava. The most important findings were the apical displacement of the septal leaflet of the tricuspid valve and the variable tethering of the leaflet tissue to the RV myocardium, as confirmed by transesophageal echocardiogram.ESC guidelines recommend surgical repair in patients with severe tricuspid regurgitation and symptoms or objective deterioration of exercise capacity (Class I, Level C) and in patients with progressive right heart dilatation or reduction of RV systolic function, regardless of symptoms (Class IIa, Level C). Our patient had no relevant symptoms in the past. In addition, the TTE showed moderate TR and no significant decline of the RV function. Therefore, conservative treatment was decided.
Conclusions: Physicians should be alerted about unusual causes of acute dyspnea in the elderly, such as congenital diseases which may occasionally remain undiagnosed and present with symptoms at advanced age.

Keywords: Ebstein Anomaly, Congenital Disease

Ebstein Anomaly in elderly people



PP-004
A case of infective in a patient with tetralogy of fallot patients that highlights the importance of multimodality imaging

Alkistis Eleni Kalesi, Evangelos Tatsis, Alexandros Pappas, Alexandros Varsos, Maria Mouratidou, Christina Floropoulou, Antigoni Sakellaropoulou, Ioannis Papadopoulos, Dimitrios Ikonomidis, Constantinos Papadopoulos and Ioannis Tsiafoutis
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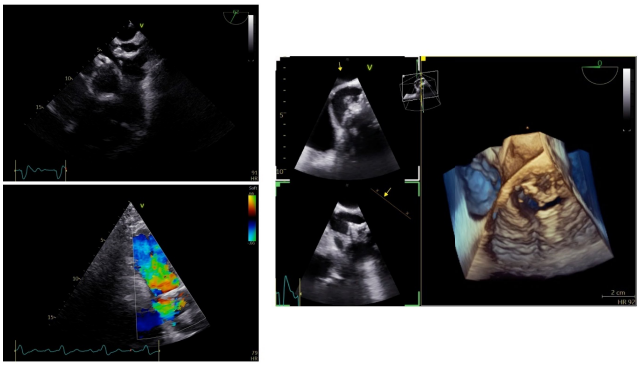
Background and Aim: The lesions' complexity and the presence of prosthetic material are known risk factors for infective endocarditis in patients with congenital heart diseases. Tetralogy of Fallot (ToF) stands out as one of the most complex cyanotic congenital heart diseases, often requiring a prosthetic valve implantation. Patients with ToF are susceptible to infective endocarditis (IE), therefore high clinical suspicion is of great importance for timely recognition and treatment of IE in those patients.
Method: We present the case of a 29-year-old male patient with a history of late surgical treatment of ToF and a pulmonary valve replacement three years ago. The patient presented at the emergency department due to syncope. At the ED the patient was hemodynamically stable, febrile and the ECG revealed sinus tachycardia of 120 bpm and right bundle branch block. Arterial blood gas analysis was also performed showing respiratory alkalosis, with a pH of 7.51, a pO2 of 55 mmHg, and a pCO2 of 30 mmHg.
Results: The patient underwent a CT pulmonary angiography that showed the presence of filling defects in lobar branches bilaterally. Considering the above findings and the patient's history, three sets of blood cultures were obtained and coagulase-negative staphylococcus was isolated in all of them, raising high clinical suspicion of infective endocarditis. The transthoracic echocardiogram performed didn't show any vegetations. The transesophageal echocardiogram however revealed a rocking motion of the sewing ring and the presence of multiple vegetations in the prosthetic pulmonary valve surface. The above findings were also confirmed by a positive 18F-FDG PET/CT scan.
Conclusions: Infective endocarditis is a common complication in patients with congenital heart diseases. Setting the diagnosis can be challenging, especially in patients presenting with atypical manifestations. Given the high mortality of this disease and the importance of timely diagnosis in successful treatment, high clinical suspicion along with the use of all available imaging modalities is essential.

Keywords: Tetralogy of Fallot, Infective Endocarditis, rare presentation, multimodality imaging, pulmonary valve prosthesis

endocarditis-toe



PP-005
A historical view of a nurse-driven outpatient clinic

Anna Wieland[1], Marie Haack[2] and Hannah Hanley[3]
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Background and Aim: The nurse-driven Adult congenital heart disease (ACHD) outpatient clinic was established at Rigshospitalet in Copenhagen almost 20 years ago. In 2012 the number of patients with ACHD in Denmark exceeded the number of children with congenital heart disease. With an increasing number of patients

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due to longevity, there was a need to optimize and organize our outpatient clinic. The team consists of four nurses with years of experience in cardiology and congenital heart disease. Our ACHD outpatient clinic aims to provide patients with easy access to highly specialized care, counseling, and treatment. Furthermore, it facilitates the transition between pediatric and adult care. Also, educating and supporting patients with ACHD and their families and ensuring a coherent patient pathway throughout Denmark, both regionally and nationally. Therefore, the aim was to optimize the outpatient clinic, to provide care for the increasing number of patients.
Method: Workshops were held to discuss and identify areas for improvement and how to structure the workload at the ACHD outpatient clinic. Themes such as transition, pregnancy counseling, surgery coordination, arrhythmia, anesthesia planning, heart failure, and advanced care planning were identified. We conducted a literature search on each theme and discussed the findings with the team. To implement our knowledge, we appointed coordinators with responsibility for each theme.
Results: We have facilitated an improved structure for the ACHD outpatient clinic, and the results are: A nurse has been appointed as transition coordinator to coordinate the transfer from pediatric to adult care and to schedule and conduct youth consultations. A nurse-led clinic for patients with arrhythmias has been established and we facilitate the MDT conference every fortnight. A pathway for patients undergoing anesthesia has been established. A nurse-led for patients with heart failure is currently in the process of being established.
Conclusions: The new structure of the ACHD outpatient clinic contributes to an increased focus on patients' special needs in the growing ACHD population. As a result of our improvements, we can see a 50% increase in transition consultations from 2020 to 2022. From 2019 to 2022 the number of phone calls more than doubled from 1700 to 3667 calls a year.

Keywords: optimize the outpatient clinic, structure the workload at the ACHD outpatient clinic, focus on patients' special needs in the growing ACHD population

PP-007
Unusual tubular communication between the right sinus of valsalva and right atrium: A rare case of aorto-right atrial tunnel

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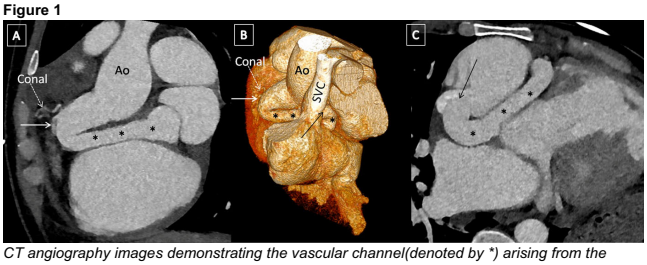
Background and Aim: Aorto-right atrial tunnels are rare extra-cardiac vascular lesions that often pose a diagnostic dilemma. MDCT in addition to being a non-invasive modality excellently delineates their anatomical features including location, course, caliber, drainage site, relation to coronaries, status of the aortic root and associated abnormalities. Moreover, it reliably differentiates them from more commonly encountered mimics like sinus of Valsalva aneurysm and coronary cameral fistula. We, hereby, describe one such case of aorto-right atrial tunnel in a middle-aged male patient who underwent successful surgical repair subsequently.
Method: The entity was diagnosed using MDCT imaging features (Figure 1).
Results: Intraoperatively, a large opening of the size of about 15 mm x 15 mm was seen in right coronary sinus adjacent to the ostium of right coronary artery. Right atrium was opened. Fistulous tract was seen opening in right atrium posteriorly near right atrium-superior vena cava junction. The tricuspid valve morphology was normal with minimal leak on saline jet test. The aortic end of fistulous opening was closed through aorta using autologous pericardium.
Conclusions: Aorto-right atrial tunnel is a rarely encountered entity and refers to an abnormal extra-cardiac vascular channel between one of the aortic sinuses and the right atrium, resulting in a left-to-right shunt at this level.Clinical presentation varies from an asymptomatic patient with an incidentally detected continuous precordial murmur to symptoms of frank congestive heart failure in decompensated cases.The tunnel if left untreated, poses certain complications due to continued patency including wall calcification, aneurysmal expansion of tunnel, congestive heart failure due to volume overload and increased risk of infective endocarditis.Treatment modalities vary and depend upon the location, size, course and drainage point with special emphasis on status of the coronary ostia. These include catheter based endovascular intervention, ligation of the aortic end of the tunnel or surgical repair under cardio-pulmonary bypass.

Keywords: Aorto-right atrial tunnel, MDCT

Figure 1



*CT angiography images demonstrating the vascular channel(denoted by *) arising from the bulbous right aortic sinus, coursing superior to left atrium and draining into the right atrium near cavoatrial junction(black arrow). Right coronary artery(white arrows) and conal branch(dashed white arrows). SVC-superior vena cava, Ao-Ascending aorta.*

PP-009
The proteomic signature of a univentricular circulation – insights from the plasma

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Background and Aim: The Fontan circulation is accompanied by substantial hemodynamic and metabolic changes. Of these, hypoxia in infancy and early childhood, a life-long reduction in cardiac output, a non-pulsatile and passive flow through the pulmonary circulation, and severe venous congestion are some of the most noteworthy. We hypothesized that the proteomic signature of this altered circulation differed from that of healthy age- and gender-matched controls.
Method: As a part of the national DANFontan trial (H-20028226), 148 adult individuals with a Fontan circulation were invited to participate. Criteria for proteomic analysis were age between 18 and 32 years. Systemic right ventricle or systemic left ventricle and tricuspid atresia or double inlet left ventricle. Age, gender, and

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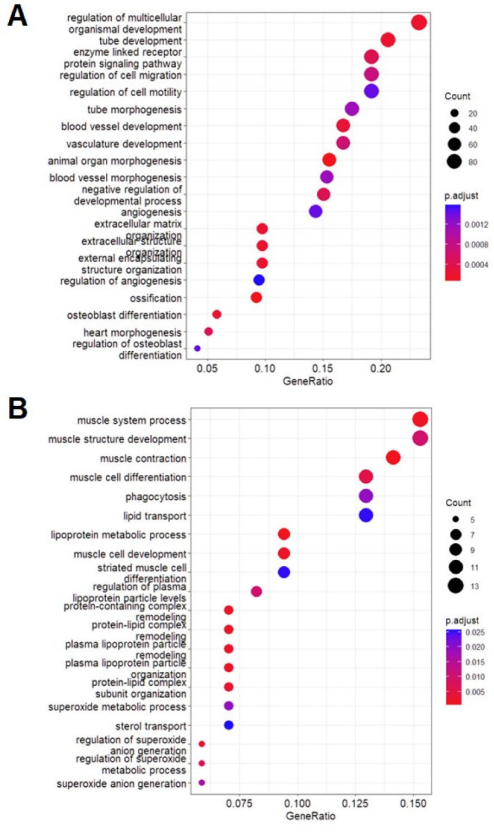
weight-matched healthy controls were matched 1:2 with cases. Relative measurements of circulating proteins were performed using the Olink® Explorer 3072 platform. To identify underlying biological schemes in the lists of significantly increased and decreased proteins, we assigned biological functions to all differentiated proteins based on the Gene Ontology Biological process database and next investigated if any categories occurred more often than expected by chance.
Results: Plasma samples from 58 individuals with a Fontan circulation and 29 controls were analyzed. Following quality control, relative quantification of 2605 unique proteins was obtained. Assigning the proteins to biological schemes and adjusting for age, gender, and BMI revealed several functions with an altered protein expression. Amongst the ones found to be upregulated, there was an overweight involved in angiogenesis, bone homeostasis, and organ morphogenesis. Proteins found to be downregulated were more often related to muscle function, lipid metabolism, and regulation of superoxides.
Conclusions: The proteomic signature of the Fontan circulation is markedly altered from that of healthy, gender- and age-matched controls. Aligning with clinical observations, our findings may help explain the increased angiogenesis, altered bone composition, reduced muscle mass, and dyslipidemia caused by an adapted metabolism. We hypothesize that universal adaptations to hypoxia play a role in many of the alterations. Although unable to show causality, our detailed proteomic findings may help bridge the gap and guide a better understanding of the Fontan circulation.

Keywords: Congenital heart defects, ACHD, Fontan, Proteomics; Angiogenesis

Figure 1



Overview of biological functions for dysregulated proteins. Overall the relative levels of 412 out of 2605 proteins were increased and 85 out of 2605 were decreased. A) Displays the biological functions most affected by increased levels of proteins (ie. 69 out of 412 proteins displaying increased levels were related to "blood vessel development" resulting in a GeneRatio of 0.17). Overall, functions related to angiogenesis, bone homeostasis, and organ morphogenesis tended to have the greatest proportion of the proteins displaying an increased level. B) Displays the biological functions covering the proteins that were decreased in individuals with a Fontan circulation. Overall, the greatest proportion of decreased proteins was found in categories related to muscle function, lipid metabolism, and regulation of superoxides.

PP-010

Sacubitril/valsartan in adult congenital heart disease patients with heart failure

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Background and Aim: Sacubitril/Valsartan showed an improvement in life expectancy in adults with heart failure (HF). Evidences about the use of these drugs in ACHD patients with HF is limited. The aim of this study is to evaluate short-term effects (6 months) and tolerability of Sacubitril/Valsartan in ACHD patients.
Method: All consecutive ACHD patients treated with Sacubitril/Valsartan for HF from February 2022 to October 2023 at "ACHD program" were included in this prospective study. According to the ventricular physiology, patients were categorized in three groups: 1) systemic left ventricle (SLV), 2) systemic right ventricle (SRV), 3) single ventricle (SV). Primary outcome was the change in New York Heart Association (NYHA) functional class. Secondary outcomes were reduction in BNP values and increase in VO2 max in cardiopulmonary exercise test (CPET). Exploratory outcome was drug tolerability.
Results: Twenty-four patients were treated with Sacubitril/Valsartan. Mean age at the treatment was 36 (+/-8.7 years old). According to the ventricular physiology, 4 patients (17%) had SLV; 9 patients had SRV (37%) and 11 patients had SV (46%). Patients had a mean follow-up of 5.7 months (SD+/-1.7 months). Seventeen patients (70%) reported improvement in NYHA functional class from NYHA III to NYHA II (p value: 0.04), independently from the ventricular physiology. In the overall cohort, BNP values presented a considerable reduction but didn't reach statistical significance (mean value 1333.4 +/- 3027 pg/ml to 506.1 +/- 573.6 pg/ml; p:0.07). In the subgroup analysis a significative reduction of BNP values was observed in patients with SRV (641.3+/-661.2 pg/ml to 445.1 +/- 501.8 pg/ml, p:0.02). Only eleven patients (46 %) presented a slight increase in VO2 max values at CPET (15.8 +/- 4.4 l/min to 15.8 +/- 2.9 l/min), however this data didn't reach statistical significance either in the overall cohort or in subgroups analisys. Only two patients discontinued therapy after a period of two months for arterial hypotension (poor tolerance).
Conclusions: Our experience suggests that sacubitril/valsartan therapy is well-tolerated in ACHD- HF patients. The use of sacubitril/valsartan is associated with improvement in NYHA functional class and with a reduction trend in BNP values.

Keywords: ACHD, Adult Congenital Heart disease, Heart failure

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Congenital Heart Surgery

PP-011

Performance and failure of surgically implanted right ventricle to pulmonary artery (RV-PA) conduit in congenital heart disease

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Background and Aim: Surgical implantation of right ventricle to pulmonary artery (RV-PA) conduit is an important component of congenital heart disease (CHD) surgery but with limited durability, and need for re-intervention. Current single center, retrospective, cohort study is reporting longterm performance of surgically implanted RV-PA conduit in a consecutive series of children and adults with CHD.
Method: Patients with CHD referred for RV-PA conduit surgical implantation (October 1997 and January 2021) have been included. Primary outcome was conduit failure defined as peak gradient above 64mmHg or need for conduit-related interventions. Longitudinal echocardiographic studies were available for mixed-effect linear regression analysis.
Results: Two-hundred and fifty-two patients were initially included. One hundred and forty-nine patients were available and elegible for follow-up data collection. After a median follow-up time of 49 months (IQR 9-132 months) the primary study endpoint occurred in 44 (29%) patients. The 1- 5- and 10-year freedom rate from conduit failure was 91%, 83% and 66%. The 10-year freedom rate from conduit failure was 46% in the non-homograft group compared to 85% in the homograft group. Similarly, the 10-year freedom rate from conduit failure was 47% in patients implanted before age of 18 years compared to 86% in patients implanted after age of 18 years. Multivariable Cox regression model identified adult age (>18 years) at implantation and pulmonary homograft as protective factors (HR 0.11, 95% CI 0.02-0.47, p-value 0.003 and HR 0.34, 95% CI 0.16-0.74, p-value 0.006, respectively). Fever within 7 days of surgical conduit implantation was a strong, independent risk factor for early (within 24 months) failure (HR 4.29, 95% CI 1.41-13.01, p-value 0.01). Longterm use of oral anticoagulant was independently associated with slower progression of peak echocardiographic gradient across conduits (p-value 0.027).
Conclusions: In patients with CHD, surgically implanted RV-PA conduit failure is faster in children and after non-homograft conduit implantation. Early fever after surgery is a strong risk factor for early failure. Longterm anticoagulation seems to exert a protective effect.

Keywords: RV-PA conduit, CHD, Cardiac Surgery

Adult Congenital Heart Disease

PP-012

Atrial membranes: Clinical problem or incidental finding

Evmorfia Ladopoulou[1], Panagiota Valaki[1], Maria Karakosta[2], Alkistis Kalesi[2], Antonios Destounis[2], Nearchos Kasinos[2], Anastasios Theodosis Georgilas[2], Niki Lama[3], Stefanos Despotopoulos[4], Sotiria Apostolopoulou[4], Eythymios Brachos[1], Nikolaos Tziolas[1], Konstantinos Tsatiris[1] and Panagiotis Zachos[5]
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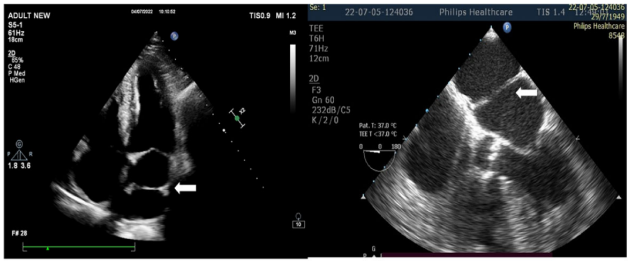
Background and Aim: Atrial membranes (AM) are thin fibromuscular bands that divide either the left (Cor Triatriatum Sinister) or the right atrium (Cor Triatriatum Dexter), thus creating three atrial compartments. Cor Triatriatum Sinister (CTS) is more frequent and causes obstructive phenomena of varying severity. They should not be confused with the supravalvular mitral annulus ring, which is located behind the appendage. It is a rare congenital anomaly (0.1-0.4%), while in approximately 80% of cases in the pediatric population is combined with other congenital heart diseases, with most common being ostium secundum atrial septal defect and anomalous pulmonary vein return.
Method: We present two cases of our clinic's Pediatric Cardiology and Adult Congenital Heart Diseases Department with incidentally detected AM.
Results: The first case concerns a 72-year-old man with a history of arterial hypertension, who came to the emergency department (ED) with progressively worsening exertional dyspnea. Transthoracic echocardiography (TTE) revealed significant mixed aortic valve disease and the presence of an AM within the left atrium, which did not cause obstructive phenomena. The patient underwent a transesophageal study (TEE), in which the findings of TTE were confirmed without coexistent congenital anomalies.
The second case concerns a 69-year-old woman with a history of surgical replacement of the aortic and mitral valves due to rheumatic attack, who was admitted to our clinic due to paroxysmal

Left Atrial Membrane



Snapshots of transthoracic (left) and transesophageal (right) ultrasound from the first case. The left atrial membrane is marked with an arrow.

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nocturnal dyspnea. During hospitalization, the patient underwent TTE and TEE in order to check the prosthetic valves, where the well-known significant mismatch between the patient's aortic annulus and the prosthetic valve (Prosthesis-Patient Mismatch) was re-checked and a right atrium AM was imaged without evidence of obstruction or the presence of another congenital malformation.
Conclusions: The finding of Cor Triatriatum in adulthood is a rare and usually an incidental finding, which does not cause particularly obstructive phenomena and symptoms. Although a benign entity, it is associated with the presence of atrial arrhythmias and cardioembolic episodes. The coexistence of AM and aortic valve stenosis in our patients should probably be considered coincidental, although the existence of a bicuspid aortic valve in patients with Cor Triatriatum is not rare.

Keywords: membranes, left atrium, right atrium, obstructive

PP-013
Gerbode defect: An extremely rare complication of atrial septal defect repair surgery

Evmorfia Ladopoulou[1], Panagiota Valaki[1], Maria Karakosta[2], Alkistis Kalesi[2], Antonios Destounis[2], Nearchos Kasinos[2], Anastasios Theodosis Georgilas[2], Niki Lama[3], Stefanos Despotopoulos[4], Sotiria Apostolopoulou[4], Eythymios Brachos[1], Nikolaos Tziolas[1], Konstantinos Tsatiris[1] and Panagiotis Zachos[5]
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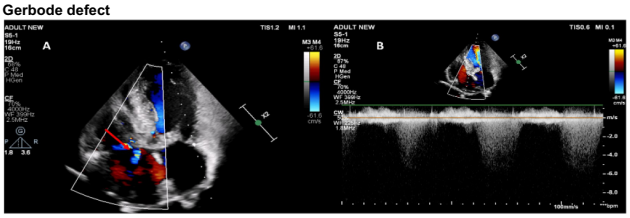
Background and Aim: Gerbode defect is a rare abnormal shunting between left ventricle (LV) and right atrium (RA). Despite that it is classified as congenital heart disease, the number of acquired cases is clearly greater. The acquired form can be iatrogenic, either after cardiac surgery (aortic/mitral valve replacement, repair of atrial/ventricular septal defect) or after percutaneous cardiac interventions (ablation of atrioventricular node, myocardial biopsy, tricuspid ring implantation) and non iatrogenic, with main representer the infective endocarditis.
Method: We report a case of the Pediatric Cardiology and Adult Congenital Heart Disease Department of our hospital, with Gerbode defect, after surgical repair of primum atrial septal defect.
Results: The case concerns a 47 years old male, with medical history of surgical closure of a primum atrial septal defect and anterior mitral leaflet cleft repair a year ago, who proceeded for first-year follow up. The transthoracic echocardiogram (TTE) revealed normal dimensions and systolic contractility of LV, borderline right ventricle dimension with normal contractility and the presence of a patch in interatrial septum area, without residual shunt. With the contribution of color Doppler, moderate both atrioventricular valves regurgitations were revealed, without signs of pulmonary hypertension, and a small, restrictive, hemodynamically insignificant communication between LV and RA, Gerbode type, with L-R flow. A transoesophageal study (TOE) followed, which confirmed the findings of TTE, while residual atrial defect was ruled out with bubble study. The patient was referred to a specialized center where he underwent a cardiac magnetic resonance which confirmed the diagnosis of postoperative Gerbode defect.
Conclusions: The surgical intervention for congenital heart disease is the second most common cause of iatrogenic Gerbode defect, with the closure of ventricular septal defect being the most common one. Only two other cases of Gerbode defect after surgery for primum atrial septal defect and mitral cleft have been reported. Its diagnosis is a challenge with TTE as the systolic blood flow, being the result of a significant difference in pressure between LV and RA, can be mistaken for tricuspid regurgitation.

Keywords: shunting, repair, atrial septal defect



A.Gerbode defect (arrow) B.Restrictive shunt

PP-014
Coexistence of atrioventricular septal defect and hypertrophic cardiomyopathy in patient with down syndrome

Evmorfia Ladopoulou[1], Panagiota Valaki[1], Maria Karakosta[2], Alkistis Kalesi[2], Antonios Destounis[2], Nearchos Kasinos[2], Anastasios Theodosis-Georgilas[2], Niki Lama[3], Stefanos Despotopoulos[4], Sotiria Apostolopoulou[4], Eythymios Brachos[1], Nikolaos Tziolas[1], Konstantinos Tsatiris[1] and Panagiotis Zachos[5]
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Background and Aim: The Congenital Heart Disease (CHD) is found in 40% of infants with Down syndrome (DS). The most common CHD in these patients are defects in the atrioventricular septum (partial or complete atrioventricular canal defects). These are often associated with chromosomal aberrations, such as in trisomy 21. Hypertrophic cardiomyopathy (HCM) is typically an isolated cardiac lesion. In the newborn or the fetus is usually transient and attributed to certain maternal metabolic disorders or to exposure to steroids.
Method: We report a case with DS, operated Atrioventricular Septal Defect (AVSD) and HCM of our Pediatric Cardiology and Adult Congenital Heart Disease Department.
Results: The case concerns a female with DS and operation in her first year for complete AVSD, who proceeded asymptomatic for yearly follow up. The transthoracic echocardiography (TTE) revealed typical asymmetric HCM without left ventricular outflow tract obstruction (LVOTO). Right ventricle was within normal dimensions, had increased wall thickness and borderline contractility. Furthermore, there was a cleft of the anterior mitral leaflet with moderate mitral valve regurgitation and the patch in the area of the crux was without residual defect. Subsequently, the patient underwent a cardiovascular magnetic resonance imaging, which confirmed the diagnosis of HCM without depicting of myocardial fibrosis.

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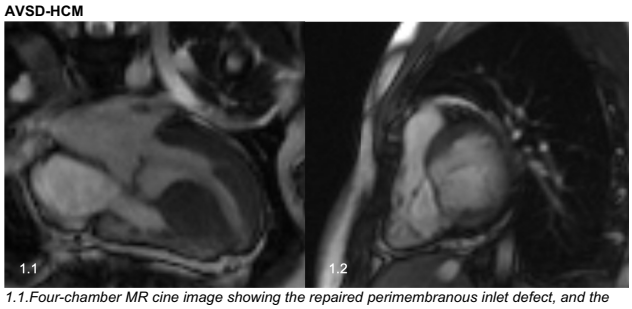
Conclusions: It is an uncommon finding the HCM phenotype as a primary lesion in patients who have CHD. Case reports have documented the occurrence of HCM in patients who have congenital atrial and ventricular septal defects, coarctation of the aorta, and tetralogy of Fallot. Moreover, in certain syndromes, such as Noonan's syndrome and LEOPARD syndrome, HCM can occur in conjunction with congenital heart disease. To the best of our knowledge, HCM has been only twice reported in association with AVSD in a newborn with trisomy 21. While exact genetic evidence is lacking at present, the combination of HCM and AVSD in Down syndrome is more likely an inherited disorder with a common genetic mechanism than a rare, sporadic event.

Keywords: Atrioventricular defect, Hyperthrophy, Genetic, Sporadic



1.1.Four-chamber MR cine image showing the repaired perimembranous inlet defect, and the asymmetric hypertrophy of interventricular septum 1.2.Short-axis MRI cine image showing septal leaflets of atrioventricular valves attached at the same level to the central fibrous body.

PP-015
Iron homeostasis in adults with complex and/or cyanotic congenital heart disease

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Background and Aim: Data on iron homeostasis in adults with complex and/or cyanotic congenital heart disease (CHD) is limited, although targeted iron treatment is a cornerstone of heart failure management in these patients. The aim of this study was to evaluate the impact of iron deficiency on clinical status in adults with complex and/or cyanotic CHD.
Method: In this cross-sectional clinical study, clinical and laboratory data were collected from n=181 patients (53.6% female; mean age 37.4 ±14.0 [18-83]) years with complex and/or cyanotic CHD. Iron deficiency was defined as serum-ferritin levels <20ng/ml (women), <30ng/ml (men) or soluble transferrin-receptor levels >4.4mg/l (women), >5mg/l (men) or transferrin-saturation <16%. Clinical status was assessed by functional classes according to Perloff. Data collection was performed between 01/2017 and 12/2022 at a specialized tertiary care center for adults with CHD.
Results: Mean oxygen saturation was 85.8 ±8.7%, n=123 (68%) patients were cyanotic (SpO2 ≤90%). Iron deficiency was more prevalent in cyanotic than acyanotic patients (58.5% vs 31.0%, p<0.001). Iron deficient, cyanotic patients were in worse functional classes, 54.2% vs. 34.3% being in functional class ≥3 (p=0.008), indicating significant manifestation of heart failure. In cyanotic adults with CHD, hemoglobin (17.1 ±3.9 vs. 17.8 ±2.6; p=0.238) and serum-ferritin levels (124.8 ±291.4 vs. 188.4 ±240.2; p=0.202) did not differ substantially between patients with or without iron-deficiency, yet oxygen saturation was lower in the iron-deficient subgroup 79.9 ±7.9% vs 83.8 ±5.0% (p=0.001). Mean corpuscular hemoglobin (MCH) and mean corpuscular volume (MCV) were lower in patients with iron deficiency (p<0.001 and p=0.002), yet mostly within the normal range.
Conclusions: We conclude that iron deficiency is a common problem in adults with complex and/or cyanotic CHD and is associated with a poorer functional class. The diagnosis of iron deficiency in these patients is of paramount importance but is challenging because hypoxia-induced secondary erythrocytosis masks anemia and pseudonormal ferritin levels mimic iron deficiency in these patients. More studies are needed to better understand iron homeostasis in cyanotic CHD patients. Based on this, targeted therapeutic strategies need to be developed to improve the treatment of heart failure in this vulnerable group of adults with CHD.

Keywords: iron, iron deficiency, adult congenital heart disease, cyanosis, heart failure

PP-016
'Pathfinder-CHD registry': A prospective, comprehensive heart failure database of adults with congenital heart disease

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Background and Aim: Heart failure is a major challenge in the management of adults with congenital heart defects (CHD). It is complicated by the complexity of the underlying CHD as well as residua from previous surgical or interventional treatment. Therapeutic approaches often differ markedly from strategies applied in acquired heart disease. The goal of this registry is to document clinical and epidemiological data, treatment approaches, and clinical outcomes of adults with CHD and heart failure based on real-world data.
Method: The *Pathfinder-CHD Registry* is a prospective, observational, web-based heart failure registry established in 2022 in Germany. Adults with CHD and all forms of manifest heart failure, previous heart failure, or at significant risk for future heart failure from abnormal ventricular function or anatomy are included.
Results: Since November 2022, n=1,377 patients (mean age 37.4 ±17.3 years; 47.5% female) met inclusion criteria and gave consent to participate in this registry with longitudinal follow-up.
Of the patients included, n=554 (40.2%) had 'complex heart anomalies' including univentricular hearts, AV-valve atresia,

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congenitally corrected transposition of the great arteries, transposition of the great arteries, truncus arteriosus, or Ebstein´s anomaly. n=432 patients (31.7%) had 'right-sided heart anomalies' including tetralogy of Fallot, pulmonary atresia, double outlet right ventricle, and pulmonary valve disease. 'Left-sided heart anomalies' (n=143, 10.4%) encompassed aortic valve disease, aortic coarctation, and interrupted aortic arch. 'Left-to-right shunt lesions' were present in n=129 (9.3%), and 'others' in n=119 (8,6%). As the primary diagnosis, n=870 (63.2%) patients had a cyanotic CHD.
The registry provides comprehensive documentation of the underlying congenital heart defects and type of heart failure as well as medical, surgical, and/or interventional treatments, comorbidities, and outcomes.
Conclusions: Containing comprehensive data on CHD-associated heart failure, the *Pathfinder-CHD Registry* database will help to evaluate disease progression, risk factors, and efficacy of different treatment strategies in the long term. The registry promotes the goal of evidence-based, personalized treatment and helps to provide predictive models to effectively address the challenges of adults suffering from CHD and heart failure.

Keywords: adult congenital heart disease, heart failure, registry, Pathfinder

PP-017

Longitudinal evaluation of renal function in patients with a fontan circulation

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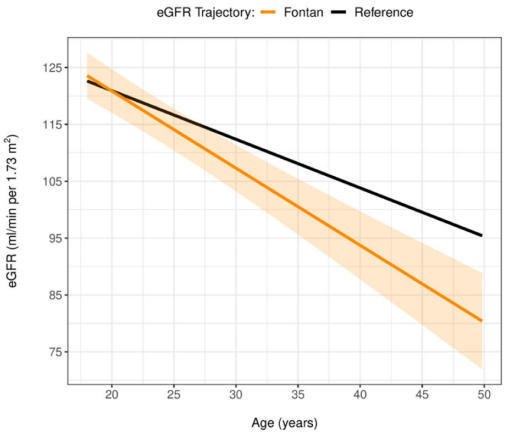
Background and Aim: Renal dysfunction is a recognized, yet underexposed co-morbidity in Fontan patients and related to poor outcome. The evolution of Fontan-associated renal dysfunction is, however, insufficiently described, and the need for longitudinal studies has recently been acknowledged. The aim of this study is to investigate the prevalence of renal dysfunction and to investigate the course of creatinine-based eGFR (eGFRcr) and its determinants over time in patients with a Fontan circulation.
Method: This longitudinal single-center study included 82 patients with a Fontan circulation. Renal dysfunction was defined as eGFR < 90 ml/min/1.73m2 and its prevalence was calculated with both creatinine (eGFRcr) and cystatin C-based eGFR (eGFRcys) at baseline. Longitudinal data for eGFRcr were collected over an 11-year period. Annual changes in eGFRcr and its determinants were investigated with linear mixed-effect models in adult Fontan patients. Additionally, the change in eGFRcr over time in the adult Fontan cohort was compared with reference values for eGFRcr from previously published normative data of a healthy Dutch cohort. NT-proBNP values were converted to a z-score in order to account for its age dependent factor.
Results: Median baseline age of the Fontan cohort was 20 (IQR [14 – 27]) years and 51% of the cohort were females. Mean baseline eGFRcr was higher than eGFRcys (108 ± 18 vs 102 ± 22 ml/min/1.73m2, $p < 0.001$). The prevalence of renal dysfunction in the adult Fontan patients was 10% for eGFRcr and 17% for eGFRcys. Creatinine-based eGFR showed to be comparable to that of healthy individuals at the start of adulthood. However, subsequent deterioration of eGFRcr was faster in the adult Fontan patients than in healthy individuals (-1.36 vs -0.86 ml/min/1.73m2 per year) (Figure). Furthermore, higher NT-proBNP z-scores at baseline were associated with a more rapid decline in eGFRcr, independent of baseline renal function.
Conclusions: Creatinine leads to a higher eGFR than cystatin C in adult Fontan patients. Creatinine-based eGFR, is comparable to healthy individuals at the start of adulthood but then declines more rapidly over time. Baseline NT-proBNP z-score is a strong predictor of accelerated deterioration of renal function in the future.

Keywords: Congenital, univentricular heart, Fontan, renal function, creatinine, longitudinal

Figure | Deterioration of $eGFR_{cr}$ in adult Fontan patients compared to healthy individuals



The orange line represents the predicted course of $eGFR_{cr}$ during adulthood in our Fontan population, as obtained from a linear mixed-effect model. The shaded area around the line is the corresponding 95% CI. The black line depicts the mean change in $eGFR_{cr}$ over time for a healthy eighteen-year-old individual.
$eGFR_{cr}$ = Estimated creatinine-based glomerular filtration rate; CI = Confidence interval.

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General Cardiology

PP-019

Incidence, current management practice and outcome of unexpected hospitalization in patients with fontan circulation in JAPAN (JAPAN fontan registry)

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Background and Aim: All-cause morbidity that required unexpected hospitalization (UEH) remains high in patients with Fontan circulation (FC) and the management practice may vary widely in Japan. Aim of the present study was to clarify the incidence, current practice and outcome of UEH in Japan.
Method: We conducted a multi-center prospective study which was consisted of 3-year registration of UEH FC patients and 3-year follow-up after the discharge.
Results: RESULTS: We followed 3227 FC patients (mean age of 14 years) in 20 Japanese institutions and 243 (7.5%) patients experienced UEH, including 7 death (2.9%) during UEH, in the scheduled 3-year-registration period. The most common cause was heart failure (HF, 19.3%), followed by infection (18.9%) and protein losing enteropathy (PLE, 16.5%), hemorrhage (13.2%), arrhythmia (9.5%), thromboembolism (TE, 2.5%), plastic bronchitis (1.6%), and others (18.5%). The most common diagnosis was mitral valve atresia (12.5%), followed by univentricular heart (9.5%) and hypoplastic left heart syndrome (8.0%). Overall median length of UEH-stay (days) was 9 and the longest stay was due to TE (20), followed by PLE (18) and HF (13). During a median follow-up of 2.0 years after discharge, 76 (32%) patients re-hospitalized with the highest cause of PLE (34.2%), followed by HF (15.8%) and hemorrhage and infection (13.2% for each). High-volume institutions (HV, $\geq$ 200 FC patients) showed a low risk of UEH (hazard ratio [HR]; 0.38, $p < 0.0001$), whereas a high rate of re-UEH (HR: 1.58, $p < 0.05$) when compared with that of non-HV. High plasma levels of alkaline phosphatase and low levels of albumin at discharge were independently associated with a high risk of re-UEH ($p < 0.05$).
Conclusions: HF, PLE, infection and hemorrhage were main causes of UEH in Japanese FC patients. Establishing management strategy for these main pathophysiology may be imperative for the better quality of life.

Keywords: Fontan circulation, morbidity, hospitalization, heart failure, protein losing enteropathy

Adult Congenital Heart Disease

PP-020

Outcomes of pregnant women with congenital heart disease attending a multidisciplinary cardio-obstetric clinic in cape town south africa

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Background and Aim: Congenital heart disease (CHD) patients are at risk for cardiovascular complications during pregnancy. Despite being the leading cause of maternal death in high-income settings, evidence from low and middle-income countries is lacking. We aimed to characterize outcomes of pregnant women with CHD referred to a multi-disciplinary combined cardio-obstetric clinic (CCOC), between 2017-2023.
Method: Pregnant women with CHD were invited and consented to participate in the PROTEA (partnerships for children with heart disease) registry. Demographics, obstetric and surgical history, and pre-partum, peripartum, and post-partum complications and events were recorded.
Results: 58 participants were enrolled over 7 years; median age was 27 years (IQR:24-32). Median booking BMI was 27 (IQR:23-35), with 29% overweight (BMI:25.0–29.9), and 34% obese (BMI $\geq$ 30). Predominant diagnoses included Ventricular Septal Defect 33% (20/61 total diagnoses), Tetralogy of Fallot 20%(12/61), Atrial Septal Defect 15%(9/61), Pulmonary Stenosis 5%(3/61), Aortic Coarctation 5%(3/61), and Atrial Ventricular Septal Defect 3%(2/61). 40 participants (69%) had a history of cardiac surgery. Most (98%, 57/58) participants had pre-existing cardiac diagnoses, however only 53% (31/58) of participants received pre-pregnancy counselling. In multigravida

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participants 58% (14/24) had a history of obstetric complications, with 75% (18/24) of pregnancies complicated by spontaneous abortion (9), therapeutic abortion (6) or intrauterine death (3). Comorbidities included angina (9), hypertension (7), asthma (4) and HIV (3). At enrolment 67% of participants presented in NYHA heart failure class 1, 23% in class 2, 9% in class 3, and 2% in class 4. During their pregnancies 19% (11/58) experienced obstetric complications for which 21% (12/58) required admission. Additionally, 12% (7/58) were admitted for cardiac complications.
Median gestational age at delivery was 38 weeks (IQR:35-40), 44% by elective caesarean section, 7% by emergency caesarean section. There were no maternal events during delivery; 2 experienced infective complications post-delivery. There were 0 maternal mortalities and 2 foetal mortalities.
Conclusions: Despite suboptimal preconceptual counselling in our population of pregnant women, we present excellent outcomes for pregnant women with a variety of CHD diagnoses treated in a multidisciplinary cardio-obstetric clinic. Future interventions should optimize preconceptual counselling, awareness of healthy weight and consolidation of the multidisciplinary heart team approach.

Keywords: South Africa, Cardio-Obstetric Clinic, Congenital Heart Disease, Pregnancy Complications, PROTEA

PP-021
Successful atrial septal defect (ASD) device closure in adults under transoesophageal echocardiography (TOE) guidance only

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Background and Aim: Transcatheter closure of atrial septal defects (ASDs) offer less invasive approaches allowing rapid recovery and earlier hospital discharge. Echocardiography plays important role in spanning diagnosis, detailed anatomical assessment, device sizing and selection, peri-procedure guidance, and post device surveillance. We present two cases of ASD device closure in adults under transoesophageal echocardiography (TOE) guidance only.
Method: Two patients with diagnosis of ASD were identified to be suitable for transcatheter device closure. They were discussed in the multidisciplinary meeting and offered the procedure.
Results: Case 1: Female, 57, mother of two children, does not do much exercise, recent shortness of breath, NYHA Functional Class III. Had ASD with significant left to right shunt (Qp:Qs 2.9:1 on cardiac MRI) and significant right heart dilatation. She is in sinus rhythm. Cardiac catheter under general anaesthetic showed mean pulmonary artery pressure of 27 mmHg. Using TOE and balloon sizing (PTSX 40X50, stop flow technique), the defect measured 26 mm. A 28 mm CeraFlex Lifetech device was used to close the ASD under TOE guidance only. The device was delivered using a 12F delivery sheath and was confirmed to be in a good position with no residual shunt. Case 2: Male, 53, shortness of breath with exercise was found incidentally to have large ASD measuring 13 mm with adequate margins and right ventricle volume overload. Catheter done under general anaesthetic and TOE guidance, Balloon sizing of the defect was done using a 25 mm PTSX balloon with stop flow technique. The defect measured 20 mm. A 20 mm Amplatzer septal occluder was delivered through a 9F delivery sheath. Device deployed into a good position with position was checked and then device no residual flow.
Conclusions: ASD device closure using TOE guidance only is feasible and successful. This allows good results without the need for radiation or contrast exposure, especially in high risk patients.

Keywords: Atrial septal defects, transoesophageal echocardiography

PP-022
Non-surgical critically ill adult patients with congenital heart disease in a general intensive care unit: Case series from a second level hospital

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Background and Aim: Most of congenital heart patients reach adulthood, even undiagnosed, and this means an increase in medical attentions, but sometimes they must be attended by non-cardiologist physicians, mainly in second level centres. In critical situations, initial stabilization is mandatory but anatomy and/or physiology must be taken into account in these cases. Clinicians in second level hospitals think they will never attend these patients and nothing can be done without advanced therapies, as extracorporeal membrane oxygenator (ECMO).
Method: We reviewed adult patients with congenital heart disease admitted from December 2018 to August 2023 in the intensive care unit (ICU) at our institution. Patients with coronary anomalies or with surgical causes of admission were excluded.
Results: There were 4 cases (50% women) with mean age 45,5 (range 23 to 71), 75% previously known and controlled. 50% were moderate or more (25% moderate, 25% severe). Congenital heart diseases were: univentricular heart palliated by Fontan surgery (failing stage); repaired ostium secundum atrial septal defect (ASD) and mechanical aortic and mitral prothesis with severe aortic leak, repaired sinus venosus ASD and unrepaired anomalous left coronary artery from pulmonary artery (ALCAPA) with rheumatic mitral regurgitation. Patients´ complaints were dyspnoea, chest pain, dizziness, palpitations. 75% of admissions were due to a cardiovascular cause. Discharge diagnoses were adult distress respiratory syndrome, type B aortic dissection, bradycardia due to beta-blockers and ventricular tachycardia. 50% were transferred, one for ECMO and other for surgical reparation. All of them survived in our centre and were discharge alive from referral centre. Patients´ characteristics are shown in image.

Table

Patient	Sex	Age	Congenital heart disease	Grade	Previously known?	Previous follow-up	reason of admission	Transfered?	reason for transferring	Final diagnosis	Discharge alive from referal centre?
1	W	36	Univentricular heart with fontan paliation. Failing fontan stage	Severe	Yes	Yes	Dyspnea	Yes	ECMO (ICU)	ADRS	Yes
2	M	52	OS-ASD (repaired). Mechanical aortic prothesis with severe leak, mitral and tricuspid annuloplasty.	Simple	Yes	Yes	Chest pain	No	No transfered	Type B aortic disection	Yes
3	M	71	SV- ASD (repaired)	Simple	Yes	Yes	Dizziness	No	No transfered	Brdicardia due to BB	Yes
4	W	23	ALCAPA (unrepaired), Severe MR (rheumatic)	Moderate	No	No	Palpitations	Yes	Surgical correction	VT	Yes

W: woman; M: man; ASD: atrial septal defect; OS: ostium secundum; SV: sinus venosus; ALCAPA: Anomalous left coronary artery arising from pulmonary artery; ICU: intensive care unit; ECMO: extracorporeal membrane oxigenator; ADRS: adult distress respiratory syndrome; VT: ventricula tachycardia

Patients´ characteristics

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Conclusions: Although rare, general intensivists of non-tertiary hospital will probably have to attend these patients in the future, even the more complicated or previously unknown cases. Because of differences from other cardiopathies, prompt consultation and multidisciplinary approach will be necessary. Despite the lack of advanced therapies, it is possible to manage without transferring or even stabilize and then transfer these patients, allowing them to survive.

Keywords: critically ill, adults, second level hospital

PP-023

Patients referred for surgical correction from an adult congenital heart disease clinic in a second level hospital

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Background and Aim: Adult congenital heart disease (ACHD) may need surgical treatment at any time of the life but due to longer time of evolution, general cardiologists tent to think once reached adulthood, nothing is necessary to do. Because of that, many patients have been discharge once they reached adulthood.
Method: We reviewed patients referred for surgical correction from our ACHD clinic. Patients´characteristics were reported.
Results: There were 7 patients (71% women), mean age 35 (range 17-71). All but one were moderate. Mean age at diagnosis was 18 (range 0 to 46). 3 patients (43%, all women) were diagnosed at born, the rest above 17. Among those diagnosed in adulthood (4 patients, 50% women) mean age was 31 (17 to 46). In 4 patients (57%) the cardiopathy was previously known, 2 with follow-up from birth, 1 with follow-up in pediatric life and then missing from 18 to 36 and then followed during 17 years by general cardiologist and 1 diagnosed at 46 without follow-up. Among those diagnosed in adulthood, only 1 patient had a previously known condition, without follow-up during 25 years. Patients diagnosed at born had 1 (valvuloplasty), 2 (surgery) and 4 (surgeryx2, arrhythmia ablation, internal cardioverter-defibrilator implantation) previous cardiac procedures respectively. 3 patients (66% women) were referred to surgery at the time of diagnosis. Mean time from diagnosis to surgical referral was 18 years (0 to 25), and among previously known was 31 years (18 to 55). Characteristics are shown in figure.
Conclusions: ACHD patients may need surgical reparation at any time in adulthood, even those with other, sometimes multiple, cardiac procedures, so they need follow-up forever. Diagnosis and referral for correction can happen in adulthood also. As evolution is longer than acquired cases, cardiologist tent to think they will always be stable and never would need surgical correction. Follow-up in specific clinics and referral to tertiary centers may be necessary in some cases.

Keywords: adults, referral, surgery

Figure

case	sex	diagnosis	Grade	Previously known	age at diagnosis	Previous intervention	Intervencions	type of intervention	follow-up	Age at follow-up	Age at reparation	time from diagnosis to reparation
1M		ASD-OP	moderate	No	25	No	None	None	Not previously known	Not follow-up	25	0
2M		ASD-OP	moderate	No	36	No	None	None	Not previously known	Not follow-up	36	0
3H		severe TR, not Ebstein	moderate	No	17	No	None	None	Not previously known	Not follow-up	17	0
4H		ASD-SV	moderate	Yes	46	No	None	None	No	Not follow-up	71	25
5M		ToF, Severe PR, VT	moderate	Yes	0	Yes	5(4)	Transanular patch, infundibular resection, VT ablation, ICD implantation.	Yes	since birth until 18 and from 36	55	55
6M		PDA+CIA-OP. Severe MR	moderate	Yes	0	Yes	5(2)	PDA closure. Closure ASD+mitral reparation	Yes	since birth	25	25
7M		AoS. Severe AR	Mild	Yes	0	Yes	5(1)	Aortic valvuloplasty	Yes	since birth	18	18

ASD: atrial septal defect; OP: ostium primum; SV: sinusvenosus; TR: tricuspid regurgitation; ToF: Tetralogy of Fallot; PR: pulmonary regurgitation; VT: ventricular tachycardia; PDA: patent ductus arteriosus; MR: mitral regurgitation; AoS: aortic stenosis; AR: aortic regurgitation; ICD: internal cardioverter-defibrilator

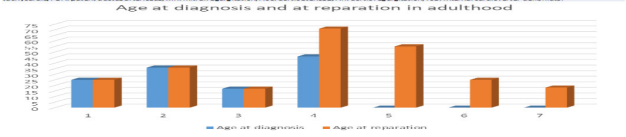

Table with patients´ characteristics and graphic with age at diagnosis and at reparation in adulthood of each patient

Cardiac Imaging

PP-024

Multimodality cardiac imaging to asess aortic valve anatomy in doubtful cases in adults. Case series from a general cardiology clinic

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Background and Aim: Because of poorer echo windows, assessing aortic valve in adults may need other techniques, as magnetic resonance (MRI) or cardiac computed tomography (cCT).
Method: We reviewed cases from an adult cardiology clinic with MRI o cCT to assess aortic valve. Characteristics were reported
Results: There were 14 cases: 36% women; mean age 60.4 years (21 to 77; 86% older than 50). All cases except three (2 women) were new ones. All patients except four had valvar dysfunction in echocardiogram and MRI, milder in the latest in 28.5% of cases. 50% patients had aortic dilatation (57% women), mean age 68 years old, ranged from 50 to 77. MRI was conclusive in all cases except three men (21,4%, mean age 68.7, range 57 to 77). Among these, only 1 cCT was performed with tricuspid valve (1 patients refused (probable tricuspid), 1 died before cCT (probable bicuspid). Definite diagnosis was achieved in 86% of patients. There were no other anomalies except in one case with left ventricular dysfunction. 64.3% of patients had any congenital valvar anomaly (definite or probable) (22% women; mean age 58 years) 78% bicuspid (BAV) with two rare cases (both men, mean age 29.5): unicuspid (without other anomalies) and quadricuspid (with aortic regurgitation). 28.5% of cases had definite tricuspid valve (75% females, mean age 61.3 years, 50% with no alterations, the others valvar and aortic affectation). 43% of cases presented definite BAV (one-third women, mean age 65.3 years, all with valvar alterations and two-thirds with aortic dilatation). 57% of patients had a definite non-tricuspid valve (25% women, mean age 56.4 years). All except one had valvar and/or aortic alterations. 50% of patients (7) had ≥60 years. Among them, there were 3 women (42%): 2 definite tricuspid, 1 definite bicuspid, and 4 men: 2 bicuspid, 1 probable bicuspid, 1 probable tricuspid. Image describes characteristics and shows images from exams.

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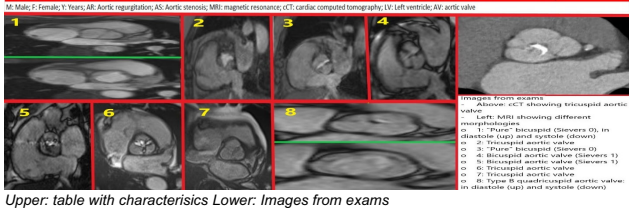
Conclusions: BAV can be even diagnosed in elderly patients. Diagnosis can be achieved by MRI, but sometimes cCT is needed, specially in men. Tricuspid anatomy might present female predominance but older cases might have no sex predominance but male predominance for BAV.

Keywords: aortic valve, congenital alterations, cardiac magnetic resonance, cardiac computed tomography

Image

case	sex	age	New case	follow-up (y)	Aortic valve anomalies (echo)	Aortic valve anomalies (MRI)	Aortic dilatation	MRI conclusive	cCT perform	Cause cCT not performed	Other anomalies	AV morphology (final diagnose)
1	M	73	Yes	0	No	No	No	Yes	No	MRI conclusive	No	Tricuspid
2	M	38	No	6	Mild-to-moderate AR	Mild AR	No	Yes	No	MRI conclusive	No	Quadricuspid
3	F	76	Yes	0	Severe AS. Mild AR	Severe AS. Mild AR	Yes	Yes	No	MRI conclusive	No	Bicuspid
4	M	58	Yes	0	Moderate AS and AR	Moderate AS. No AR	Yes	Yes	No	MRI conclusive	No	Bicuspid
5	M	73	Yes	0	Severe AS	Severe AS	Yes	Yes	No	MRI conclusive	No	Bicuspid
6	M	59	Yes	0	Severe AS	Severe AS	No	Yes	No	MRI conclusive	No	Bicuspid
7	M	76	Yes	0	Severe AS	Severe AS	No	Yes	No	MRI conclusive	LV disfunction	Bicuspid
8	F	50	No	10	Moderate AS	Moderate AS	Yes	Yes	No	MRI conclusive	No	Bicuspid
10	M	72	Yes	0	Severe AS	Moderate AS	Yes	No	No	Refused	No	Bicuspid (probable)
9	M	77	Yes	0	No	No	Yes	No	No	Death	No	Tricuspid (probable)
11	M	57	Yes	0	No	No	No	No	Yes	Performed	No	Tricuspid
12	F	69	No	3	Moderate AR	Mild AR	Yes	Yes	No	MRI conclusive	No	Tricuspid
13	F	68	Yes	0	Moderate AS	Moderate AS	No	Yes	No	MRI conclusive	No	Tricuspid
14	F	51	Yes	0	No	No	No	Yes	No	MRI conclusive	No	Tricuspid

M: Male; F: Female; Y: Years; AR: Aortic regurgitation; AS: Aortic stenosis; MRI: magnetic resonance; cCT: cardiac computed tomography; LV: Left ventricle; AV: aortic valve



Upper: table with characterisics Lower: Images from exams

Genetics, Basic Science and Myocardial Disease

PP-025

P.HIS2476ARG: Variant of unkown significance (VUS) in flnc GEN possibly pathogenic in a teenager with hypertrophy and elevated intracardiac pressures

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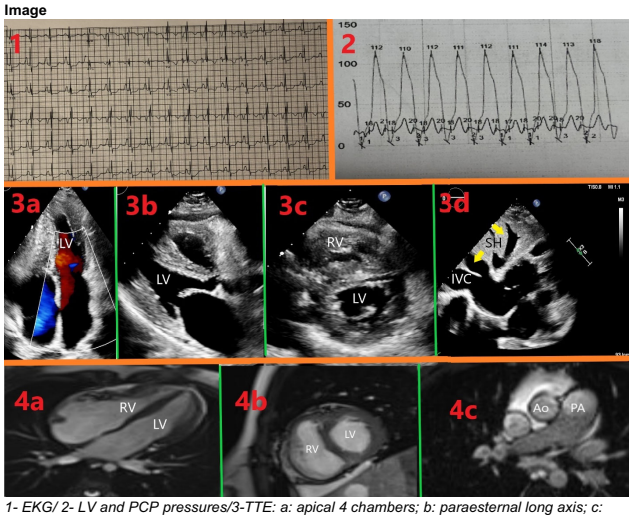
Background and Aim: Some mutations are not clearly cause of a disease. They are called variants of unknown significance (VUS). Some of them could potentially cause a phenotype, and some data can be taken into account, such as, mutations in similar region or concordance between genotype-phenotype among relatives of a patient.
Method: A 15 years and 4 months Romanian female teenager was referred to our adult congenital heart disease and pulmonary hypertension clinic due to alterations in electrocardiogram (EKG) and signs pulmonary hypertension (ratio pulmonary artery (PA)/aorta (Ao)>1 in cardiac magnetic resonance (MRI). Transthoracic echocardiogram (TTE), MRI, cardiopulmonary exercise test (CPET), cardiac catheterisation (CC) and panel of myocardiopathies with 225 genes (NGS) in a blood sample, as well as abdominal echography, were performed to assess the cause.
Results: The patient had neither personal nor familiar history of any cardiac or extracardiac diseases. Development was normal, but she referred short of breathness "all her life" and the mother confirm that, adding: "she was a calm baby, a lazy child, always very quiet, in contrast to her little stepsister". Examinations showed a severely hypertrophic right ventricle, thickness of left ventricle in the upper normal limit, biatrial dilatation, the right one bigger, pericardial effusion, no fibrosis o signs of deposit disease, inferior vena cava and suprahepatic veins dilatation, impaired exercise capacity, mild postcapilar pulmonary hypertension but very elevated left ventricle filling pressures without dip-plateau as well as very high NT-proBNP (2.121 pg/mL). Abdominal echography was unremarkable. Among genes tested, there was a mutation in FLNC gen (p.His2476Arg), a VUS with potential association with disease because missense variants in the same functional region had been associated to hypertrophic/restrictive myocardiopathy, with high rate of heart failure during follow-up. Images from exams (EKG, ETT, MRI, CC) are shown in image.
Conclusions: Although a VUS is present, it is potentially the cause of the phenotype, taking into account mutations in similar region. In the future, investigation of relatives and reported cases in other patients and families would clarify its possible pathogenicity. Collaboration between clinical cardiologists, cardiologists specialized in familiar cardiopathies and geneticists is mandatory to face these cases.

Keywords: VUS, FLMN, biventricular Hypertrophy, restrictive myocardiopathy

Image



1- EKG/ 2- LV and PCP pressures/3-TTE: a: apical 4 chambers; b: paraesternal long axis; c: paraesternal short axis; d: subcostal/4-MRI: a: apical 4 chambers; b: paraesternal short axis; c: great vessels LV: left ventricle, PCP: pulmonary capillary pressure, RV: right ventricle, IVC: inferior vena cava, SH: suprahepatic veins (yellow arrows), Ao: aorta, PA: pulmonary artery

Adult Congenital Heart Disease

PP-026

Vitamin D insufficiency correlates with impaired exercise capacity in adults with congenital heart disease

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Background and Aim: 25-hydroxy vitamin D (25(OH)vitD) is important in the normal function of multiple organs and supports exercise performance and vascular health. VitD deficiency is highly prevalent in the general population. Exercise capacity is reduced in patients with congenital heart disease (CHD). However, literature

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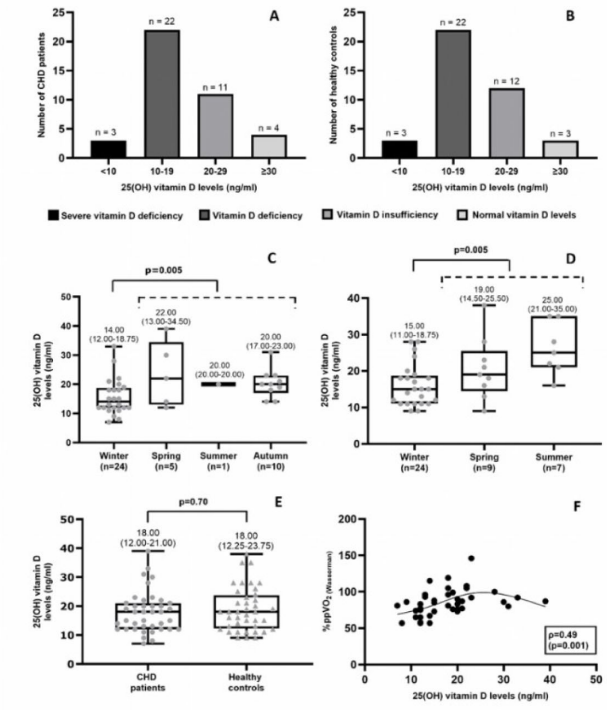
on vitD in CHD is limited. Lower vitD levels have been reported in children with CHD and are associated with skeletal muscle deficits in Fontan patients. The aim of this study was to analyze vitD levels in adult CHD patients as compared with healthy controls, and to examine associations with peripheral arterial function and exercise capacity.
Method: Forty adults with CHD (20 males, median (Q1-Q3) age 30 (24-42) years) performed a maximal cardiopulmonary exercise testing (RER≥1.10). Levels of vitD were compared with 40 age- and gender-matched healthy controls. In addition, peripheral arterial function was measured by arterial tonometry at the fingertip in all CHD patients. Associations between vitD levels and percent-predicted peak oxygen consumption (%ppVO2) and reactive hyperemia index (RHI) were investigated in CHD.
Results: Thirty-six of 40 CHD patients (90.0%) and 37 of 40 healthy controls (92.5%) had reduced vitD levels (<30 ng/ml)(Figures A-B). A seasonal effect in vitD values was visible in the two groups (p=0.005 for both)(Figures C-D). VitD levels did not significantly differ between the patients and healthy controls (Figure E). Exercise capacity, but not peripheral arterial function, significantly correlated with vitD levels in adults with CHD (Figure F). Even after adjusting for season as a confounding factor, the relationship between vitD and exercise capacity remained statistically significant (p=0.018).
Conclusions: Serum levels of vitD did not significantly differ between adults with CHD and healthy controls. Nevertheless, vitD insufficiency is prevalent in adults with CHD and correlates with impaired exercise capacity. It remains to be elucidated whether vitD supplementation confers additional benefits to exercise in case of vitD insufficiency in CHD.

Keywords: Adults with congenital heart disease, Congenital heart defects, Vitamin D, Exercise capacity

Distribution of vitD levels and seasonal effect in vitD levels in CHD (A,C) and healthy controls (B,D); comparison of vitD levels between CHD and healthy controls (E); and correlation of vitD with exercise capacity in CHD (F)



Data are presented as median (Q1-Q3), whisker plots indicate min-max; Abbreviations: %ppVO2 = percent-predicted peak oxygen consumption (reference values following

PP-027
Overweight and obesity in the adult congenital heart disease population

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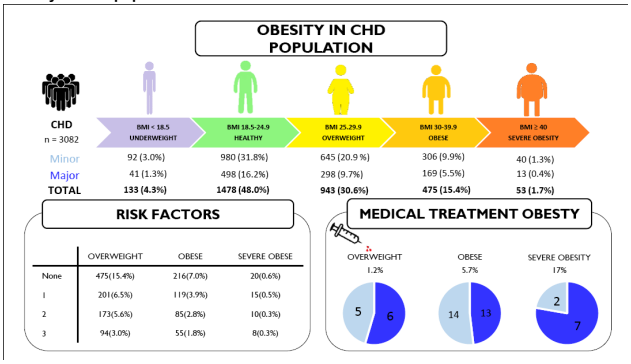
Background and Aim: Overweight and obesity is an increasing concern worldwide and associated with increased cardiovascular morbidity and mortality. ACHD patients are, due to improved survival, ageing and at risk for premature acquired heart disease. Overweight/obesity is one of the contributing risk factors to acquired heart disease. The aim of this study was to describe the prevalence of overweight/obesity among adult CHD patients in the eastern part of Denmark, describe the prevalence of overweight/obesity in major and minor CHD patients and finally look at how many patients are medically treated for overweight/obesity.
Method: A single center retrospective cross-sectional study. All danish ACHD patients seen between 2021 and 2023 at Rigshospitalet, Copenhagen, Denmark were included if BMI was present.
Results: A total of 3.082 ACHD patients were included in the study, and of those 33% had major CHD. Almost half of the ACHD patients (48%) had an increased BMI with the distribution of overweight 31%, obesity 17%, and severe obesity 1,7%. The distribution between minor and major CHD patients were equal. AVSD (65/110), and Ebsteins anomaly (20/37) were among the CHD diagnoses with the highest occurrence of overweight/obesity. A total of 49 patients were on weight reducing medication and the distribution was 1.2% of the patients with overweight, 6% with obesity and 17% with severe obesity
Conclusions: Overweight and obesity among CHD patients is common and a concern with the increased risk of premature acquired heart disease. There is no difference between major and minor CHD patients and overweight/obesity. Despite a high percentage of CHD patients with overweight/obesity very few patients were on medical therapy for overweight. The overweight/obesity in a young population calls for action in regards of preventive cardiology to reduce the risk of premature aquired heart disease in CHD patients.

Keywords: adult congenital heart disease, body mass index, aquired heart disease, overweight, obesity

Obesity in CHD population



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Cardiac Dysrhythmias and Electrophysiology

PP-028
Left cardiac sympathetic denervation in 12-year-old boy with cardiac ryanodine receptor 2 (RYR2) EXON 3 deletion syndrome

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Background and Aim: Cardiac ryanodine receptor 2 (RYR2) exon 3 deletion syndrome is a rare cardiac ryanodinopathy similar to catecholaminergic polymorphic ventricular tachycardia. This syndrome can present with a variable tachycardia-bradycardia phenotype and life-threatening exercise- and stress-induced ventricular arrhythmias. Implementation of antiarrhythmic medication is challenging due to the frequent coexistence of sinus node dysfunction and atrioventricular node conduction disorders.
Method: Case report.
Results: A 12-year-old boy experienced ventricular fibrillation while heavy drumming in a fanfare corps. After ten minutes of cardiopulmonary resuscitation and subsequent defibrillation, the arrhythmia was terminated. In the hospital, the first *ECG* showed sinus rhythm with recurrent bigeminy alternated by a bradycardic atrial escape rhythm and a QTc prolongation of 480 ms, which resolved in the following days. *Echocardiography* and *cardiac MRI* were normal except for initially slightly reduced function. *Cardiac catheterization* revealed normal coronary arteries and endomyocardial biopsies without inflammation. During vomiting, a non-sustained polymorphic ventricular tachycardia (VT) occurred. On stimulation with isoproterenol and later during exercise treadmill testing, a stepwise deterioration in ventricular arrhythmia score (VAS) from 0 to VAS 4 non-sustained VT occurred. Holter ECG showed bradycardia with sinus node dysfunction. *Family history* revealed repeated resuscitations and pacemaker dependency in the maternal great-grandmother, and sudden cardiac death of the maternal grandmother at the age of 38 years after pacemaker placement due to sinoatrial and atrioventricular block. *Genetic testing* revealed the exon-3 deletion in the RYR2 gene NC_000001.11(NM_001035.2):c.(168+1_169-1)_(273+1_274-1)del. Treatment with nadolol and flecainide was initiated and stepwise up-titrated to 0.8 and 3.9 mg/kg BW/d, respectively. Due to bradycardia, further increase was impossible although exercise treadmill testing repeatedly showed the occurrence of bigeminy at a heart rate of 120 bpm. Hence, left cardiac sympathetic denervation from the lower half of the stellate ganglion to thoracic ganglia T4 was performed. Afterwards, treadmill testing showed only VAS 1 with isolated premature ventricular contractions.
Conclusions: Bradycardia may impede sufficient antiarrhythmic therapy with beta-blockers and class Ic antiarrhythmics in RYR2 exon 3 deletion syndrome. Left cardiac sympathetic denervation may be a feasible, safe, and effective treatment escalation also in children.

Keywords: RYR2 exon-3 deletion syndrome, cardiac ryanodinopathy, catecholaminergic polymorphic ventricular tachycardia, left cardiac sympathetic denervation, pediatrics.

PP-029
Wolf-parkinson-white syndrome and 3 accessory pathways. ablation guided by 3D transoesophageal echocardiography and electro-anatomic navigation system

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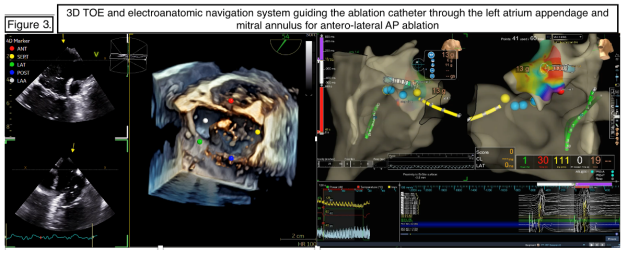
Background and Aim: Wolf-Parkinson-White Syndrome (WPW) is the most common cause of ventricular pre-excitation in paediatrics. In 2-10% of the cases we might find more than one accessory pathway (AP), happening mostly in congenital heart disease patients. We want to describe the strategy we used in a challenging case of a teenager with structurally normal heart and a total of 3 APs, where the electrophysiology study (EPS) was guided by a 3D transoesophageal echocardiography (TOE) as well as electro-anatomic navigation system.
Method: Descriptive case report of a challenging and unusual case of 3 APs and its strategy for a successful ablation.
Results: 15-year-old male diagnosed with WPW Syndrome. He had been treated with anti-arrhythmic drugs as a neonate for up to 1 year. Later in life, due to recurrence of palpitations and evidence of ventricular pre-excitation, he was referred to our centre for EPS (Figure 1). He had a normal structural heart. The exercise test showed ventricular pre-excitation up to a heart rate of 203 bpm. The EPS showed 3 different APs, each one with a different ventricular pre-excitation morphology in the 12 lead ECG (Figure 2) that we could successfully ablate. The first AP was a Parahisian AP, performed with cryoablation via internal jugular vein. The second AP was a postero-lateral left AP. The third AP was an antero-lateral left AP located between the left atrial appendage and the mitral annulus. Both left APs where ablated with radiofrequency with a transseptal approach. Both the electro-anatomic navigation system and the use of TOE with a 3D mini probe 9VT were essential for the correct mobilisation of the catheter

Figure 3



3D TOE and electro-anatomic navigation system guiding the ablation catheter through the left atrium appendage and mitral annulus for antero-lateral AP ablation

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ablation through the mitral annulus as well as for the guidance of the transseptal puncture (Figure 3).
Conclusions: The existence of 2 APs is a common finding during an EPS. Otherwise, having up to 3 APs in a structural normal heart is a very uncommon finding. The use of TOE with 3D mini probe and electro-anatomic navigation system was the key to a successful ablation in such a challenging case.

Keywords: Wolf-Parkinson-White Syndrome, accessory pathway, electrophysiology study, ablation, 3D TOE, electro-anatomic navigation system

PP-030

Arrhythmogenic challenges in pediatric TOF: A comprehensive case study on pulmonary valve replacement and electrophysiological considerations

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Background and Aim: Pediatric patients with Tetralogy of Fallot (TOF) undergoing pulmonary valve replacement may be at risk of life-threatening arrhythmias. An 18-year-old patient with Di George's syndrome, TOF, and a history of corrective surgery in infancy, exhibiting chronic pulmonary insufficiency and RV dilatation. Following pulmonary valve replacement through catheterization, the patient experienced self-limited episodes of ventricular bigeminy/trigeminy, culminating in a life-threatening episode of ventricular fibrillation. Diagnostic catheterization excluded mechanical compression of the valve. Despite a negative electrophysiological study (EPS) and after a multidisciplinary assessment, implantable cardioverter-defibrillator (ICD) implantation was performed for secondary prevention of sudden cardiac death.
Method: Only two articles from recent studies by French research groups evaluate the significance of EPS before pulmonary valve replacement, however in adult patients with TOF.
Results: Given the absence of a Pediatric protocol for these patients, we have developed a protocol addressing the potential management of this specific population (Table 1):

1. When a pulmonary valve replacement (either percutaneous or surgical) is considered, some preliminary tests should be assessed such as ECG, Holter-ECG, Ergomespirometry, Echocardiography, Cardiac Angio-CT/Angio MRI and then complete the evaluation with an EPS.
2. If the EPS is positive, ablation can be planned before valve replacement. If this ablation procedure is not effective, the indication for pulmonary valve replacement should be surgical, enabling the potential for concurrent cryoablation during the same procedure.
3. If the EPS is negative, pulmonary valve replacement can be performed.
4. Two months after valve replacement, another EPS can confirm whether the patient does not require further procedures or whether ablation or ICD implantation is necessary.

Conclusions: This protocol proposes a comprehensive preoperative assessment, emphasizing the role of EPS and complementary studies to identify and manage potential arrhythmogenic substrates. EPS becomes a valuable tool in identifying high-risk patients, offering opportunities for targeted ablation, and aiding decision-making regarding ICD implantation. This comprehensive approach ensures a thorough evaluation of pediatric TOF patients, potentially improving outcomes and reducing the incidence of life-threatening arrhythmias post-pulmonary valve replacement. The proposed protocol serves as a valuable guideline for clinicians managing similar cases, emphasizing the importance of tailored evaluations to optimize patient outcomes in pediatric TOF populations.

Keywords: Tetralogy of Fallot, EP study, Pulmonary valve replacement, Arrhythmia, Ventricular fibrillation, ICD

PP-031

Cardiac magnetic resonance abnormalities in children, adolescents and young adults with idiopathic ventricular arrhythmia and normal echocardiogram

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Background and Aim: Ventricular arrhythmia (VA) of unknown etiology in pediatric and young adult patients with normal echocardiogram and preserved left ventricular function presents a significant diagnostic challenge. The aim of this study was to determine the prevalence and predictors of cardiac magnetic resonance (CMR) abnormalities in children, adolescents and young adults with idiopathic VA.
Method: 89 patients (median age 17 years, age range 10 to 44 years) from a prospectively kept, electronic registry of a tertiary center, with VA of unknown origin and normal echocardiographic finding, underwent CMR (1.5 Tesla) with late gadolinium enhancement (LGE) during 2015-2022. Exercise stress tests and 24h Holter ECG monitoring were done prior to CMR. Patients were stratified into two groups based on the presence of CMR abnormalities. Binary logistic regression was used to determine independent predictors of pathological CMR finding.

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Results: Symptoms were present in 39% (n=35), including chest pain in 7% (n=6), syncope at rest in 11% (n=10), exercise-related syncope in 7% (n=6), and palpitations in 14% (n=13). Exercise related symptoms were present in 16% (n=14). Abnormal CMR was present in 35% (n=31), while LGE was found in 7% (n=6) of patients. Mean premature ventricular complex (PVC) burden was 15.2±14.3%. Non-sustained ventricular tachycardia (VT) was present in 29% (n=26), while sustained VT was present in 15% (n=13). Male sex (odds ratio (OR) 4.16, 95% confidence interval (CI): 1.48-11.65, p<0.05), exercise-related symptoms (OR 4.33, 95% CI: 1.30-14.41), polymorphic PVC (OR 3.68, 95% CI: 1.08-12.48, p<0.05), PVC burden >15% (OR 5.18, 95% CI: 1.36-19.70, p<0.05) were significantly associated with CMR abnormalities. In multivariate analysis, exercise-related symptoms (OR 3.95, 95% CI: 1.02-15.24) and PVC burden >15% (OR 3.54, 95% CI: 1.12-11.16, p<0.05) remained significantly associated with CMR abnormalities. There was no statistically significant association between abnormal exercise stress test and CMR findings.
Conclusions: In our cohort of children, adolescents and young adults with idiopathic VA and normal echocardiogram, the abnormal CMR finding was present in more than third of patients. Exercise related symptoms and PVC burden >15% were significantly associated with abnormal CMR finding.

Keywords: extrasystoles, exertion-related symptoms, idiopathic, magnetic resonance

PP-032
Correlation between electrocardiographic phenotype and genotype in children with rasopathy syndromes

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Background and Aim: The RASopathy syndromes constitute a phenotypically similar group of malformation syndromes caused by dysregulation of the RAS/MAPK and PI3K/Akt pathways. Collectively, they are one of the largest groups of multiple congenital anomalies known, affecting approximately 1 in 1000 individuals. Hypertrophic cardiomyopathy (HCM) is the second most common cardiac abnormality found in the syndrome. Characteristic electrocardiographic (ECG) features are thought to be present in RASopathy-associated HCM (R-HCM), but this has never been systematically studied and its link with RASopathy type and genotype is still poorly understood. This study aims to determine the correlation between ECG findings on RASopathy-associated HCM patients and RASopathy syndrome type as well as genotype.
Method: Data from Great Ormond Street Hospital electronic health records of children (<18 years old), from the period of 1985 till 2023, presenting with RASopathy - associated HCM was collected. Patients included have baseline ECG available within a year of first date of assessment. Repeated and blinded systematic ECG assessment was carried out to reduce bias. Association between ECG phenotype and RASopathy type and genotype was analysed. Link between ECG and mortality was carried out using logistic regression.
Results: Eighty four patients were included in the study with median age 6 years (IQR 11-19.5). The majority of patients have Noonan syndrome (n=59, 70.2%), with the most common genetic variants being unknown (32.1%), PTPN11 (29.8%), RAF1 (13.1%), HRAS (8.3%) and, RIT1 (6.0%). Common ECG features identified were axis deviation (56.5%), left [LVH] (47.6%) or right ventricular hypertrophy [RVH] (52.4%), and conduction delay (45.2%). No significant association between ECG phenotype and RASopathy syndrome type as well as RASopathy genotype was found. No significant link between specific ECG phenotype and mortality was found.
Conclusions: Several ECG features common in R-HCM were identified, with some being unique to the syndrome such as extreme axis deviation. No significant association between ECG phenotype and RASopathy type as well as genotype was found. Possible reason for results are due to the small study sample size. Larger multi-centred studies are required to confirm these findings.

Keywords: RASopathy syndromes, Noonan syndrome, electrocardiography, genotype

PP-033
Arrhythmic mitral valve prolapse – prevalence, characteristics and high-risk features in children and young adults

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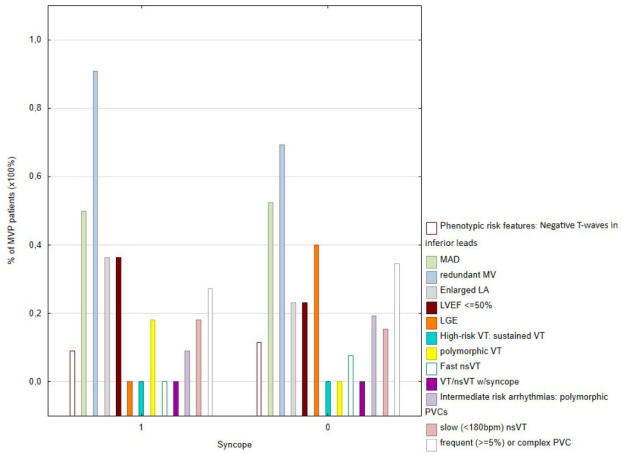
Background and Aim: Arrhythmic mitral valve prolapse (AMVP) is a rare, yet potentially fatal syndrome. Recently, high-risk markers for malignant arrhythmias (As) have been identified in adults with mitral valve prolapse (MVP) and risk stratification scheme have been proposed. Little is known about prevalence to AMVP, characteristics of the syndrome and risk markers in children and young adults.
Method: A computerized database of high-volume, tertiary invasive pediatric cardiology/congenital heart defects department was searched to identify all patients discharged with an ICD-10 code I34.1 (MVP) between 2018 and 2023 (6y). Patients characteristics, including arrhythmic risk factors, diagnosis of AMVP and last survival were noted.
Results: Of the 9378 patients hospitalized, 37 had MVP (0.4%, median age 14y [IQR=5], 35% male, 30% with syncope, 16% pre-syncope), including 8 (22% of MVP) with AMVP (frequent

Phenotypic and arrhythmic risk factors in MVP population with and without syncope



MAD - mitral annulus disjunction, MV - mitral valve leaflets, LA - left atrium, LVEF - left ventricular ejection fraction, LGE - late gadolinium enhancement in MRI, VT - ventricular tachycardia, ns - non-sustained, PVC - premature ventricular complex

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[≥5% PVC burden] or complex As [nsVT, VT, or VF]). One patient (2.7%) had ICD implanted (MVP and genetically confirmed LQTS2), one had loop recorder implanted. 94% had at last 1 phenotypic risk factor, 11% at least 1 high-risk and 32% intermediate-risk arrhythmic factor. 30% had one, 16% two, 27% had three and 21% >3 risk factors. During median 2y (IQR 3), all were alive.
Conclusions: One in 250 patients admitted to tertiary invasive pediatric cardiology department has MVP, among those with MVP, one in five presents with AMVP. Although risk factors are common, outcomes among young AMVP patients seem to be good. Overlapping with primary electrical disease may flag a worse prognosis. Identification of new risk factors for children is warranted.

Keywords: mitral valve prolapse, arrhythmia, sudden cardiac death, ventricular tachycardia, prevention, risk stratification

PP-034
QRS fragmentation and ventricular arrhythmias inducibility in tetralogy of fallot

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Background and Aim: Repaired Tetralogy of Fallot (TOF) patients show long-term risk of ventricular arrythmias and sudden cardiac death. Currently, risk assessment is performed through noninvasive/invasive tests including symptoms, ECG (QRS duration and QRS fragmentation, QRSf), imaging tools (echocardiography, cardiac magnetic resonance CMR) and intracardiac electrophysiology study (EPS). However, predicting the risk is challenging. QRSf is related to right ventricle (RV) myocardial fibrosis and dilatation, it appears to be a mortality predictor. The relationship between QRSf, RV dilatation and ventricular arrhythmia inducibility at EPS is unknown.
The aim of the study is to investigate the relationship between QRSf in operated TOF patients and arrythmia inducibility at EPS.
Method: This is a retrospective study. We analyzed the 12-lead ECG in every patient with RV dilatation (RV Vol>130 ml/m2) who underwent EPS. QRSf was defined as: QRS with two additional notches in the R/S wave in ≥ 2 contiguous leads on a 12-lead ECG, not related to the bundle branch block. QRSf was divided into none, mild (≤3 leads), moderate (4 leads) or severe (≥5 leads). QRSf was correlated with RV 3D volume at echocardiography and EPS findings. Data are reported as: mean +-SD.
Results: 50 adolescent and adult patients with repaired TOF (66% males), with age at EPS 20.2+-8.6 years, BSA 1.6+-0.2 m2, QRS duration 144.2+-27.8 ms, RV Vol 161.7+-40.6 ml/m2, were included. QRS fragmentation was present in 40 (80%) patients, who also showed RV dilatation (164±26 ml/m2) and QRS duration 149+-24 ms. Five (10%) patients had moderate QRSf and 35 (70%) patients severe QRSf with RV Vol 161±27 ml/m2. Ventricular tachycardia was induced at EPS in 10 (35%) patients who were also symptomatic for palpitation; 90% of them showed severe QRSf with RV dilatation (132.9±25 ml/m2), and one patient (10%) showed moderate QRSf with RV dilatation (RV Vol 163 ml/m2).
Conclusions: TOF patients with RV dilatation and severe QRSf showed more ventricular arrhythmia inducibility at EPS. Therefore, severe QRSf seems a marker of arrythmia risk.

Keywords: QRS fragmentation, ventricular arrythmias, intracardiac electrophysiology study

PP-035
Dual mechanism (atrial flutter and AVRT) in the setting of wolff-parkinson-white syndrome leading to sustained foetal tachycardia

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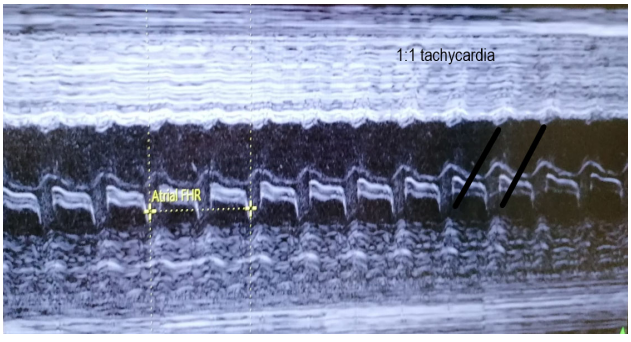
Background and Aim: To present a fetus with dual mechanism tachycardia in the setting of WPW syndrome
Method: A 34 w + 0 d pregnant lady was referred for foetal SVT. The initial scan showed SVT throughout scan at 220 - 230 bpm, no hydrops or AV valve regurgitations, normal structure. The M mode trace and SVC Aorta doppler showed 1: 1 atrioventricular conduction ratio tachycardia with retrograde P wave likely AVRT (1), (2). Mother was started on Digoxin. Subsequent scans showed persistent tachycardia with episodes of flutter and AVRT (3). Digoxin was increased and Flecainide was added but tachycardia did not slow down or convert to sinus rhythm- digoxin 250mcg TDS and Flecainide 100mg BD.
. Baby was born in stable condition in 39 weeks with wide complex SVT with heart rate of 220(4). Adenosine was applied 100,200,300 mcg/kg which slow down the tachycardia temporarily and revealed atrial flutter (5). Baby was intubated and received DC cardioversion with 1 joule/kg and converted to sinus rhythm (6). The 12 lead ECG after DC cardioversion showed sinus rhythm, WPW (Wolff-Parkinson-White) (7).
Baby was started on Flecainide 1 mg /kg TDS and Flecainide level was taken as baby was treated antenatally with Flecainide. Baby remained stable in sinus rhythm but 24 Holter fitted next day revealed Very frequent runs of SVT - Longest run lasting 136 beats at 233 bpm with 1:1 short R- P tachycardia likely AVRT (8).
Results: The dose of Flecainide was increased to 2mg/kg /TDS and baby remained stable ever since with no breakthrough episodes and no preexcitation on the ECG.
Conclusions: Atrial flutter (AFL) is the second most common fetal tachyarrhythmia,1 1 which accounts for 25% of fetal tachyarrhythmias (1). The electrophysiologic mechanism of tachycardia is intra-atrial macro-re-entry, like adult AFL. There are a few reports in the literature that describe the association of fetal atrial flutter with AVRT in the setting WPW syndrome (2). Our case revealed this rare combination and showed that careful antenatal and postnatal

AVRT and flutter



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rhythm analyses is very important as it helped the medical management and justified a long-term postnatal antiarrhythmic treatment.

Keywords: Fetal CVT, atrial flutter, AVRT (atrioventricular reciprocating tachycardia), WPW(WOLFF-PARKINSON-WHITE SYNDROME)

PP-037

Radiofrequency ablation (RFA) of post-infarct ventricular ectopy (VE) in a child

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Background and Aim: We report a case of RFA of post-infract scar related VE in a child with repaired congenital heart disease (CHD). To best of our knowledge this has not been reported in a child.
Method: A 10 year old boy (weight – 25 kg) presented with frequent monomorphic VE'S. He had undergone cardiac surgery during neonatal period after postnatal diagnosis of transposition of great arteries, ventricular septal defect (VSD) and coarctation of the aorta (CoA). He underwent an arterial switch operation, CoA repair and VSD closure which was complicated by intraoperative coronary complications needing subsequent revision of his coronary artery surgery. Subsequent cardiac catheter showed infracted left ventricular (LV) inferior and septal wall with minor narrowing of the circumflex artery. This resulted in only mildly reduced cardiac function until he developed high VE burden (43%) leading to severe impairement of LV function. CT/MRI scanning confirmed unobstructed coronaries, reduced systolic function (EF 35%) with regional wall motion abnormality and full thickness scarring in mid inferior wall of LV. The VE'S were monomorphic with right bundle branch block (RBBB) and superior axis morphology. There was no improvement with medical therapy so RFA was planned.
Results: The electrophysiology study (EPS) was performed under GA via left femoral vein as right was occluded. Electroanatomical mapping (CARTO, Biosense Webster) with activation mapping demonstrated scarring around the septal and inferior area and earliest signal in the midseptal area. Initial RFA was performed on right side of septum with no change. Later access to LV was obtained through PFO and on mapping early signals were obtained at LV anterior septum. RFA using irrigated tip ablation catheter led to abolition of ventricular ectopy. Post procedure holter showed ectopy burden 1% and LVEF improved to 45%.
Conclusions: VE in setting of surgical scars in CHD is not uncommon. However, presentation at very young age similar to post infact scar related VE/VT in adults is uncommon. This is the first report of a successful RFA of a post infarct VE in a child and highlights that RFA can be considered early in the management.

Keywords: Ventricular ectopy, congenital heart disease, Radiofrequency ablation

PP-038

The morbidity and mortality of patients with congenital complete heart block treated with permanent pacemaker implantation: Single centre experience

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Background and Aim: There are limited studies assessing the long-term outcomes of patients with congenital complete atrioventricular block (CCHB) treated with a permanent pacemaker. Aim To undertake a retrospective study to determine the long-term incidence of cardiac morbidity and mortality in subjects with CCHB treated with pacemaker implantation and identify associated risk factors.
Method: A retrospective cohort analysis of patients treated with a permanent pacemaker for CCHB at East Midlands Congenital Heart Centre between 2010 and 2021 was performed. The primary outcome was a composite of death, left ventricular systolic dysfunction, heart failure, or need for upgrade to cardiac resynchronization therapy (CRT). Statistical analysis was performed using SPSS (median and interquartile range (IQR) for demographics and T-student test, Spearman's correlation and Chi-square test for comparisons). Statistical significance was set at $P<0.05$.
Results: Forty nine subjects (59% female; median age at last visit 21 years) were included. The demographics, clinical characteristics and clinical outcomes are presented in Table 1. One in five patients also had a co-existing congenital heart defect with seven out of nineteen having congenitally corrected transposition of great arteries (ccTGA). Five patients (10%) reached the primary outcome with the majority of these (four out of five) having an underlying congenital heart defect (CHD). One death occurred due to heart failure at age five in a patient with ccTGA. A significant

Figure 1: Demographics, clinical characteristics and clinical outcomes in patients with congenital complete heart block

Demographics and clinical characteristics		
	Number of patients or median value	IQR
Patients	49	
Current age (years)	21	15
Female	29 (59%)	
Antibody positive	15 (31%)	
Anti-Ro	11 (22%)	
Anti-LA	5 (10%)	
Negative	8 (16%)	
Unknown status	26 (53%)	
Congenital heart defect	19 (19%)	
Age of diagnosis (years)	0	1.4
Temporary pacing	3 (6%)	
Age at implantation (years)	1.5	10
Duration of epicardial pacing (years)	11	15
Duration of pacing (years)	14	15
Clinical outcomes		
Heart failure	5 (10%)	
Systolic dysfunction		
Mild	15 (31%)	
Moderate	4 (8%)	
Severe	1 (2%)	
Upgrade to CRT	2 (4%)	
Lead fracture	7 (14%)	
Lead dislodgement	2 (4%)	
Pacemaker malfunction	1 (2%)	
Infection	6 (12%)	
Number of implants	2 (4%)	1
QRS duration (msec)	148	44
Death	1 (2%)	

Figure 1: Demographics, clinical characteristics and clinical outcomes in patients with congenital complete heart block. CRT: cardiac resynchronisation therapy. Numbers presented median and interquartile range or numbers of patients and percentage as appropriate.

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association was found between the incidence of heart failure and the presence of maternal antibodies and the presence of CHD (Pearson Chi-square: 0.043 and 0.047, respectively). There was a significant negative correlation between QRS duration and fractional shortening (Spearman's ρ=-0.418, P=0.024) and ejection fraction (Spearman's ρ:-0.691, P=0.013).
Conclusions: Our cohort of patients had a lower incidence of heart failure and mortality compared to other published studies. Patients with CHD and positive maternal antibodies were at greater risk for the development of heart failure and may represent a group who should be considered for conduction system pacing at an early stage.

Keywords: Congenital complete heart block, heart failure, pacemaker

PP-039
Pulsed electrical field ablation cured scar-related right ventricular tachycardia after failed radiofrequency ablation in congenital heart disease
Christian Scheckenbach[1], Johannes Nordmeyer[1], Malte Kranert[2], David Heinzmann[2], Meinrad Gawaz[2] and Jürgen Schreieck[2]
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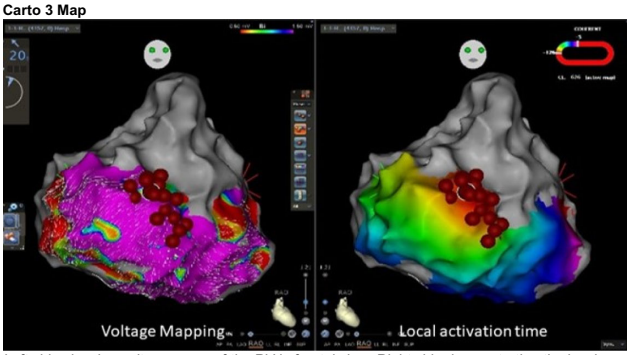
Background and Aim: Ventricular tachycardia (VT) in the context of congenital heart disease (CHD) often differs from that of post-ischemic patients in adult cardiology. VT in CHD can occur in younger age groups and often requires specific approaches due to prior surgical procedures and special hemodynamic conditions.
Method: case presentation
Results: We report the case of a 30-year-old CHD patient with first episode of VT requiring electric cardioversion. He previously had surgical repair of tetralogy of Fallot in first year of life. He subsequently underwent bioprosthethic pulmonary valve replacement and plication of the right ventricle (RV) at the age of 23 years because of severe pulmonary regurgitation and RV dilatation. After the reported first episode of VT he was referred for evaluation of internal cardioverter-defibrillator (ICD) implantation and electrophysiologic study. A recent cardiac MRI study revealed an RV enddiastolic volume of 130ml/m2 and reduced systolic RV function (ejection fraction 11%). The voltage map of the RV showed a low voltage area at the lateral site of the RV. After inducing a hemodynamically stable VT the RV activation mapping showed earliest activation within the lateral scar, however, a part of the cycle length was missing, pointing out to a concealed intra-/epicardial conduction. Therefore, irrigated radiofrequency (RF) ablation up to 40 Watt was applied at the site of earliest signals, however, the VT still persisted. To achieve deeper impact of ablation energy facing scare tissue because of prior surgery we switched to pulsed electrical field ablation (PEF) ablation via a contact force-controlled RF catheter (Centauri, Galvanize). After a few PEF applications using the monopolar 25A energy pulses of the system at the site where RF failed, the VT converted to sinus rhythm and could not be induced any longer. Since aggressive ventricular stimulation induced another different, hemodynamically unstable VT and due to suppressed RV function ICD implantation was planned.
Conclusions: Pulsed electrical field ablation as a new type of non-thermal ablation technique is known to prevent adverse side effects to noncardiac tissue like the phrenic nerve or esophagus. In this case, we demonstrate the feasibility and usefulness of PEF for ablation of post-surgery, scar-related VT in adult CHD.



Left side showing voltage range of the RV in frontal view. Right side demonstrating the local activation time of the induced ventricular tachycardia.

Keywords: Arryhthmias, ventricular tachycardia, electrophysiology, pulsed electrical field ablation, congenital heart disease

PP-040
Longevity of four different epicardial pacemaker leads in neonates, infants and young adolescents
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[1]Pediatric Heart Center Vienna, Department of Pediatric Cardiology, University Children's Hospital, Medical University Vienna, Austria; [2]Pediatric Heart Center Vienna, University Hospital of Cardiac Surgery, Medical University Vienna, Austria

Background and Aim: Internal pacemaker therapy was introduced in the mid-1950s, initially with an epicardial approach. Since then, therapy for "grown-up" patients has orientated towards endovascular placement with an increasingly wide range of leads. However, for young children and patients with univentricular circulations, the epicardial approach remains the only option, albeit with a limited lead portfolio. Here we present our analysis of follow-up data from four epicardial leads routinely used in childhood.
Method: In this retrospective monocentric study, data from patients ≤18 years at lead implantation between 7/2004 and 5/2021 were evaluated for pacing thresholds and incidence of revision procedures. The implanted electrodes were the passive-fixating, steroid-eluting Medtronic© 4965 (unipolar) or 4968 (bipolar) or the active-fixating, bipolar Greatbatch Medical© 511211/2 (non-steroid-eluting) or 1084T (steroid-eluting). Only patients with a follow-up ≥1 year were included.
Results: 144 leads were implanted in 83 patients (48 males with 86 and 35 females with 58 leads) at an average age of 3.02±3.76 years. Of these, 108 leads were implanted as first-time and 36 as re-do interventions. In 36 patients lead position was atrial, 59 RV, 27 LV, and 22 univentricular. The initial pacing threshold of 0.96 ±0.51V (mean) over 0.44±0.16ms increased 2.2±2.9-fold during the first year and up to 5.5±12.7-fold by the end of follow-up (mean 59.1±49.0 months). In 47 leads, the pacing threshold was persistently elevated >1V after 27.2±32.4 months (mean) after implantation, with an increase rate of 14.0±19.4 times. Of these, 5/22 were a Medtronic© 4965 (22.7%), 27/88 a type 4968 (31.4%), 9/26 a Greatbatch Medical© 1084T (34.6%) and 6/8 a 511211/2 (75%). This proportion was significantly higher for the non-steroid-eluting electrode than for the other three (p=0.009, p=0.013 and p=0.044, respectively).
Conclusions: Greatbatch Medical© active-fixing leads are a viable alternative to Medtronic© passive-fixating electrodes, particularly for ventricular stimulation. In terms of pacing thresholds and finally

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longevity, the non-steroid-eluting 51121/2 is significantly inferior. As the company has withdrawn the steroid-eluting 1084T from the market in 2021 and the unipolar 4965 is hardly used anymore, the limited selection of epicardial leads is now alarming.

Keywords: epicardial pacing, lead longevity, childhood

PP-041

Cardiac resynchronisation therapy (CRT) by right ventricular pacing in a child with complex congenital heart disease: A case report

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Background and Aim: Cardiac resynchronization therapy (CRT) has attained established status in adult practice and in selected cohort of paediatric patients. The results of CRT in children with congenital heart disease (CHD) remain variable with the cohort of patients with left bundle branch block (LBBB) where right ventricle (RV) is chronically paced being the most likely target. The data regarding CRT in setting of right bundle branch block (RBBB) remains rare. We present a case of child with CHD and postoperative complete heart block (CHB) who was paced epicardially from left ventricle (LV) leading to RBBB and was resynchronised by RV pacing through the transvenous route. AIM: To highlight a rare case of CRT by hybrid approach through pacing RV rather than LV.
Method: Retrospective case review.
Results: A child with CHD (Dextrocardia, pulmonary atresia, multiple ventricular septal defects (VSD's) and bilateral superior vena cava) underwent a Rastelli procedure at 17 months of age and developed postoperative heart block. She was paced using an epicardial dual chamber permanent pacemaker via left atrium and ventricle. She developed symptoms of heart failure over time with ejection fraction (EF) of 30% and BNP levels of more than 35,000 ng/L. The heart failure persisted despite transcatheter closure of some of the additional VSD's. The ECG's showed paced rhythm with RBBB and QRS duration of 160 ms.
At 31 months of age the patient underwent hybrid CRT implantation by adding a transvenous lead into RV via right subclavian vein and tunnelling this to a CRT generator (Medtronic, Solara CRT-P MRI SureScan) in the abdomen. The old LA and LV leads were maintained. The QRS duration post CRT was 110 ms.
Following CRT implantation she showed consistent improvement in symptoms of heart failure (BNP 268 ng/L) and now 2 years post CRT implantation EF remains at 50%
Conclusions: CRT with pacing from the RV in the context of heart failure and RBBB should be considered in children where other causes of heart failure have been ruled out.

Keywords: CRT, Congenital heart disease, dysarrhythmia

PP-042

Hereditary hcn4 mutations as a cause of fetal and postnatal inappropriate sinus tachycardia

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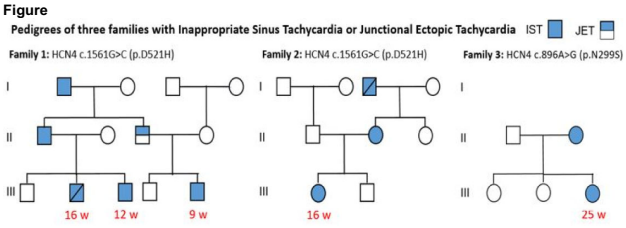
Background and Aim: Inappropriate sinus tachycardia (IST) is characterized by an unjustified fast sinus heart rate. Possible etiologies include a primary abnormality of the sinus node or an autonomic nervous system disorder. IST is an exceptional cause of persistent fetal tachycardia and hereditary forms have not been reported. We explored the possibility of genetic associations with fetal IST.
Method: Cardiac arrhythmia panel or whole-exome sequencing were used to identify candidate genes in three families with fetal IST (Figure). We then used a mutant mouse model and human pluripotent stem cell (hPSC)-derived cardiomyocytes to establish the pathogenicity of identified gene variants.
Results: In Family 1, a hereditary arrhythmia cause was first suspected after two consecutive fetuses of a healthy mother presented at 12 and 16 gestational weeks with a long VA tachycardia >200 bpm. A paternal sibling has junctional ectopic tachycardia (JET) and his second child was diagnosed with a fetal long tachycardia of 190 bpm at 9 weeks. In Family 2, the mother was treated with metoprolol for tachycardia-mediated cardiomyopathy when, at 16 weeks, her fetus presented with a long VA tachycardia of 205 bpm. Genetic testing revealed a novel heterozygous missense HCN4 variant (D521H) affecting 9 members in both families, including the four fetuses. Seven require chronic treatment with ivabradine or a β-blocker for heart rate control, while two of the carriers have died. When compared with wild-type controls (WT), faster heart rates were also observed in mutant mice (heterozygous: 763.3 ±35.3 bpm; homozygous: 807.2±38.7 bpm vs WT: 549.7±10.8 bpm; $P<0.001$) and hPSC-derived cardiomyocytes (heterozygous: 74±4 bpm vs WT: 62±5 bpm; $p=0.063$) segregating the D521H mutation. The above observation triggered genetic testing of a 12-year old girl in Family 3, who had been treated since before birth for "atrial ectopic tachycardia" but was eventually found to have IST at an average rate of 125 bpm and reduced ventricular function. She and her mother carry a different heterozygous HCN4

Figure



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variant (N299S) and are now treated with ivabradine for heart-rate control.
Conclusions: This is the first report of hereditary gain-of-function mutations in pacemaker HCN4 channels to cause persistent fetal sinus tachycardia.

Keywords: inappropriate sinus tachycardia, HCN4 channels, funny current, channelopathies, fetal

PP-043
Characteristics of patients who underwent placement of implantable cardioverters defibrillators in pediatric population in latvia 2014-2023

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Background and Aim: Pediatric population undergoing placement of implantable cardioverters defibrillators (ICD) is a heterogeneous group of channelopathies, cardiomyopathies and structural heart diseases. Children's Clinical University hospital (CCUH) is the only hospital in Latvia were pediatric patients with heart rhythm disorders, electronic rhythm devices and different cardiac conditions are treated. The aim of the study was to analyze indications and outcomes of all the patients <18 years old undergoing ICD implantation between 2014 and 2023 performed in CCUH.
Method: We reviewed the medical records and home monitoring results of all the patients <18 years old undergoing ICD implantation between 2014 and 2023 performed in the CCUH in Latvia
Results: Between 2014 and 2023, a total of 11 pediatric patients (55% boys) underwent ICD implantation in CCUH at mean age 12.8±5.5years.73% of indication were aborted sudden cardiac death (ventricular fibrillation with successful resuscitation), 18% VT due to hypertrophic cardiomyopathy (Danone disease), 9%(n=1) ventricular tachycardia (VT) due to gigantic fibroma of left ventricle, 82% of ICD were with home monitoring, 18% (n=2) subcutaneous ICD, all single chamber devices. All patients underwent genetic examination (55% positive), 36% of them with channelopathies. All patients received medications (82% beta blockers (BAB), 9% BAB with mexiletine, 9% flecainide. There was 1 case with purulent complications with reimplantation of ICD but no overall mortality (follow up time 4.4±2.8 years).
Conclusions: pediatric patients with ICD are heterogeneous group of diseases which are frequently genetically determined, complication rates are low, home monitoring possibilities allow more effective and safer follow up.

Keywords: Implantable cardioverters defibrillators, Pediatric cardiology

PP-044
Unavoidable anthracycline-application for acute lymphoblastic leukemia in a boy with long qt syndrome – a case report

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Background and Aim: Acute lymphoblastic leukemia (ALL) is the most common malignant cancer type in children. Its treatment requires anthracycline-application, which can disturb heart rhythm and electrocardiographic time intervals. Long QT syndrome (LQTS) is a rare disorder that affects the heart's repolarization potentially leading to severe cardiac arrhythmias and sudden cardiac death. In the following, we present a case of unavoidable anthracycline-application for ALL in a boy with LQTS.
Method: At time of admission, the 5-year-old boy was in a stable general condition. Staging revealed common-ALL. LQTS type 1 (variant of the KCNQ1/KCNE2 genes) was diagnosed molecularly. The boy took propranolol. No history of syncope was observed. The ECG at admission showed a sinus rhythm with a QTc of 490 msec at a heart rate of 90 bpm. Echocardiography revealed normal cardiac function but a small pericardial effusion at apical level. Electrolytes were in normal range.
Results: For the first anthracycline-application (30.4 mg daunorubicin/m² BSA over 60 min), the patient was transferred to our ICU enabling close cardiac monitoring and the possibility of prompt cardioversion. During and two hours after anthracycline-application a distinct elevation of QTc was demonstrated (QTc pre-application: 590 msec at a heart rate of 99 bpm, QTc during application: 617 msec at a heart rate of 95 bpm, QTc two hours post-application: 630 msec at a heart rate of 66 bpm). During his ICU stay, the patient showed a constant sinus rhythm. No adverse cardiovascular events were observed. Cardiac function was not altered. Twelve hours after anthracycline-application, QTc returned to 590 msec at a heart rate of 58 bpm. For the management of chemotherapy-induced nausea and vomiting, the patient received aprepitant, one of the few not QTc-altering antiemetic agents available. Further anthracycline-applications were uneventful.
Conclusions: The anthracycline-application led to a distinct QTc increase in a boy with LQTS. No severe cardiac arrhythmias were present during/after anthracycline-application. Nonetheless, a QTc of 630 msec due to anthracycline-application seen in this case can substantially increase the risk for severe cardiac arrhythmia, such as Torsade de pointes ventricular tachycardia. We therefore recommend close cardiac monitoring on an ICU in patients with QTc alterations who require anthracycline-treatment.

Keywords: Long QT Syndrome, Acute Lymphoblastic Leukemia, Anthracycline-Application

PP-045
Types of premature ventricular contractions in large a population-based sample of schoolchildren

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Background and Aim: It is essential to obtain accurate estimations of the actual prevalence of PVCs from large cohorts in order to interpret electrocardiographic abnormalities appropriately. The aim of our study was to provide data on the prevalence of premature

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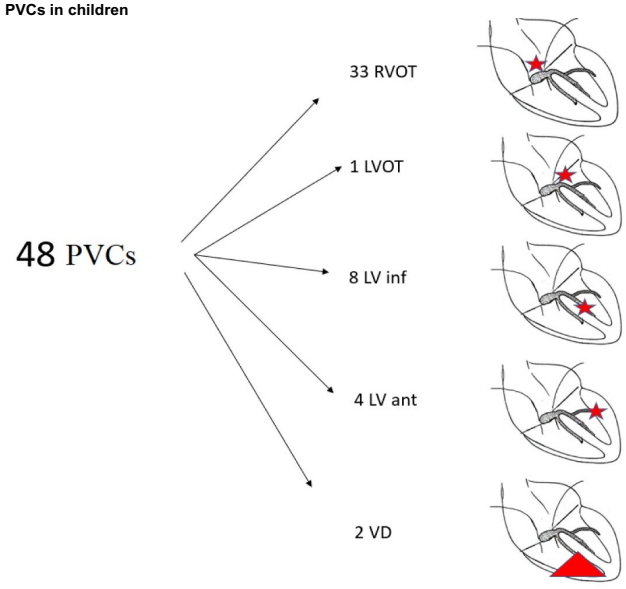
ventricular contractions in the general population of children from Romanian schools.
Method: We analysed prevalence of disturbances of rhythm in a population of 24,316 healthy, asymptomatic elementary school children, 48.3% males and 51.7% females, aged from 6 to 18 years. Children with known cardiac disease were excluded from the study. We analysed the prevalence of premature ventricular contractions using 6 seconds duration 12-lead ECG.
Results: The prevalence of PVCs was 0.19%. The morphology of PVCs was: RVOT in 33 children, LVOT in 1 child, posterior fascicular LV in 8 children, anterior fascicular in 4 children, RV inferior in 2 children.
Conclusions: Most children from our cohort (95.8%) had an outflow tract or fascicular morphology of PVCs.

Keywords: premature ventricular contractions, outflow, fascicular, children



PP-046
Atrioventricular reentrant tachycardia induced by fever due to acute cholecystitis and septic shock unmasked intermittent WPW syndrome

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Background and Aim: The difficulty of diagnosis and treatment of supraventricular tachyarrhythmias in children are challenging in pediatric practice. We want to present a case of tachycardia with narrow QRS complexes, that later unmasked an intermittent WPW syndrome in a febrile patient with acute cholecystitis and septic shock
Method: A 4 years old female was admitted to the ED for: tachycardia with narrow QRS complexes, fever for 2 days, nausea and vomiting.
Results: On admission to the ED, the general status was altered, no fever, hemodynamic stable, BP = 106/70 mmHg, tachycardia with narrow QRS complexes, HR = 270-320 b/min, retrograde P waves, with no response to vagal maneuvers, no response to Adenosine 3 doses, considered an atrioventricular reentrant tachycardia (AVRT). Amiodarone was started, initially with the loading dose, then continuous infusion (15 microgr/kg/day), under which the tachyarrhythmia still persisted. In ICU, low dose of Propranolol orally was associated, with a slight decrease in HR to 210 b/ min, but also with an important decrease of BP (being in a septic state). The decision was to convert to Landiolol continue infusion (7.5 microgr/kg/min), to be able to manage the blood pressure. After 30 min the conversion to sinus rhythm was observed, with a heart rate of 90 b/min. The next day, the ECG showed an intermittent WPW syndrome, with alternative conduction 1 to 1. From the third day, the ECG was in permanent sinus rhythm. Abdominal ultrasound reveals acute cholecystitis and antibiotic therapy was initiated with Cefort and Gentamicin, and then changed to Meronem. The evolution was favorable with maintenance of sinus rhythm under orally Amiodarone and Propranolol. Holter ECG was completely normal. Electrophysiology study (EP) was performed with ablation of the accessory pathways.
Conclusions: Fever can trigger tachyarrhythmias such as AVRT and even to demask undiagnosed WPW syndrome, which is very rare in the same patient. The use of Landiolol in refractory tachycardias from the septic shock, can be of great benefit, being able to control the blood pressure due to the short time of metabolization. EP study is mandatory in such patient with two accessory pathways.

Keywords: atrioventricular reentrant tachycardia, WPW syndrome, Landiolol, septic shock

PP-047
KCNQ1 mutation presenting with complete heart block (CHB) and long QT syndrome (LQTS)

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Background and Aim: 2:1 Heart block and sinus bradycardia are well known with Type 2 and Type 1 LQTS respectively. CHB in association with Type 1 LQTS has not been described. We describe a rare case of a child with CHB and symptomatic Type 1 LQTS.
Method: A 20-month-old girl presented with an episode of syncope at home and subsequently was noted to have CHB. She underwent transvenous permanent pacemaker implantation using a Selectsecure 3830 lead (Medtronic inc, MN, USA) and Microny II SR + (Abbott,USA) pacemaker. She remained well for 3 years and then presented with out of hospital arrest during which automatic external defibrillator (AED) was used to restore sinus rhythm. Due to technical issues data could not be downloaded from the AED. While on intensive care unit an episode of polymorphic VT was noted. ECG showed CHB with QTc of 550 ms. On retrospective review her ECG's prior to pacemaker implantation had shown similar QT intervals. She made a full neurological recovery. Based on the finding a clinical diagnosis of LQTS was made.
Results: A hybrid implantable cardioverter defibrillator (ICD) was implanted using existing pacemaker lead by using subcutaneous anteroposterior Medtronic 35 cm Superior Vena Cava coils (Medtronic inc, MN, USA) with ICD generator in the abdomen. Genetic testing showed her to be heterozygous for pathogenic KCNQ1 mutation. She remains well on beta blocker therapy with Nadolol.

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Conclusions: CHB in setting of LQTS is not well described. The described HB are in the form of 2:1 Block and generally seen with KCNH2 and sometimes SCN5A mutation. KCNQ1 mutation is more commonly associated with bradycardia. To the best of our knowledge this is the first report of KCNQ1 mutation associated with CHB and LQTS.

Keywords: Long QT syndrome, Complete heart block, ICD

PP-048

Prevalence and impact of arrhythmias during midterm follow up after the extracardiac fontan procedure

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Background and Aim: Fontan patients are at increased risk to acquire rhythm disorders which may contribute to heart failure and Fontan related long-term complications such as plastic bronchitis and protein losing enteropathy. The extracardiac Fontan type might have benefits regarding the absence of arrhythmias, but long-term data are rare. Further, rhythm disorders as cause for Fontan failure are described diverse in the literature. Therefore, our aim was to evaluate the prevalence of arrhythmias in this extracardiac Fontan cohort as well as their risk for death.

Method: This retrospective single center study includes patients born between January 1994 and August 2013 (>10 years of age) who underwent an extracardiac Fontan procedure at our center. We reviewed medical charts, including electrocardiograms, Holter monitoring, current medication and need for pacemaker therapy until August 2023. Primary outcome measure was death of any cause among patients with relevant arrhythmias and statistical analysis was performed using Fisher's exact test.

Results: Of 321 Fontan patients with a median age of 12.6 years (IQR: 8.4), 285 (=88.8%) patients were in sinus rhythm or stable junctional escape rhythm (=non-relevant) at most recent electrocardiogram and had no history of any significant rhythm disorder post Fontan. Etiology of relevant arrhythmias (36/321, =11.2%) leading to antiarrhythmic therapy or pacemaker implantation are outlined in Table 1. 17/321 (=5.3%) patients with bradycardia required pacemaker therapy at a median age of 5.3 years. The risk for PB/PLE was significantly increased in patients with relevant arrhythmias ($p = 0.003$; OR: 4.2, 95% CI: 1.75, 10.06) compared to controls. Moreover, patients with relevant arrhythmias had a significantly increased risk of death ($p = 0.006$; OR: 5.0, 95% CI: 1.72, 14.43). In four patients death was due to cardiac failure, one patient suffered a sudden cardiac death and one patient died due to pulmonary complications. Two patients without documented arrhythmias died suddenly at home.

Conclusions: Extracardiac Fontan patients have a very low prevalence of relevant arrhythmias at our center. However patients with arrhythmias are at increased risk for adverse events such as PB/PLE or death. Therefore, these patients need thorough monitoring and radical medical and electrophysiological therapy to avoid a failing Fontan circulation.

Keywords: Fontan, single ventricle, arrhythmia, pacemaker, protein losing enteropathy, plastic bronchitis

Table 1

Table 1 – Patients with relevant arrhythmias	N= 36/321 (11.2%)
Male, n (%)	21 (58.3)
Patients in sinus rhythm, n (%)	**19** (52.8)
- recurrent supraventricular tachycardia	3
- Sinus node dysfunction + PM	3
- atrial flutter	1
- IART	3
- AVNRT	3
- intermittent tachycardic junctional rhythm	2
- intermittent AV block II	1
- ventricular tachycardia	3
Patients without sinus rhythm, n (%)	**17** (47.2)
- junctional escape rhythm + PM	3
- junctional escape rhythm + betablocker	2
- junctional escape rhythm + atrial flutter + PM	1
- AV block II	4
- AV block II + st.p. ventricular fibrillation + PM	1
- AV block III	1
- AV block III + PM	5
Pacemaker therapy, n (%)	17 (47.2)
Median age at pacemaker insertion, years (IQR)	5.3 (5.3)
Antiarrhythmic medication, n (%)	17 (47.2)
PB/PLE, n (%)	9 (25.0)
Death, n (%)	6 (16.7)
PM, pacemaker; IART, Intra-atrial re-entrant tachycardia; AVNRT, Atrioventricular nodal reentrant tachycardia; AV, atrioventricular; st.p., status post; PB, plastic bronchitis; PLE, protein losing enteropathy; IQR, interquartile range;	

Extracardiac Fontan patients with relevant arrhythmias.

PP-049

Permanent pacemaker implantation with hybrid endocardial perventricular approach in infancy: May be an alternative route in the absence of epicardial

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Background and Aim: Despite the advancements in technology, establishing the optimal implantation technique for pediatric patients with a pacemaker indication remains challenging, as factors such as age, body weight, venous occlusions, the presence of congenital heart diseases (intracardiac shunts, univentricle physiology etc.), and prior surgical procedures play a crucial role in determining the most appropriate implantation approach.

Method: A 4-month-old female infant, weighing 4 kg, diagnosed with Down syndrome, developed complete atrioventricular (AV) block following complete atrioventricular septal defect (AVSD) repair. The patient, closely monitored with a temporary pacemaker, was planned for permanent pacemaker implantation on the 10th postoperative day due to the persistence of complete AV block. Given that epicardial pacemaker leads were not available in our country at the time, an alternative method for endocardial lead implantation was pursued, as demonstrated in this case report.

Results: After obtaining written informed consent from the patient family procedure was performed under general anesthesia. Medtronic Select Secure 3830 MRI conditional, active fixation lumenless leads with length 59 cm was placed in this case. A prior sternotomy had been performed, and an introducer sheath was inserted through a pursestring suture placed on the right ventricular apicoanterior free wall. (Figure 1a) A screw-in endocardial lead was inserted through the introducer sheaths under florouscopy

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Figure 1.

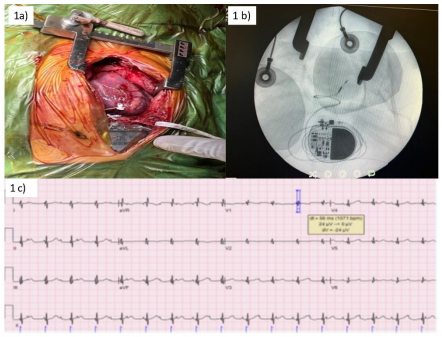

Figure 1a. Sternotomy with endocardial lead placement using hybrid approach Figure 1b. Post procedural chest x-ray with new transventricular endocardial screw-in lead; the pulse generator is located in the subxiphoid epigastric subcutaneous Figure 1c. ECG showing ventricular pace rhythm with nearly narrow QRS.

guidance into ventricular apicoseptum towards to midseptal area (Figure 1b).After confirming that the lead position and measurements were within normal limits, it was dissected and removed using an introducer cutter.Subsequently, it was affixed to the ventricular surface with pursue stitches. Leaving a sufficient epicardial loop, the epicardial battery (Vitatron single chamber generator) was implanted in the subdiaphragmatic region (initial setting was VVIR 90-150/bpm). Also the paced QRS duration (55-60 ms) was very narrow and IRBBB patern- like left bundle area pacing was shown in 12 lead ECG (Figure 1c) There were no procedural complications. Duration of hospitalization was 5 days and the patient was discharged under stable conditions and is being followed up.

Conclusions: We suggest that placement of endocardial lumenless leads by a transventricular approach provides an alternative to an epicardial system in children for epicardial lead malfunction or in the absence of an epicardial lead.

Keywords: hybrid, pacemaker, children

PP-050

Wearable devices in paediatric cardiology: An evaluation of their accuracy, diagnostic value and patient satisfaction

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Background and Aim: Wearables are increasingly being used by paediatric cardiology patients for heart rate (HR) and rhythm monitoring. They could overcome limitations in comfort and battery life of the conventional Holter ECG. Yet, the validation of wearables in paediatric populations is not well established. Therefore, the aim of this study is to investigate the accuracy of HR measurements from two wearables—a multi-sensor wristband and a smart ECG-shirt—under natural conditions in children attending a paediatric cardiology clinic. Additionally, we examined the influence of patient and measurement characteristics on HR accuracy, the efficacy of the ECG-shirt in arrhythmia detection and patient satisfaction with these wearables.

Method: Children indicated for 24h-holter monitoring were equipped with the Holter (gold standard), together with both wearables. HR agreement was assessed using Bland-Altman analysis, and accuracy was defined as percentage of HRs within 10% of Holter values. We conducted subgroup analyses based on BMI, gender, age and moment of wearing, among other factors. A blinded paediatric cardiologist analysed ECG-shirt data for rhythm classification. Patient satisfaction was measured using Likert-scale questionnaires.

Results: Thirty-one participants (mean age 13.2±3.6 years; 45% female) were included. Both wearables demonstrated good agreement with Holter readings (wristband bias –1.3 BPM; 95% Limits of Agreement [LoA]: –30.7 to 28.1. ECG-shirt bias –1.3 BPM; 95% LoA: –21.6 to 24.1). Mean accuracy was 84.4% (±8.6%) for the wristband and 86.7% (±11.2%) for the ECG-shirt. ECG-shirt accuracy was notably higher in the first 12 hours (94.9±7.4%) compared to the latter 12 (78.6±17.3%, $p<0.001$). Higher accuracy was observed at lower HRs (wristband: low vs. high HR: 90.4±9.7% vs. 78.8±10.6%, $p<0.001$; ECG-shirt: 89.8±14.7% vs. 83.8±12.0%, $p<0.005$). Based on ECG-shirt readings, 84% (26 of 31) of rhythms were correctly classified as normal or abnormal. Patient satisfaction scores were significantly higher for both the wristband (3.8±0.5, $p<0.001$) and ECG-shirt (3.6 ±0.8, $p<0.001$) compared to the Holter (2.6±0.7).

Conclusions: The multi-sensor wristband and smart ECG-shirt demonstrate good accuracy in paediatric HR monitoring and provide higher patient comfort than conventional monitoring. The ECG-shirt also shows potential in arrhythmia detection. While further development is warranted, these wearables show promise in enhancing diagnostics, therapeutic monitoring and patient safety in paediatric cardiology.

Keywords: wearable technology, heart rate monitoring

PP-051

Which implantable cardioverter defibrillator in paediatric patients with hypertrophyc cardiomyopathy?

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Background and Aim: Hypertrophic cardiomyopathy (HCM) in children is associated to a risk of sudden cardiac death (SCD) of 1,2-1,5% per year. Paediatric-specific risk models for SCD have been developed and implantable cardioverter defibrillators (ICDs) are implanted when indicated. Nevertheless, HCM clinical features can change during follow-up (after myectomy, for high dosage drug needed) and it can be challenging to decide which ICD system is favorable. The aim of this study is to show follow-up data of HCM children implanted with ICDs in our Centre, focusing on ventricular pacing need and on inappropriate functioning due to disease progression.

Method: From October 2008 to October 2023 we retrospectively enrolled 23 patients (11 males, 12±2 years old at ICD implant) with HCM and ICD. Five patients had a syndromic HCM (Danon disease, Noonan (2 pts), Leopard and Timothy syndrome); one child had a coronary artery anomaly. Ten patients had obstructive HCM and five of them underwent surgical myectomy during follow-up.

Results: Nineteen patients received an ICD for primary prevention of SCD (82.6%). Transvenous ICDs (TV-ICDs) were 11 (47.8%); subcutaneous ICDs (S-ICDs) were 10 (43.5%); 2 patients received epicardial systems. At follow-up (59 months) appropriate shocks were recorded in 9 children (39%, 2 with S-ICD, 7 with TV/epicardial ICD); inappropriate shocks were reported in 5 patients (22%, 3 with S-ICD, 2 with TV-ICD): in one of S-ICD patient

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the inappropriate therapy was due to changes in ECG vectors after a large surgical myectomy. Patients with obstructive HCM received TV-ICDs in 5 cases (50%). Ventricular pacing in patients with TV/epicardial ICD was near-zero (0-1%) except for a patient with iatrogenic complete AV block after surgical myectomy. In one patient with the epicardial ICD the ATP (antitachycardia pacing) algorithm stopped a ventricular tachyarrhythmia. Complications due to pocket revision or lead repositioning were recorded in 3 cases (13%).
Conclusions: In our cohort of HCM children with ICDs there were no significant differences in outcomes among ICDs implanted. In one patient with obstructive HCM surgical myectomy after the implant was a problem for successive S-ICD therapies. Ventricular pacing was not necessary in almost all patients.

Keywords: Sudden cardiac death, Hypertrophic cardiomyopathy, Implantable cardioverter defibrillator

PP-052
Ivabradine as antiarrhythmic drug in paediatric patients
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Background and Aim: Ivabradine (IVA) is a selective inhibitor of the potassium channel HCN4, responsible for generation of the "funny current". For its selective mechanism of action IVA reduces heart rate without affecting inotropy. It has been used for years in systolic heart failure and chronic angina, validated for inappropriate sinus tachycardia and recently used in rate or rhythm control of automatic atrial or junctional arrhythmias. In this study we report our experience in the use of IVA as antiarrhythmic drug in children with focal atrial tachycardia (FAT) or idiopathic junctional tachycardia (JT).
Method: From February 2019 to October 2023, we retrospectively enrolled 11 patients treated with IVA at our Institution for FAT or JT. Tachycardia was diagnosed using standard 12-lead electrocardiography and confirmed by 24-h Holter monitoring. Echocardiography was performed in all patients and tachycardia-induced-cardiomyopathy (TIC) was diagnosed with left ventricular ejection fraction (LVEF) < 50%, excluding other causes of cardiomyopathy.
Results: Patients enrolled (11, 5 males, 8±5 years) had FAT in 9 cases (2 patients had a congenital heart disease surgically treated time before), JT in 2 cases. In 4 children TIC was present at diagnosis (mean LVEF 43%). IVA was administered per os every 12 hours as monotherapy in 4 children and in association to other drugs (amiodarone, flecainide and/or propranolol) in the others. IVA was effective in slowing down or terminating the arrhythmia after 48 hours from the maximum tolerated dosage in 8 cases (73%). In 4 of these patients it was given as monotherapy (1 JT, 3 FAT), in other 4 cases IVA was associated to amiodarone or flecainide. Among children with TIC only one experimented successful control of the JT and recovery of LVEF. IVA dosage was variable (range 0,05 mg/kg/die – 0,3 mg/kg/die). During follow-up (1,6±1,4 years) in 3 patients successfully treated with IVA, the arrhythmia was absent at 24-h Holter monitoring in wash-out (1 JT, 2 FAT).
Conclusions: Efficacy of IVA as antiarrhythmic drug in children with automatic tachyarrhythmias (FAT or idiopathic JT) is fair (73%) in our cohort. Future studies, prospective and multicenter, could clarify efficacy and safety of IVA in these rare patients.

Keywords: automatic tachyarrhythmias, children, ivabradine

PP-053
Phenotypic and genotypic presentation of pediatric inherited primary arrhythmias – a long-term countrywide survey
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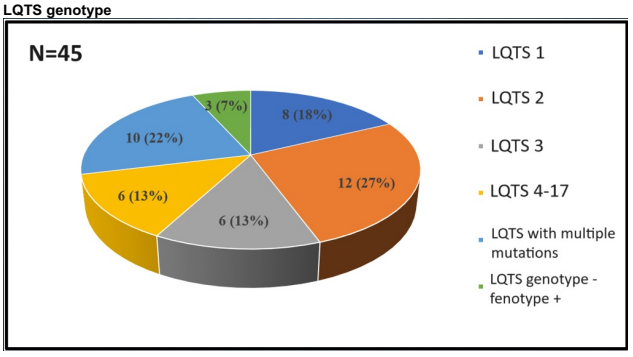
Background and Aim: Primary arrhythmia syndromes (PAS) are inherited electrical disorders of the heart where pathologic gene mutations cause abnormal structure and function of cardiac ion channels. Phenotypically they are usually manifested as malignant ventricular arrhythmias.
The aim of the study is to present phenotypic and genotypic characteristics of a nationwide cohort of patients diagnosed with PAS in a specialized tertiary care center with countrywide referrals during 15 consecutive years (September 1st, 2008 to August 31st, 2023) Analyzed characteristic comprised clinical symptoms, family history, ECG findings, genetic testing, and administered therapies.
Method: Retrospective chart review-based analysis of a pediatric countrywide cohort (0-19 years old), diagnosed with PAS and managed by a specialized pediatric cardiology center. Symptoms attributable to PAS were defined as syncope of unknown etiology, seizures, and aborted sudden cardiac death (SCD). Associated family history was defined as positive in the presence of a channelopathy or SCD. Causative mutation was defined as the presence of a pathogenic/likely pathogenic mutation or a variant of unknown significance in the case of clinical or ecg signs of disease. Statistical analysis by means of chi-square test, T-test or Wilcoxon test was applied.
Results: During the 15-year-period 61 patients diagnosed with PAS were assessed. The most prevalent syndrome was long QT syndrome (LQTS) in 49 patients (80%), followed by catecholaminergic polymorphic ventricular tachycardia (CPVT) in 6 (10%) and Andersen-Tawil syndrome in 3 (5%).



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Comparing the symptomatic (43%) and the asymptomatic (57%) group of LQTS patients at the time of the diagnosis, significant differences were observed in corrected QT interval, Schwartz's score, proband status, family history of LQTS and occurrence of recurrences. Most prevalent gene mutation was KCNH2 (27%). Aborted SCD and fetal arrhythmias were each detected in 24% of symptomatic patients. All subjects (n=5) with LQTS and sinoatrial node dysfunction had mutation in KCNQ1 gene. ICD was implanted in 13 (27%). KCNH2 mutation with malignant arrhythmia presentation was managed by ICD implantation.
Conclusions: LQTS was the most prevalent channelopathy found during 15 years in patients with PAS, with type 2 being the most frequent. ICD was implanted in 27% of PAS patients, mostly in carriers of KCNH2 mutation.

Keywords: arrhythmia, children, cardiac channelopathy, long QT syndrome

Cardiac Imaging

PP-054
Effect of sars-cov-2 infection on cardiac function in children and adolescents with duchenne muscular dystrophy

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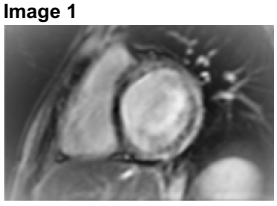
Background and Aim: In this case series, we describe the diagnosis of myocarditis in asymptomatic children and adolescents with Duchenne muscular dystrophy (DMD) after a mild illness with COVID-19. These patients had abnormal electrocardiogram (ECG) and echocardiogram (EEG) findings that were not present prior to COVID-19 diagnosis, which led to a referral for cardiac magnetic resonance imaging (cMRI). These patients were referred for magnetic resonance imaging (cMRI) because of electrocardiographic (ECG) and echocardiographic pathological findings that were not present prior to COVID-19 diagnosis.
Method: Four patients aged 6–12 years old with DMD, good mobility, and no cardiorespiratory symptoms underwent cardiac evaluation <1 month after diagnosis of COVID-19 disease to rule out potential cardiac complications and deterioration of the underlying disease. Evaluation included an ECG, echocardiogram, holter rhythm monitoring and cMRI.
Results: Two patients showed extrasystolic ventricular arrhythmia in their ECG. Left ventricular dilatation was observed on transthoracic echocardiography (TTE) in all patients, with a moderate reduction in the left ventricle's ejection fraction (EF: 45-55%) at first. cMRI confirmed the above findings and identified the presence of severe myocardial inflammation in all patients, based on an abnormally elevated myocardial T2 ratio, late gadolinium enhancement, natural T1 mapping, T2 mapping and extracellular volume fraction, in correspondence with simultaneous disturbance of left ventricular function. In all cases, treatment with ACE inhibitors, carvedilol and eplerenone was initiated. Two out of four patients developed episodes of non-sustained ventricular tachycardia on holter monitorin in the following 6 months of follow up, in combination with deterioration of left ventricular function with EF<35-40% on TTE. Prophylactic implantation of a cardioverter-defibrillator was carried out in the above two patients.
Conclusions: In conclusion, assessment of cardiac function in asymptomatic DMD patients after COVID-19 disease demonstrates the diagnostic value of cMRI in detecting myocarditis for early therapeutic intervention.

Keywords: Duchenne muscular dystrophy, myocardial dysfunction, COVID-19, cardiac MRI

Image 1



Late Gadolinium Enhancement was shown in cMRI of all four patients of our study.

PP-055
Cardiac involvement and therapeutic management in patients with duchenne muscular dystrophy

Aikaterini Giannakopoulou[1], Antonios Belegrinos[2], Evangelia Grigoraki[3], Marina Katsalouli[1] and Evangelos Karanasios[4]
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Background and Aim: Duchenne muscular dystrophy (DMD) is caused by mutations in the dystrophin gene on chromosome Xp21. Almost all patients with DMD who survive to adulthood, develop dilated cardiomyopathy (DCM) until their third decade of life. Clinical presentation includes peripheral muscle weakness, signs and symptoms of DCM/heart failure and chronic respiratory failure. Object of this study was the early diagnosis of DCM in children and young adolescents, aiming to early therapeutic intervention.
Method: 22 male patients with DMD mean age 12.5+1.8 years were studied. The following parameters of left ventricle systolic function were evaluated through transthoracic echocardiography (TTE): ejection fraction (LVEF), fractional shortening (FS), segmental mobility and diastolic function via tissue Doppler(TDI). Mitral E wave velocity (Ea) and E/Ea ratio were evaluated, as well as global strain with speckle tracking echocardiography (STE). Study population complementary underwent magnetic resonance imaging (cMRI). Late Gadolinium Enhancement (LGE) on T1 sequence was studied to highlight any myocardial fibrosis.
Results: No patient reported symptoms and all had normal respiratory function. Sixteen patients had normal LVEF ≥ 65% and FS>35% without segmental hypokinesias. In twelve of them (75%), cMRI showed LGE in the inferior and lateral wall with involvement of the interventricular septum, despite normal TTE. Six patients (27%) with mildly affected LVEF ejection fraction (<55%) on TEE had extensive and diffuse LGE on cMRI. STE showed a global longitudinal strain of less than -18% in 2 (9%) patients. One of them showed ventricular arrhythmia on Holter monitoring. On TDI no patient demonstrated signs of diastolic dysfunction. There was no correlation of the neurological status of patients to cardiac findings (p=NS). Based on these early

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findings, 18 patients (81.8% of total population) were treated gradually as first line prevention with perindopril, carvedilol and spironolactone, aiming to delaying progression of DMD-DCM.
Conclusions: DCM and heart failure are major complications of DMD. Management is based on regular monitoring with electrocardiography and TTE. 2D strain echocardiography and cMRI can help in early diagnosis of cardiac involvement and improve prognosis through initiation of appropriate cardioprotective treatment.

Keywords: Duchenne muscular dystrophy, echocardiography, cMRI, cardioprotective medical treatment

PP-056

A gigantic congenital right atrial appendage aneurysm in an infant: Ten-year follow-up

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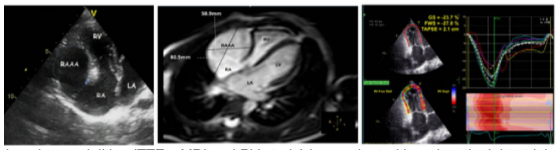
Background and Aim: Gigantic right atrial appendage aneurysm (RAAA) is a rare condition usually discovered during the third decade of life after being symptomatic. We present an asymptomatic RAAA discovered early during basic screening of an infant, its natural history and a ten-year follow-up, because of parents' refusal to operation.
Method: A 10-month-old male infant was referred due to a heart murmur before planned partial nephrectomy due to right upper renal atrophy with an ipsilateral duplicated collecting system. Electrocardiogram (EKG), transthoracic echocardiography (TTE), 24-hour holter monitoring, and cardiac magnetic resonance imaging (cMRI) were performed.
Results: EKG was normal. TTE revealed a giant RAAA with extensive communication with the right atrium (RA). Basal diameter of right ventricle was 17 mm (normal range for body surface area = 0.46 m2 ≤ 18). No thrombus was detected within the aneurysm. The ventricles had normal contractility and dimensions. The right ventricle was not compressed by the aneurysm and blood flow through tricuspid valve was normal. cMRI showed a giant RAAA (39 × 33 mm) with thick walls and a wide communication (27 mm at diastole and 15 mm at systole) with the RA. The right coronary artery was not compressed by the RAAA despite being in contact with it for 25 mm. Cardiac surgery was recommended, but due to the parents' refusal, the child was given acetylsalicylic acid 5 mg/kg BW daily to avoid thrombosis. Mild physical activity and frequent cardiological monitoring were recommended. At 10-year follow-up, the boy remains asymptomatic, the aneurysm continued to grow: 80 mm × 59 mm with a total volume of 110 mL (cMRI 7 years later). No arrhythmias were detected in 24-hour holter recordings nor thrombi in the RAAA. Right ventricular function remained normal as assesed via right ventricular strain.
Conclusions: RAAA can remain asymptomatic for a long time, especially in children. cMRI provides useful information about size, wall thickness, the course of the right coronary artery and potential sites of right ventricular compression by the aneurysm. The recommended treatment is surgical excision. In patients who do not desire surgery, close monitoring and treatment are required to avoid life-threatening complications.

Keywords: right atrial appendage aneurysm, infant, cMRI

Image 1



Imaging modalities (TTE, cMRI and RV strain) in a patient with a gigantic right atrial appendage aneurysm.

PP-057

Friedreich's ataxia- associated childood hypertrophic cardiomyopathy: The experience of a tertiary hospital

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Background and Aim: Friedreich's ataxia (FA) is a very rare autosomal neurodegenerative disease (1:50000) that occurs in childhood and early adolescence.Majority of patients (>60%) develop left ventricular hypertrophy and die due to cardiovascular complicationsm,with an average life expectancy of 40 years. Purpose of this study is to describe cardiovascular involvement of FA and its optimal treatment in a Neuromuscular Disease Unit (NMD) of a tertiary hospital.
Method: 403 patients with neuromuscular diseases were monitored in MNN. Only seven of these (1.7%), (3 men and 4 women) suffered from FA with mean age 17.8 years and a range of values from 11 to 31 years old. Genetic comfirmation was performed in all patients. The mean age of diagnosis was 10 years old and was due to neurological symptoms. All patients underwent electrocardiogram (ECG), echocardiography (TTE), Holter monitoring and cardiac magnetic resonance imaging(cMRI). Average follow-up was 8 years.
Results: Only two out of seven patients with FA (28.5%) had normal cardiac function in ECG and TTE. ECG findings of the rest five patients were repolarization disturbances, increased potentials and profound negative T waves in 80%. TTE showed concentric hypertrophy without left outflow obstruction in all five patients (wall thickness: 14.15mm+0.9) and normal ejection fraction. In Holter monitoring four patients had isolated monomorphic supraventricular and ventricular extrasystoles. Loop recorder was implanted in one patient which, 3 years later, didn't record life-threatening arrhythmias and was removed. All five patients had persistently elevated troponin I levels. These patients underwent gadolinium-enhanced cMRI which demonstrated extensive diffuse fibrosis and increased end-diastolic wall thickness. One of these patients died at 19 years of age due to worsening heart failure, initially developing left ventricular hypertrophy, subsequential progressive left ventricular wall thinning, decreased ejection

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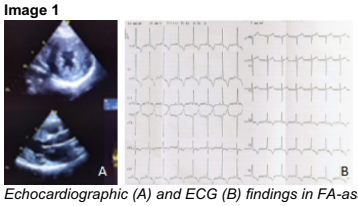
fraction (<25-30%) and ventricular arrhythmias. All patients were treated with ACE inhibitors, eplerenone, and coenzyme Q10 because of its similarity to idebenone, a promising drug in some studies.
Conclusions: FA is a rare, inherited, multisystem disorder of mitochondrial function with neurological and cardiac complications. Systematic close monitoring of cardiac complications, proper treatment and the development of new therapeutic strategies are required to improve the prognosis of this disease.

Keywords: Friedreich's ataxia, Hypertrophic cardiomyopathy, optimal medical therapy

Image 1



Echocardiographic (A) and ECG (B) findings in FA-associated hypertrophic cardiomyopathy in one of our patients.

PP-058
Giant ra diverticulum- an uncommon cause of palpitations
Akriti Gera, Pankaj Bajpai, Rajesh Sharma, Shefali Yadav and Anil Bhan
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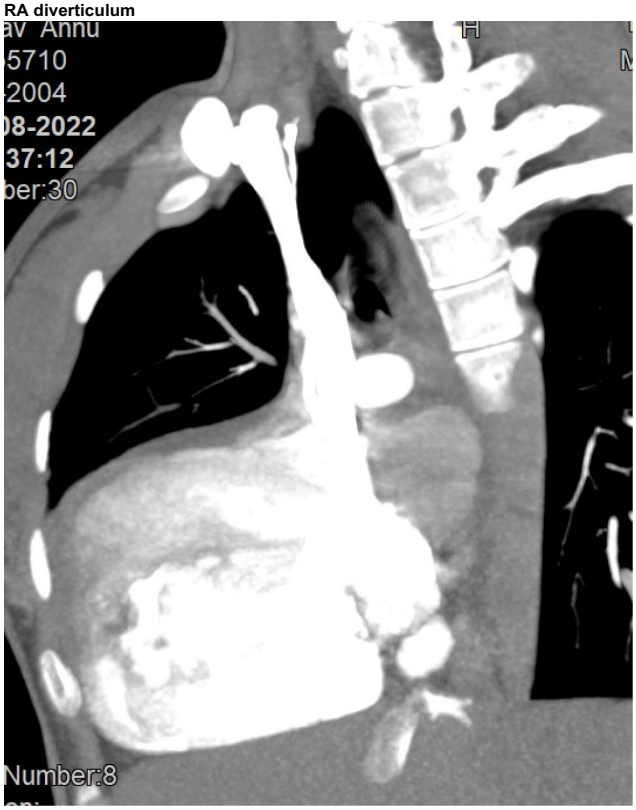
Background and Aim: Right atrial (RA) diverticulum is a very rare congenital anomaly with unclear aetiology. It is usually asymptomatic and incidentally detected, and hence only about 20 cases have been diagnosed worldwide. We report a case of a symptomatic giant RA diverticulum.
Method: We retrospectively reviewed a single case and collected data from records. Review of literature on RA diverticulum was done.
Results: We report the case of an 18-year-old female who presented to us with palpitations. She was hemodynamically stable and cardiovascular examination was unremarkable. ECG showed sinus tachycardia, with a heart rate of 92 beats per minute. 2D echocardiography showed a huge (96X87mm) diverticulum with wide neck (40mm) on the lateral wall of right atrium (RA) compressing right ventricle (RV), with no effect on the tricuspid valve. Spontaneous echogenic contrasts were seen in IVC and RA diverticulum. CT Pulmonary Angiogram showed a large wide neck diverticulum (7.8 x 9.1 x 8.6 cm) arising from the right anterolateral wall of the right atrium compressing and displacing the right ventricle across the midline to the left side. Our patient underwent resection of RA diverticulum and patch closure using bovine pericardium. Post operatively, there was no leak through RA patch and no residual RA aneurysm. Biventricular function was normal. On follow up visits, she is asymptomatic and doing well.
Conclusions:

1. Single RA diverticulum is an uncommon but treatable cause of palpitations. A thorough evaluation is hence warranted in all patients.
2. Giant RA diverticulum may be complicated by atrial arrhythmias, thromboembolism and sudden death and hence are treated surgically.

Keywords: Right atrial diverticulum, Arrhythmia, Isolated right atrial aneurysm

RA diverticulum



Contrast CT showing right atrial diverticulum

PP-060
Multimodality imaging and functional assessment in patients with systemic right ventricle and biventricular physiology
Alice Pozza[1], Martina Avesani[1], Elena Reffo[1], Irene Cattapan[1], Annachiara Cavaliere[2], Sofia Piana[1], Anna Molinaroli[1], Biagio Castaldi[1] and Giovanni Di Salvo[1]
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Background and Aim: Right ventricle (RV) might support systemic circulation, resulting in systemic RV (sRV). Biventricular versions of sRV include congenitally corrected transposition of the great arteries (cc-TGA) and dextro-transposition of the great arteries (D-TGA) after Mustard or Senning procedure. sRV may cause a high burden of morbidity and premature mortality from heart failure. A close follow-up is therefore mandatory. Echocardiographic assessment of sRV is challenging. Cardiac magnetic resonance imaging (CMR) allows functional evaluation of sRV, while cardiopulmonary exercise test (CPET) provides information about functional performance and exercise limitations.

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We aimed to compare a range of advanced echocardiographic parameters with CMR-derived parameters and to evaluate their correlation with exercise performance.
Method: Retrospective single centre study of 22 patients with sRV and biventricular circulation. Patients recruited had a clinical visit, Echo and/or CMR scan within one year from last follow-up visit (September 2022-September 2023). When available, data from CPET was also collected. Qualitative variables are reported as median values, standard deviation or percentage.
Results: 59.1% female, 13 cc-TGA; of the 9 TGA Mustard/ Senning patients studied, 55.5 % had undergone Senning repair. Most patients were clinically asymptomatic. RV EF derived from 3D echocardiography was 47%± 8 SD (30-59%). Median RV EF derived from CMR was 49.5% (41-73%). FAC derived from basal echocardiography was 37%± 7 SD (19-50%) and from CMR 34.16±10.45 SD (19-61%). E/E' was inversely correlated with peak atrial longitudinal strain of right atrium above sRV (ρ -0.9) and peak VO2 (ρ -0.8). Similarly, a significant correlation was found between CMR-derived RV EDV and echocardiographic-derived RV EDA (ρ 0.8). When cc-TGA and D-TGA patients were compared separately, the two cohorts did not differ for CPET data. By analyzing CMR-data, cc-TGA population showed better EF and GLS parameters (p 0.002). By echocardiographic assessment, cc-TGA population showed a bigger indexed atrial EDA (p 0.04).
Conclusions: Echocardiography is a valid technique to screen for sRV dilatation and to assess systolic function as compared to CMR. Further investigations of diastolic function may help in better understating the role of diastole in exercise performance.

Keywords: Systemic right ventricle, Advanced Echocardiography, Cardiac Magnetic Resonance Imaging

PP-061
Role of CT scan in newborns and infants with congenital heart disease

Alice Pozza, Irene Cattapan, Jennifer Fumanelli, Elena Reffo, Martina Avesani, Roberta Biffanti, Alessia Cerutti, Domenico Sirico, Biagio Castaldi and Giovanni Di Salvo
Pediatric Cardiology Unit, Department of Women's and Children's Health, University of Padova, Italy

Background and Aim: Improvements in diagnostic techniques brought to a better understanding of congenital heart disease (CHD), thus allowing a more comprehensive clinical and surgical management. Echocardiography (Echo) is the first imaging modality because of easy availability, limited costs and safety. A larger interest in the use of computed tomography scan (CT) has been achieved, although its application is generally limited because of safety concerns and low image quality.
We aimed to evaluate differences in diagnostic performance of Echo and CT, to investigate how clinical and surgical management are affected, accordingly to each imaging technique.
Method: Retrospective longitudinal single center study, involving 22 patients aged <1 year (11 newborns, 11 infants) diagnosed with CHD between 2020-2023. Newborns diagnosed with CHD undergone Echo (mean age 1.5 days) and results were discussed to evaluate future surgical plan. For those whose cardiac anatomy was challenging a CT was performed (mean age 10 days). Infants diagnosed with CHD underwent CT as part of the surgical planning (mean age 162.91 days). CT data were compared to the most recent Echo assessment. Main indications to perform advanced imaging were revised.
Results: In newborn population main indications for CT were: evaluation of coronary artery anatomy (4), multiple aorto-pulmonary artery collaterals (2), great vessels abnormalities (3 aortic arch anomalies, 1 pulmonary artery anomaly, 1 anomalous venous return). Good agreement Echo vs CT in 6 cases, while in 5 cases CT added details to diagnosis. In infants CT was performed in case of challenging coronary artery anatomy (2), anomalous venous returns (3); better definition of intracardiac anatomy (6). In 7 cases Echo and CT reports were comparable, whereas disagreed in 4 cases, mainly due to coronary artery anomalies and definition of pulmonary venous return anatomy. Only in one case CT findings significantly changed the surgical plan for patients.
Conclusions: Intracardiac anatomy is well depicted either by Echo or CT. CT plays an incremental role in the assessment of extra-cardiac and vascular anatomies, notoriously difficult to be assessed by Echo. Echo is superior to CT to assess valvular anatomy and function. Multimodality imaging is of paramount importance to plan surgical and interventional management of complex CHD.

Keywords: Echocardiography, Computerized Tomography scans, Multimodality imaging

PP-062
Cardiac - cerebrovascular hemodynamics and acquired neonatal brain injury during perinatal transition in transposition of the great arteries

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Background and Aim: Neonates with transposition of the great arteries (TGA) are vulnerable to acquired clinically silent brain injuries, including stroke and white matter injury (WMI), which has been shown to predict neurodevelopmental outcomes in this population. NeoDoppler is a non-invasive tool for continuous cerebral blood flow velocity monitoring that can be used for studying the effect of hemodynamic changes on cerebral blood flow. We aim to describe the cardiac and cerebrovascular physiology during the transitional period to understand the mechanisms involved in the generation of WMI.
Method: This is a prospective, observational cohort study that enrolled mothers pregnant with fetuses with TGA. The newborns were followed from birth until cardiac surgery with regular imaging of the cardiovascular and cerebrovascular physiology in the first 48 hours using Echocardiography and NeoDoppler, respectively.

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Cerebral magnetic resonance imaging was performed pre-operatively, and WMI classified according to a previously published WMI score.
Results: Seven infants were enrolled. Clinical characteristics, surgical and interventional timings are outlined in Table 1, along with the imaging modalities used for monitoring. Among the subjects, five patients underwent a balloon atrial septostomy. Two infants had WMI identified on pre-operative MRI (SK2 & SK6), while stroke was observed in one patient (SK7). There was a trend of increasing left and right ventricular cardiac outputs over the first 48 hours of life (Figure 1). The augmentation in cardiac output for infant SK6 was not as prominent, with sustained lower combined, left and right ventricular outputs. NeoDoppler monitoring on this particular infant revealed sustained lower peak systolic and end-diastolic velocities from 6 hours of age (Figure 2). On pre-operative MRI, SK6 was the infant with extensive bilateral WMI.
Conclusions: A granular approach to the medical management of neonates with TGA using precision cardiac and cerebrovascular monitoring may potentially identify infants at the highest risk. This strategy could help in the development of preventative measures to mitigate against common and significant brain injuries.

Keywords: Transposition of the Great Arteries, Neuromonitoring, Acquired Brain injury, Echocardiography, NeoDoppler, MRI

Table and figures describing the clinical characteristics, NeoDoppler findings and interventional history (Table 1), right and left ventricular outputs calculated by Echocardiography (Figure 1), and NeoDoppler parameters measured on infant SK6 (Figure 2)

Table 1. Clinical characteristics, NeoDoppler findings and interventional history together with result of pre-operative MRI of 7 neonates with Transposition of the Great Arteries. *Mean (std) velocities and Resistive Index from a 2 min recording at 6 and 12 h of life. TAV = time averaged maximum velocity, EDV = end diastolic velocity, PSV = peak systolic velocity*

	SK1	SK2	SK3	SK4	SK5	SK6	SK7
Gestational age	38+6	39+5	39+1	38	39+4	39+5	38
Apgars (1,5 mins)	7,8	3,8	9,9	8,8	7,8	5,5	4,7
Weight (g)	4190	3350	3225	3000	3680	3000	3820
Head circumference (cm)	36	33	33	32,5	37	33,5	35
Ventricular Septal Defect	No	No	No	Yes	Yes (+ Coarctation)	No	No
Balloon Atrial Septostomy	Yes	No	Yes	Yes	Yes	No	Yes
Continuous 48 hour NeoDoppler	No	No	Yes	Yes	Yes	Yes	Yes
NeoDoppler at 6 & 12 hours*	6 12	6 12	6 12	6 12	6 12	6 12	6 12
Mean PSV (Std) (cm/sec)	- -	- -	16.4 (2.7) 14.8 (1.4)	18.1 (1.1) 17.7 (1.7)	12.9 (0.7) 11.1 (1.09)	7.2 (0.9) 6.7 (0.2)	16.2 (1.7) 15.9 (1.8)
Mean EDV (Std) (cm/sec)	- -	- -	3.3 (1.0) 2.8 (0.3)	1.9 (0.3) 1.6 (0.2)	3.1 (0.7) 1.9 (0.4)	1.8 (0.1) 0.7 (0.7)	4.2 (0.4) 1.7 (1.0)
Mean TAV (Std) (cm/sec)	- -	- -	8.9 (1.6) 7.4 (0.9)	7.2 (1.1) 7.3 (0.96)	6.9 (0.5) 5.5 (0.6)	4.3 (0.1) 2.7 (1.1)	9.5 (1.0) 8.9 (1.0)
Mean Resistive Index (Std)	- -	- -	0.80 (0.04) 0.81 (0.02)	0.89 (0.02) 0.91 (0.01)	0.76 (0.02) 0.83 (0.03)	0.75 (0.05) 0.87 (0.02)	0.74 (0.02) 0.93 (0.02)
Depth (mm)	- -	- -	15-18 14-17	15-16 15-18	14-17 14-17	23-27 23-27	11-15 12-16
Cerebral NIRS	67 61	67 61	60 60	64 64	64 61	64 66	- -
Pre-operative MRI result	Small focus of ischemia	Mild WMI	No injury	No injury	Nonspecific T2 hyperintensity	Severe WMI, bicitate peak on MRS	Large right posterior cerebral artery territory infarction
Postnatal age at Arterial Switch Operation (day)	3	8	7	7	3	9	6

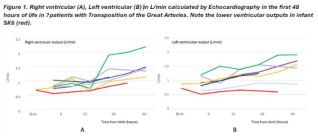

Figure 1. Right ventricular (A), Left ventricular (B) in L/min calculated by Echocardiography in the first 48 hours of life in 7 patients with Transposition of the Great Arteries. Note the lower ventricular outputs in infant SK6 (red).

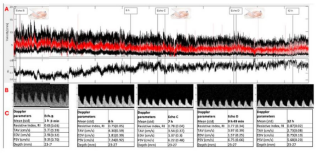

Figure 2. NeoDoppler parameters measured on infant SK6. (A) The trend of NeoDoppler data in the first 12 hours of life. (B) Examples of NeoDoppler tracings taken from a 1 minute recording, each example having the same scale. (C) Mean values (standard deviation) from a 1 minute recording during serial echocardiograms and at 6 & 12 hours of life. TAV = time averaged maximum velocity, EDV = end diastolic velocity, PSV = peak systolic velocity, RI = resistive index.

PP-063

Anomalous coronary artery from the opposite sinus: differences in the clinical-instrumental presentation profile in a pediatric and adult cohort

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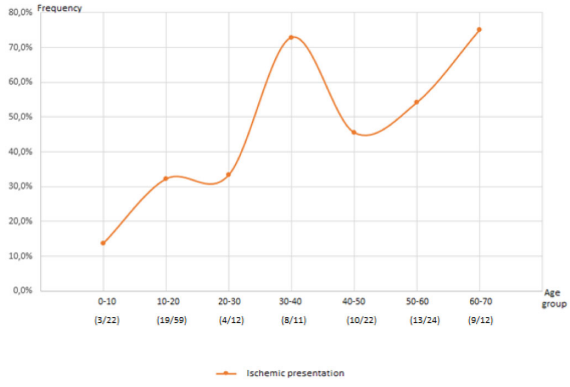
Background and Aim: Anomalous Coronary Artery from the Opposite Sinus (ACAOS) is one of the most challenging topic in cardiology. Mainly due to the not linear presentation and clinical significance, although awareness of their associated fatality is spreading. This study assesses the manifestation's pattern frequency and significance of ACAOS across different ages.
Method: We retrospectively reviewed imaging and clinical data, stratifying patients by age to identify age-specific patterns in the condition's manifestation. A total of 162 patients diagnosed with ACAOS through coronary CT were included; 67.9% were male, with a median age at presentation of 19.4 years. The most prevalent anomalies were the right ACAOS (62.3%), followed by the left ACAOS (30.2%) and circumflex anomalous origin (7.4%). High-risk anatomical features included: interarterial course (74%), intramural course (58%), and slit-like ostium (53.7%). An ischemic presentation occurred in 40.7%, with 14% experiencing major cardiac events (myocardial infarction, exertional syncope, aborted sudden cardiac death).
Results: We identified an age cut-off of 30 years related to a symptomatic presentation in ACAOS. The patients > 30 years old had a significantly more frequent presentation with angina (33% vs 18.5%, p<0.001) and acute coronary syndrome (10% vs 2.2%, p=0.03), whereas those < 30 years were more frequently asymptomatic at the diagnosis (53% vs 29%, p=0.008). Risk factors associated with an ischemic presentation were age at diagnosis (OR:1.035, p<0.0009), slit-like ostium (OR:2.5, p=0.007), intramural course (OR:2.3, p=0.013) and interarterial course (OR:2.4, p=0.028). Variables associated with major cardiac events were proximal vessel diameter (OR:1.23, p=0.036) and left ACAOS (OR:3, p=0.017), although left ACAOS resulted in being a risk factor for major cardiac events (OR 3.3, p:0.016) at multivariable analysis.
Conclusions: The ACAOS in the pediatric and adult populations have significantly different presentation modalities. In particular, adults with an age > 30 years had higher rates of ischemic manifestation. In addition to the known anatomical risk factors (i.e. slit-like ostium, intramural course, interarterial course), age at diagnosis is associated with an ischemic presentation, suggesting that asymptomatic pediatric and young adult ACAOS requires long–term follow-up because there may present ischemia at an older age.

Keywords: ACAOS, cardiac ischemia.

Figure 1



Course of ischemic sign and symptoms stratified by age group. It can be observed that after the age of 30, there is an increase in the incidence of ischemic presentation

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PP-067

Premature closure of the ductus arteriosus in a newborn, challenges of diagnosis. clinical case report

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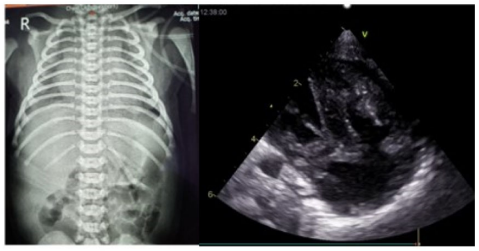
Background and Aim: Premature closure of the ductus arteriosus (DA) is rare and without prenatal sonography its diagnostics may be complicated. Premature closure of the DA causes increased pressure in the right ventricle, resulting in right ventricular hypertrophy (RVH) and pulmonary hypertension (PH) and can be fatal. Our aim is to present a clinical case of a newborn admitted to NICU with respiratory distress and diagnosed with severe RVH and PH, caused by premature closure of the DA.
Method: A newborn boy was admitted to the neonatal ICU 5 hours after delivery with severe respiratory failure. From maternal history it is known that the mother has suffered from viral infection a week before labor, which was treated with high doses of over-the counter ibuprofen. Additionally, alcohol consumption and smoking during pregnancy were disclosed. Prenatal sonography before labor was not performed. On examination: a neonate appeared acutely ill, presenting with respiratory failure and was oxygen dependent. Chest retractions, peripheral cyanosis were evident. On auscultation: bronchial breathing, coarse crackles, and 3\6 systolic ejection murmur were auscultated.
Results: Chest roentgenography showed cardiomegaly and areas of consolidation at the right lobe of the right lung. Therefore, congenital pneumonia was suspected. Echocardiography was performed due to the cardiomegaly and revealed severe hypertrophy of the right ventricular wall – 0.7 mm (Z-score +3,45), moderate tricuspid insufficiency with right ventricular pressure of 70 mmHg. Atrial septal aneurysm without perforation was visualized and no detectable flow through the DA was found. Coarctation of aorta was ruled out, therefore, a premature closure of PDA as a cause of RVH was suspected. The infant was placed on CPAP, his general state improved and RVH was resolved during the following weeks.
Conclusions: Fetal premature DA closure may cause symptoms of circulatory failure and even result in intrauterine death. This pathological process can be either idiopathic or could be explained either by abnormally low levels of circulating prostaglandin or maternal intake of prostaglandin synthase inhibitors, as was in our case. Prenatal ultrasound plays an important role in diagnosis. Urgent cesarian section may improve neonatal condition. Supplemental oxygen and time are the needed treatment for RVH to resolve.

Chest Rentgenography and Echocardiography



Chest rengenography shows cardiomegaly Echocardiography reveales severe hypertrophy of the right ventricular wall

Keywords: patent ductus arteriosus, premature closure, right ventricular hypertrophy, pulmonary hypertension, newborn, prostaglandin

PP-068

Hypoattenuated leaflet thickening after transcatheter pulmonary valve implantation with the venus p-valve: A call for action

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Background and Aim: The Venus P-valve is a recently developed self-expanding valve, designed to be implanted in dilatated native right ventricle outflow tracts (RVOTs). The optimal antithrombotic therapy after successful transcatheter pulmonary valve implantation (TPVI) with this valve remains unclear. Subclinical leaflet thrombosis, detected as a hypoattenuated leaflet thickening (HALT) on computed tomography (CT), can be associated with reduction of leaflet motion (RELM), increased gradients, and adverse clinical outcomes. The study aimed to assess the incidence of HALT and RELM after TPVI with the Venus P-valveTM.
Method: In this retrospective study, data on patients undergoing TPVI with the Venus P-valveTM between January and April 2023 was analyzed. After the procedure, the single antiplatelet therapy (aspirin, 100 mg daily) was prescribed for at least 6 months, except for patients on life-long oral anticoagulation therapy who continued with the anticoagulation only. CT was performed 6 months after-TPVI. Documentation of HALT was defined as hypo-attenuated thickening with or without rigidity of one or more cusps.
Results: In the study period 8 patients received a Venus p-Valve. Seven patients performed CTA (mean age 46 ± 13 years, including 4 females). Five patients were discharged on aspirin and 2 patients on anticoagulation therapy. CTA identified HALT of at least one leaflet in 5 patients while of all three leaflets in 2 patients (Figure 1). Leaflet rigidity was seen in 3 patients and in 90 % of the cusps where HALT was identified. Between the two patients without HALT, only one was on anticoagulant therapy. The mean pressure gradient was 10±3 mmHG on the echocardiogram at 6 months after the procedure. Only one patient with HALT developed mild-to-moderate pulmonary regurgitation. None of the patients showed clinical symptoms of aggravated heart failure or thromboembolism on CT.
Conclusions: The Venus P-valveTM is a self-expendable valve that can be implanted successfully in patients with significant pulmonary regurgitation and large RVOT. The incidence of HALT and RELM was significant in our population. Further studies are necessary to confirm this phenomenon in patients after TPVI with Venus p-valve, to identify any predictors and, to understand its clinical and evolutionary significance.

Keywords: Venus P-Valve, transcatheter pulmonary valve implantation, antithrombotic therapy

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PP-069

Agenesis of the left pericardium: And yet it moves!

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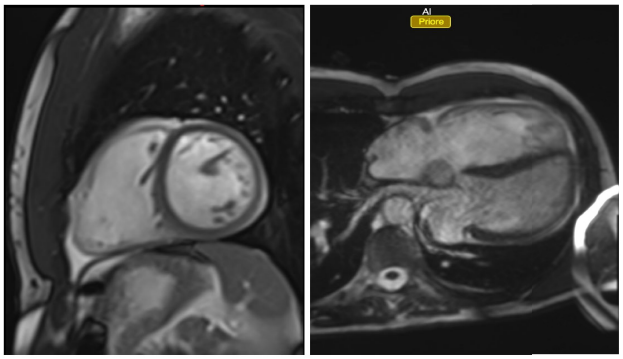
Background and Aim: We report a case of left pericardial agenesia in a 61 year old man presenting palpitations, without syncope. Clinical picture was not specific on clinical field: atypical chest pain, systolic murmur 2/6 Levine on the second left interspace were found.
Method: EKG showed sinus rhythm, clockwise deviation of axis in the precordial leads. Frequent isolated polymorphic premature extrabeats. Holter EKG was perfomed: frequent polymorphic extrabeats (9834/24 ore) with some couplets and triplets, without sustained ventricular tachycardia episodes. Starting metoprolol 100 mg per day the arrhythmic burden decreases significantly. Chest X-ray showed marked levocardia and dilatation of pulmonary trunk. Levoposition of the heart, lung interposition between the diaphragm and the base of the heart and between the aorta and pulmonary artery. Echocardiography showed a classic abnormal swinging motion of the heart in the chest, a paradoxical ventricular septal movement, apparent right-sided heart enlargement, with unusual bulging of the apex of the right ventricle.
Results: Magnetic resonance imaging (currently the gold standard for diagnosing) confirmed the absence of the left pericardium, and a left pericardial agenesia diagnosis was performed.

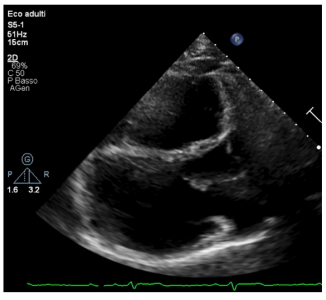
Left pericardial agenesis

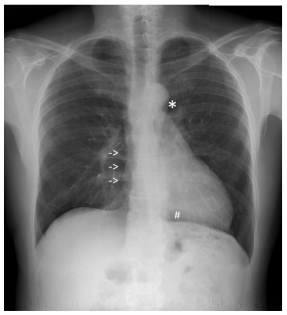
Cardiac magnetic resonance imaging short-axis view and for chambers view: *note the apex pointing posteriorly*
And dilation of right ventricle mimiking ARVD



Echocardiography *demonstrated a structurally and functionally normal heart with left heart deviation and no pericardial effusion.*

Posteroanterior chest X-ray: *overlap of the right cardiac border to the vertebral column(->), the main pulmonary artery convexity with interposition of lung tissue between them *, interposition of lung tissue between the diaphragm and the Heart #*





Conclusions: Patients have a similar life expectancy to those without pericardial defects; however in rare cases, herniation and strangulation of cardiac chambers can be life threatening and lead to sudden cardiac death.

Keywords: Left pericardial agenesis, Echocardiography, Magnetic Resonance Imaging

PP-070

4D flow MRI in patients with univentricular hearts following TCPC: Correlation of kinetic energy with other functionional properties

Anja Hanser[1], Melanie Hofmeister[1], Petros Martirosian[2], Fritz Schick[2], Michael Hofbeck[1], Andreas Hornung[1], Renate Kaulitz[1], Patrick Krumm[3], Konstantin Nikolaou[3], Jürgen Schäfer[3], Johannes Nordmeyer[1] and Ludger Sieverding[1]
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Background and Aim: In this study, we measured kinetic energy (KE) by means of 4D MRI in the ventricles of patients with univentricular hearts after total cavopulmonary connection (TCPC). The results were correlated to stroke volume (SV), enddiastolic volume (EDV), endsystolic volume (ESV), body surface area (BSA) and ejection fraction (EF).
Method: We prospectively examined 33 patients aged 19.8(14.6;30.2) years [median(Q1;Q3)] 14.3(9.7;24.9) years after TCPC surgery with a 4D-Flow sequence on a 3.0 T MRI scanner. Examinations in coronal orientation were performed with respiratory gating, voxel size 2.5 mm3, FoV 300 x 400 x 160 mm3 (covering the heart and the proximal great vessels). Scan time was 19:43 [15:01;22:35] mins. The ventricle was segmented and the kinetic energy was calculated using a post-processing software. Over one cardiac cycle 20 measuring points were calculated and divided in systole, early-diastole and late-diastole. Values of kinetic energy were correlated to stroke volume (SV), enddiastolic volume (EDV), endsystolic volume (ESV), body surface area (BSA) and ejection fraction (EF). Statistical analysis was performed using Pearson´s correlation (r) with r>0.5 indicating a significant correlation.
Results: Average KE over the entire cardiac cycle was correlated to SV (r=0.804*, p=<0.001), to EDV (r=0.674*, p=<0.001), to ESV (r = 0.516*, p=0.002), to BSA (r = 0.440, p=0.010) and to EF (r=-0.068, p=0.708). Average KE in systole was correlated to SV (r=0.695*, p=<0.001), to EDV (r=0.422, p=<0.015), to ESV (r=0.237, p=0.184), to BSA (r=0.456, p=0.008) and to EF (r=0.079, p=0.663). Average KE in diastole was correlated to SV (r=0.743*, p=<0.001), to EDV (r=0.775*, p=<0.001), to ESV (r=0.674*, p=<0.001), to BSA (r=0.328, p=0.063) and to EF (r=-0.203, p=0.258).
Conclusions: In our cohort, statistically significant correlations were found between KE and SV, EDV and ESV. Overall, the highest correlation was found between average KE over the entire cardiac cycle and SV. Thus, higher values of KE were associated with a larger stroke volume, and vice versa. Further investigations will be required to use KE as a new marker to describe Fontan haemodynamics.

Keywords: 4D-Flow, TCPC, kinetic energy

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PP-071

Assessment of the aortic wall parameters in children with normally functioning bicuspid aortic valve

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Background and Aim: Clinical presentation of bicuspid aortic valve (BAV) provides to BAV-related aortopathy. The gradual dilation of aortic dimensions increases the risk of dissection. Abnormal histopathological structure of the aortic wall may cause irregular haemodynamic parameters of the wall including distensibility, stiffness index and aortic strain.
Method: Study group (SG) of 46 children with BAV and control group (CG) of 34 healthy children didn't differ statistically for: age, body weight, height, BSA, HR or BPs and BPd. Both SG and CG were divided into 2 groups: under and over 11 years of age. The TTE protocol included M-mode scan of the aorta on the three levels: aortic root (AR), ascending aorta (AA) and aortic arch to assess systolic (AoDS) and diastolic (AoDD) diameters on every level and in 15 heart cycles. In offline analysis mean values of diastolic and systolic diameter for each level were obtained. Blood pressure (BPs and BPd) were measured during the M-mode recording. The Stiffness Index (SI), Distensibility Index (DI) and Aortic Strain for three levels were calculated based on formulas: DI=2(AoDS-AoDD)/((BPs-BPd)×AoDD), SI=logBPs/BPd/[((AoDS-AoDD))/AoDD]×10^(-3), Strain(%)=100×((AoDS-AoDD))/AoDD.
Results: Analysis revealed statistically higher aortic SI, reduced aortic DI and its Strain on the AR and AA levels in children with BAV compared to the CG in both younger and older children ($p<0.05$ for all analyses). No statistically significant differences were observed depending on sex ($p>0.05$). Comparing both groups of children with BAV: under and over 11 years of age, for aortic SI, DI and strain, no statistically significant differences were revealed for the aortic SI and strain, while the DI-2 was lower in older than in younger children ($p= 0.044$). A two-way analysis of variance revealed that the difference in the DI-2 level between older and younger children was higher in the CG than in the SG (weak/medium effect). A negative correlation was obtained between DI-2 and BSA, p=0.021, tau-b=-0.15.
Conclusions: Impaired parameters of the aortic wall in BAV subjects are present since the earliest years of life, and the DI of the AA level deteriorates with age. BSA affects the DI of the AA, which is a valuable indicator for formulating healthy lifestyle recommendations.

Keywords: Bicuspid aortic valve, stiffness index, distensibility index, aortic strain.

PP-074

Early detection of cardiac dysfunction in children with organic acidurias: An assessment using the two-dimensional strain

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Background and Aim: Organic acidurias (OADs) are primarily represented by propionic acidemia (PA) and methylmalonic acidemia (MMA). Cardiac complications are well-documented in propionic acidemia, and there are few reported cases of cardiomyopathies in methylmalonic acidemia. The aim of our study is to evaluate the left ventricle global longitudinal strain (LV GLS) in MMA and PA patients, compare it with the left ventricle ejection fraction (LVEF) method and compare it to the pediatric general population data.
Method: In this monocentric retrospective study, 26 OADs patients of la Timone University Hospital were included. Demographic, clinical, electrocardiographic and echocardiographic (LVEF, LV GLS) data from October 2022 to July 2023 were collected. Mean LV GLS in MMA and PA patients were compared to LV GLS in the pediatric general population.
Results: LVEF was similar between MMA and PA patients and in normal range (66,27 %± 6,24 vs 61,41 %± 11,02; $p = 0.18$). LV GLS was significantly lower in PA patients than in MMA patients (-15,82% ± 5,89 vs -21,19% ± 3,57; $p = 0.015$). LV GLS was significantly lower in PA patients than in the general pediatric population ($p = 0.03$).
Conclusions: To our knowledge, this is the first study comparing LV GLS between MMA and PA patients, and with the general pediatric population. Patients with propionic acidemia may have impaired global longitudinal strain even if their LVEF is normal. GLS is a useful tool for cardiac follow-up in pediatric patients with OA and could detect early cardiac dysfunction in those patients, leading to major therapeutic decisions.

Keywords: Organic aciduria, propionic academia, methylmalonic academia, global longitudinal strain, cardiac strain

PP-075

Potential use for strain in the diagnosis and follow up of kawasaki disease

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[1]Department Pediatric Cardiology, Hospital Universitario Puerta Del Mar, Cádiz, España; [2]Department Pediatric Nephrology, Hospital Universitario Puerta Del Mar, Cádiz, España

Background and Aim: The detection of myocardial dysfunction by conventional echocardiography is unusual and challenging in Kawasaki disease (KD). We present two cases where the measurement of global longitudinal strain of the left ventricle (GLS) by speckle tracking echocardiography (STE) allowed to detect subclinical myocardial dysfunction in both, acute and chronic stage of the disease.
Method: The first case was a 9-year-old female with a clinical kawa-shock, and mild coronary aneurysm at anterior descending coronary artery (ADCA) but normal EF (67%) on echocardiography. Subclinical myocardial dysfunction was proven by STE (Figure 1). Both findings resolved during acute phase after receiving standard Kawasaki therapy. (IVIG plus steroids). The second case was an 18-year-old male with a Kawa-shock when 8 years-old and a giant aneurysm in ADCA and a moderate aneurysm at circumflex coronary artery as sequelae. During follow-up he was always asymptomatic with normal serial echocardiograms (except for aneurysms), treadmill tests and cardiac MRI. Evaluation with STE demonstrated depressed myocardial function at territories irrigated by those coronaries (Figure 2).
Results: STE imaging has proven to be useful in the follow up and the diagnosis of myocardial affection in patients with Kawasaki disease where conventional echocardiography shows normal values even with coronary aneurysm. As we can see in the cases the affection of the myocardium corresponds with the cardiac magnetic resonance results as well as with the coronary affected which irrigates the territories where we can find the dysfunction.

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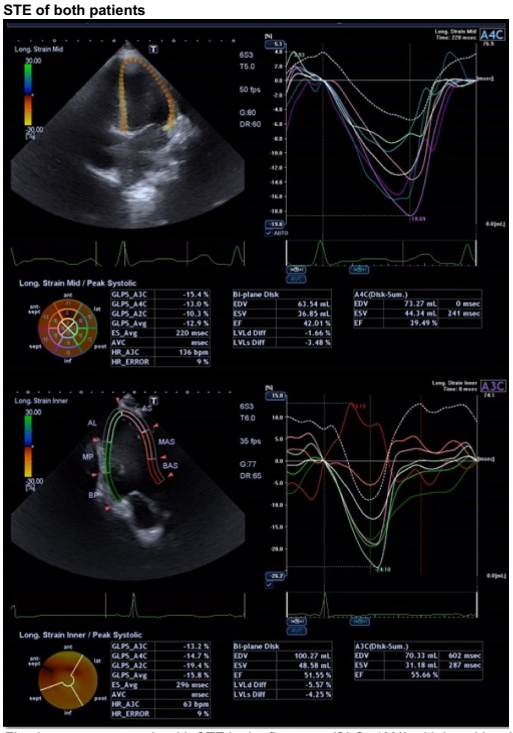
Conclusions: In Kawasaki disease the conventional echocardiography is normal in most of the patients, however the STE measurement allows to detect myocardial alterations previously unnoticed being more sensitive and a fundamental factor in the diagnosis of this and possibly other pathologies. In the follow up, it allows to detect systolic dysfunction early in these patients, detecting if they require myocardial remodelling treatment to maintain systolic function several years after the acute onset of the pathology. More studies are needed, with a longer follow-up and bigger sample size.

Keywords: Kawasaki disease, Stain, myocardial dysfunction, echocardiography, imaging.

STE of both patients



First image corresponds with STE in the first case (SLG -12%) with hypokinesia areas at anteroseptal and posteroinferior level (Corresponding with the anterior descending artery). Two weeks after the acute onset the STE normalized being the patient asymptomatic. Second image corresponds to the STE of the second patient after 10 years follow up still presenting GLS diminishment (-15%) and in the bull´s eye image red areas of ventricular disfunction in the affected coronary (Anterior descending and circumflex artery) areas of myocardial irrigation.

Cardiac Dysrhythmias and Electrophysiology

PP-076
Sotalol as treatment of short QT syndrome

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[1]Universidad Autonoma de Barcelona, Hospital del Oriente de Asturias, Hospital Central de Asturias, Oviedo, España; [2]Hospital Central de Asturias, Oviedo, España

Background and Aim: Short QT syndrome (SQTS) is a rare inheritable cardiac channelopathy presenting as a sudden cardiac death (SCD) in 34% of patients. SQTS is characterized by an abnormally short QT interval and an increased risk of ventricular and atrial arrhythmias. Diagnosis is based on an electrocardiogram, symptoms (palpitations or SCD) and genetic tests. Only 25% of patients have a positive genetic test. Five subtypes of SQTS have been described. ICD is the main treatment of these patients but in childhood pharmacology therapy could be indicated. Quinidine has been described as an effective treatment but has many side effects. Sotalol has also been used in these patients and with less side effects but is not recommended in SQTS1 (KCNH2 mutation). We studied the effect of sotalol in a patient diagnosed with SQTS.
Method: One patient of 2.5 years old with diagnose SQTS was analysed. The patient had been referred at age of 12 months because of a heart murmur and have been diagnosed with a muscular ventricular septal defect and short QT interval on electrocardiogram (338ms). The patient was asymptomatic. Genetic test showed KCNJ2 mutation (pAsp172As) (SQTS3). No family history of SCD was reported. Parents have a normal electrocardiogram. Quinidine was refused as an initial treatment because of its side effects. ICD was not indicated. We decided to use sotalol as a pharmacological treatment of SQTS.
Results: Sotalol was initiated a 30mg/m2/8h and increased since 35mg/m2/8h. Slight prolongation of QT interval was observed (2,9%) respect to baseline. No side effects were showed, including arrhythmic events.
Conclusions: SQTS is a rare cardiac channelopathy with an increased risk of ventricular and atrial arrhythmias. ICD is the mean treatment but in childhood pharmacological treatment could be used. Although quinidine is the most used drug in these cases, sotalol could become a competitive option especially in other subtypes of SQTS.

Keywords: SQTS, sotalol.

Cardiac Imaging

PP-077
4D Echocardiography: Left ventricle normal volumes

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Background and Aim: Left ventricle (LV) evaluation is essential for diagnosis, treatment, and prognostic of congenital heart diseases. 4D echocardiography (4D-ECHO) has become a significant advance in the field of cardiac imaging, being an innovative option in pediatric patients. However, there are few studies that include children and no standardized normality values are available. The main objective of this study was to establish normal LV values in pediatric patients using 4D-ECHO, compared to 2D-ECHO
Method: Observational, analytical, and cross-sectional study carried out in a tertiary children's cardiology hospital from November 2021 to May 2022. We included patients with a normal LV and weight less than 40 kg. Demographic, electrocardiographic and echocardiographic (M mode, 2D and 4D) data were collected.
Results: A total of 133 patients were scanned, 10 patients were excluded for not meeting the inclusion criteria, 11 had poor acoustic window and 4 were lost in the system. Finally, 108 patients were analyzed, 59% were male. Mean age was 5.4 years (3,9 SD). Mean heart rate was 100bpm (27 SD) and mean blood pressure was 102/60 mmHg (9.3/ 7.7SD), 97% had sinus rhythm.

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Normality values were established corrected for age, weight, height, and body surface area (by Haycock` formula).
Statistically significant correlation was observed between the measurement of LV volumes in 4D-ECHO and manual and semi-automatic 2D ($p<0.01$), as well as in the measurement of ejection fraction by 4D-ECHO with manual ($p=0.05$) and semi-automatic 2D ($p<0.01$).
Conclusions: Normal LV values were obtained in 4D-ECHO with good correlation with other echocardiographic modalities. The establishment of these values could be useful in the assessment of pediatric patients, due to the reproducibility and accessibility of this technique.

Keywords: left ventricle, 4D-echocardiography, 2D-ecochardiography

PP-078

Establishing the local paediatric reference range for T1, T2 parametric maps in a local chinese population

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Background and Aim: According to the joint consensus statement by the Society for Cardiovascular Magnetic Resonance (SCMR) and the European Association of Cardiovascular Imaging (EACVI) on cardiovascular magnetic resonance mapping of T1 and T2, it is recommended to generate machine-specific local reference ranges. We aim to do that at the local children's hospital, which is the tertiary referral center for all paediatric cardiology cases in the region.
Method: The Hong Kong Children's Hospital conducted a study to establish local reference ranges for T1 and T2 parametric maps on their 1.5T MRI system (Aera, Siemens Medical Solutions, Erlangen, Germany). They recruited 18 healthy pediatric Chinese volunteers, with a mean age of 10.5 +/- 3.2 years old. The participants' mean heart rate was 84 +/- 13.8 bpm. T1 mapping was performed using a modified Look-Locker inversion recovery (MOLLI) sequence, and T2 mapping was done using a gradient echo pulse sequence. Two experienced cardiac imagers analyzed the images.
Results: The mean T1 mapping value obtained was 981 +/- 47 msec (mea +/- standard deviation), while the mean T2 mapping value was 48 +/- 3 msec. The researchers compared their local mean T1 mapping values with those published by other centers and found them to be similar, although their standard deviation was larger due to the smaller sample size. The local T2 parametric mean and standard deviations were similar to published pediatric references.
Conclusions: In conclusion, this study provides local reference ranges for T1 and T2 parametric maps on a 1.5T MRI system using a cohort of healthy pediatric Chinese volunteers. These findings contribute to the quantitative usage and reporting of T1 and T2 mapping data in their clinical setting in Hong Kong.

Keywords: Parametric mapping T1, T2 reference range

PP-079

CMR predictors of the right ventricle end diastolic pressure in repaired tetralogy of fallot: A new CMR approach

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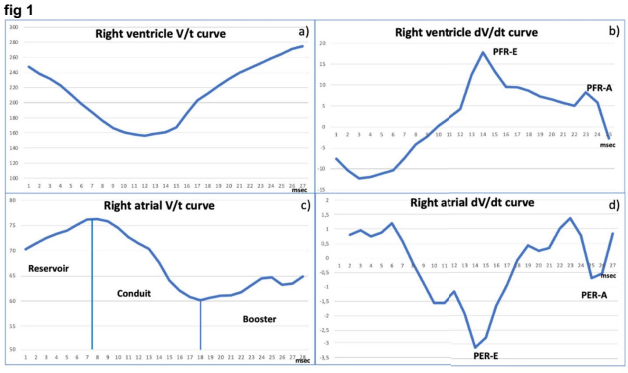
Background and Aim: BACKGROUND: In patients with repaired Tetralogy of Fallot (rTOF), poor outcomes are associated with progressive biventricular systolic and also diastolic dysfunction. However, definition of RV diastolic function is still challenging. AIM: To identify non-invasive CMR parameters predictors of RV end diastolic pressure (RVEDP) invasively measured at catheterization (CC), by introducing a new method of diastole evaluation, based on the simultaneous analysis of bi-atrial and bi-ventricular volumes variation across the cardiac cycle.
Method: We identified rTOF patients who underwent CC and CMR. In addition to standard CMR parameters, we analyzed diastolic function parameters from right and left atrial and ventricular volume/time (V/t) curves, derived from the volume curves (dV/dT) and correlated them with RVEDP (Fig.1). Furthermore, we identify CMR parameters associated with RVEDP $\geq$ 12 mmHg.
Results: This study included 35 patients (18 males, age 31 $\pm$ 15 years) with rTOF and 14 age-matched controls. Mean follow-up time at study entry from primary repair was 27.6 $\pm$ 11.6 years. Compared with control group, rTOF patients showed an opposite behavior of CMR parameters between right and left chambers: RV demonstrated prevalent systolic dysfunction (lower RVEF and elastance), whereas left chambers showed impaired LV filling, with significantly higher isovolumetric pulmonary veins transit ratio (IPTR). Moreover, a linear inverse relation was found between RVEDP and parameters describing both RV systolic function (i.e elastance and cardiac index) and diastolic function (RV atrial-peak filling rate and isovolumetric caval vein transit volume). Patients with RVEDP $\geq$ 12 mmHg showed abnormal RV

fig 1



PFR-E: early peak filling rate PFR-A: atrial peak filling rate
PER-E: early atrial peak empting rate PER-A: late atrial peak empting rate

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relaxation (lower RA emptying fraction; lower ICTV) and LV passive filling (greater LAVi conduit; higher LA early-peak emptying rate index). Based on univariate analysis, a score system was generated, including LA max Vi, LA reservoir peak Vi, LA conduit peak Vi, LA booster peak Vi. The score system demonstrated 100% sensitivity and 87% specificity to predict MACE.
Conclusions: The analysis of ventricular and atrial V/T curves showed pathophysiological insights of biventricular dysfunction in rTOF: progressive RV systolic dysfunction, while diastolic dysfunction is the main adaptation change of the left heart. Moreover, parameters of left atrial diastolic function are prognostically associated with MACE in this selected population.

Keywords: repaired Tetralogy of Fallot. diastolic function, cardiac magnetic resonance, end diastolic pressure

PP-080
Organisation of paediatric echocardiography laboratories and Governance of echocardiography services and training in Europe

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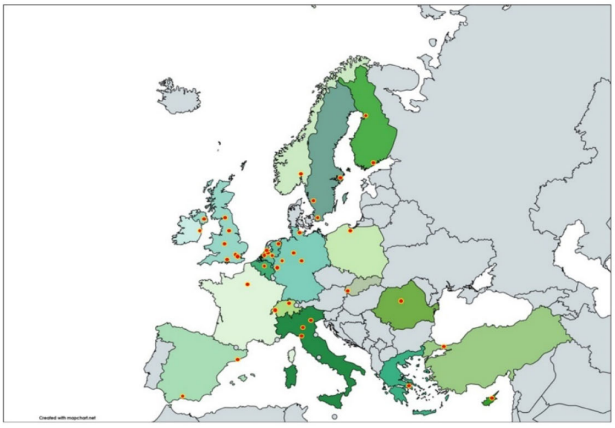
Background and Aim: There is limited data on the organisation of paediatric echocardiography laboratories in Europe. This study sought to assess the current status of provision of echocardiography services, training, quality improvement and research across European paediatric cardiology centres.
Method: A structured questionnaire was circulated across all 95 European Association of Paediatric Cardiology (AEPC) affiliated centres. The aims were to evaluate: 1) facilities in paediatric echocardiography laboratories across Europe, 2) accredited laboratories, 3) medical/paramedical staff employed, 4) time for echo studies and reporting, 5) training, teaching, quality improvement, and research programs.
Results: Respondents from forty-three centres (45%) in 22 countries completed the survey (Figure). Thirty-six centres (84%) have a dedicated paediatric echocardiography laboratory lab, only 5 (12%) of which were EACVI-AEPC accredited. The median number of echocardiography rooms was 3 (range 1-12), and echocardiography machines was 4 (range 1-12). Only half of all the centres have dedicated imaging physiologists and nursing staff, while the majority (79%) have specialist imaging cardiologist(s). The median (range) duration of time for a new examination was 45 (20-60) minutes, and for repeat examination was 20 (5 to 30) minutes. More than half of responders (58%) have dedicated time slots for reporting, while the others report at the end of the examination.
An organised training program was present in 33 centres (78%) with 9 centres (23%) having a training "bootcamp" for new starters, and 37% used a simulator for training. Forty-four percent undertake quality assurance, and 79% perform research, of which 42% was funded. Published research on imaging was achieved in 71% of centres. Most of the respondents (59%) believe that research in not well organized in Europe. They expressed a desire for improving research through greater cooperation among centres

Figure



Participating Echocardiography Centres

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with multicentre studies and registries, greater time for research, funding, standardization and guidelines in imaging. Guidelines for performing echocardiography were available in 32 centres (74%).
Conclusions: Facilities, staffing levels, study times, standards in teaching/training and quality assurance vary widely across paediatric echocardiography laboratories in Europe. Greater supports and investment to facilitate improvements in staffing levels, equipment and governance would potentially help improve European paediatric echocardiography laboratories.

Keywords: Paediatric Cardiology, Echocardiography, Laboratories, Training, Education, Governance

PP-081

ECHO- and electrocardiographic findings in newborns with inadequate intrauterine growth during pregnancy – a copenhagen baby heart study

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Background and Aim: Being born small for gestational age (SGA, birthweight <10th percentile) is associated with both increased neonatal mortality and cardiovascular disease later in life. Associations with fetal growth restriction (FGR, birthweight <3rd percentile) are particularly strong. Echocardiographic studies with up to 200 participants have shown that SGA and FGR are associated with structural and morphological alterations in the neonatal heart and fetal electrocardiography (ECG) studies have reported alterations in the fetal cardiac conduction system. We investigated associations between FGR, SGA, and echo- and electrocardiographic parameters in a large population-based cohort of newborns.
Method: The Copenhagen Baby Heart Study is a population-based cohort study with prenatal inclusion and postnatal cardiac examination including echocardiography and 8-lead ECG of more than 25,000 infants born in the period 2016-2018. Using multiple linear regression, we compared left ventricular echocardiographic parameters and ECG measurements in newborns with FGR (n=972) and SGA (birthweight ≥3rd and <10th percentile, n=2,106) with measurements in infants born appropriate for gestational age (AGA, birthweight ≥10th percentile and <90th percentile, n=20,468). We adjusted our analyses for sex, gestational age at birth, weight, length, and age at cardiac examination.
Results: Newborns with FGR or SGA had significantly smaller left ventricular (LV) posterior wall thicknesses and volumes, compared with infants born AGA (mean adjusted differences: -0.05 and -0.04 mm); interventricular septal diameter was markedly smaller in newborns with FGR than for infants born SGA (mean adjusted difference: -0.04 mm). Both groups also had shorter uncorrected QT intervals, although once QT intervals were corrected for heart rate, only FGR remained associated with shorter QT intervals (mean adjusted difference, QTc Fridericia: -2.32 ms). Infants born with SGA also tended to have reduced amplitudes of the R- and S-waves in V6 and larger amplitudes of the S-wave in V1 compared with children born AGA (Table 2).
Conclusions: Even after adjustment for their smaller sizes, newborns with FGR and SGA have smaller LV dimensions and differences in electrocardiographic parameters compared with infants born AGA. Follow-up examinations of these children are necessary to determine whether these abnormalities persist beyond the neonatal period and how they implicate cardiovascular health later in life.

Keywords: fetal growth restriction, intrauterine growth restriction, small for gestational age, growth restriction, ECG, echocardiography

Table 1 and Table 2

Table 1. Comparison of echocardiographic left ventricular measures in infants born with fetal growth restriction (n = 972) or small for gestational age (n = 2,106) and infants born appropriate for gestational age (n = 20,468).

	Fetal Growth Restriction		Small for Gestational Age	
	Mean adjusted difference (CI)	P-value	Mean adjusted difference (CI)	P-value
Heart rate (bpm)	0.37 (-1.15, 1.89)	0.63	1.23 (0.22, 2.24)	0.02
Stroke volume (mL)	-0.02 (-0.15, 0.11)	0.74	-0.09 (-0.17, 0)	0.05
Fractional shortening (%)	0.00 (0.00, 0.00)	0.94	0.00 (0.00, 0.00)	0.12
MV E (cm/s)	1.01 (0.07, 1.96)	0.04	-0.97 (0.34, 1.61)	<0.01
MV A (cm/s)	0.63 (-0.31, 1.58)	0.19	1.04 (0.4, 1.67)	<0.01
MV DecT (ms)	0.21 (0.00, 0.43)	0.05**	-0.03 (-0.18, 0.11)	0.67
LVPWd (mm)	-0.05 (-0.09, -0.01)	0.01	-0.04 (-0.06, -0.01)	<0.01
LVIDs (mm)*	-0.10 (-0.2, -0.01)	0.03	-0.13 (-0.2, -0.07)	<0.001
LVIDd (mm)*	-0.14 (-0.26, -0.03)	0.01	-0.16 (-0.23, -0.08)	<0.001
IVSd (mm)*	-0.07 (-0.11, -0.03)	<0.001*	-0.03 (-0.06, -0.01)	<0.01
ESV (mL)	-0.01 (-0.1, 0.07)	0.75	-0.08 (-0.14, -0.03)	<0.01
EDV (mL)	-0.05 (-0.23, 0.14)	0.61	-0.17 (-0.29, -0.05)	<0.01

95% confidence interval (CI), beats per minute (bpm), mitral valve early peak velocity (MV E), mitral valve atrial peak velocity (MV A), mitral valve deceleration time (MV DecT), left ventricle posterior wall an end-diastole (LVPWd), left ventricle internal diameter in end-systole (LVIDs), left ventricle internal diameter in end-diastole (LVIDd), interventricular septum in end-diastole (IVSd), end-systolic volume (ESV), end-diastolic volume (EDV).
* Estimates for fetal growth restriction is significantly lower than estimates for small for gestational age (<0.05)
** Estimates for fetal growth restriction is significantly higher than estimates for small for gestational age (<0.05)
Reference group: infants born appropriate for gestational age. All estimates are adjusted for newborn's age, weight, and length at the time of cardiac examination, gestational age at birth, and sex.

Table 2. Comparison of electrocardiographic measures in infants born with fetal growth restriction (n = 544) or small for gestational age (n = 1,384) and infants born appropriate for gestational age (n = 14,076).

	Fetal Growth Restriction		Small for Gestational Age	
	Mean adjusted difference (CI)	P-value	Mean adjusted difference (CI)	P-value
Heart rate (bpm)	2.55 (0.61, 4.5)	<0.01	2.58 (1.32, 3.84)	<0.001
QT interval (ms)	-3.46 (-5.87, -1.05)	<0.01	-1.76 (-3.33, -0.18)	0.03
QTc_Fridericia (ms)	-2.32 (-4.49, -0.15)*	0.04	-0.08 (-1.5, 1.33)	0.91
QTc_Bazett (ms)	-1.30 (-3.65, 1.04)	0.28	1.22 (-0.31, 2.75)	0.12
QRS duration (ms)	-0.42 (-0.98, 0.14)	0.14	-0.36 (-0.73, 0.01)	0.05
PR interval (ms)	0.79 (-0.29, 1.88)	0.15	0.06 (-0.64, 0.77)	0.86
QRS axis (degrees)[b]	0.00 (-0.02, 0.03)	0.72	0.00 (-0.02, 0.01)	0.70
Max amplitude of the R-wave in V1 (uV)[b]	0.00 (-0.05, 0.04)	0.85	-0.01 (-0.04, 0.02)	0.64
Max amplitude of the R-wave in V6 (uV)[c]	-0.56 (-1.25, 0.12)	0.10	-0.63 (-1.07, -0.19)	<0.01
Max amplitude of the S-wave in V1 (uV)[d]	0.25 (-0.06, 0.56)	0.11	0.27 (0.07, 0.47)	<0.01
Max amplitude of the S-wave in V6 (uV)[b]	-0.04 (-0.11, 0.03)	0.32	-0.07 (-0.12, -0.02)	<0.01

95% confidence interval (CI), beats per minute (bpm)
* Estimates for fetal growth restriction is significantly lower than estimates for small for gestational age (<0.05)
[a] Due to skewness of some of our variables, transformations were necessary to obtain a normal distribution
[b] Linear scale
[c] Boxcox-transformed to the power of 0.47
[d] Boxcox-transformed to the power of 0.37
Reference group: infants born appropriate for gestational age. All estimates are adjusted for the newborn's age, weight, and length at the time of cardiac examination, gestational age at birth, and sex.

Table 1. Comparison of echocardiographic left ventricular measures in infants born with fetal growth restriction (n = 972) or small for gestational age (n = 2,106) and infants born appropriate for gestational age (n = 20,468 Table 2. Comparison of electrocardiographic measures in infants born with fetal growth restriction (n = 544) or small for gestational age (n = 1,384) and infants born appropriate for gestational age (n = 14,076).

PP-082

Fetal 3D Cine cardiovascular mri: improved diagnostic quality with region-optimized virtual coils

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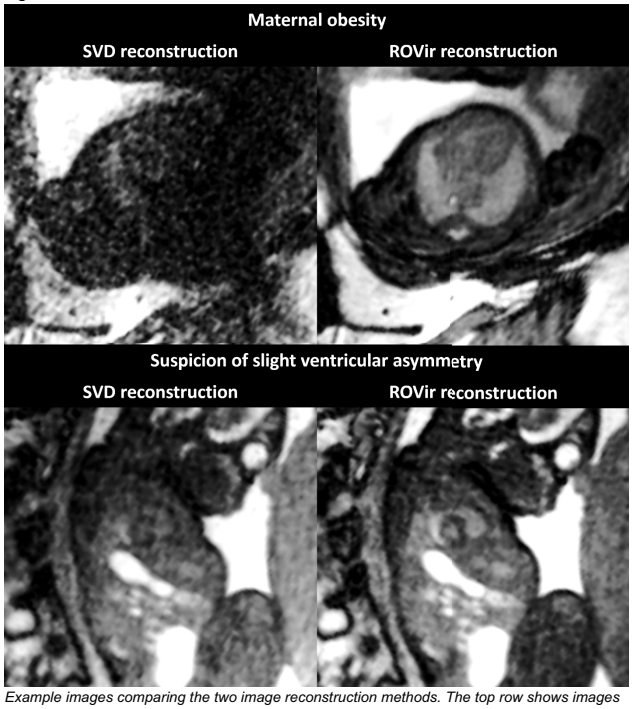
Background and Aim: Fetal cardiovascular magnetic resonance (CMR) imaging can assess congenital heart defects prenatally. The first true fetal CMR 3D cine acquisition was developed to overcome image planning challenges and to reduce examination time (Piek et al., MRM 2022). However, increased image contrast and temporal resolution are needed to translate the method to clinical routine. The aim was to apply the Region-Optimized Virtual (ROVir) coil combination method (Kim et al., MRM 2020) to the true fetal 3D cine CMR acquisition as to highlight CMR signal from the fetus and particularly the fetal heart while suppressing artefacts originating from maternal tissue and particularly fat, which in turn is expected to increase diagnostic quality.
Method: Fetal CMR 3D cine data were acquired as previously described (Piek et al., MRM 2022). In short, a transversal radial slab-selective balanced steady-state free precession research sequence with isotropic 1.9mm voxels was used (1.5T MAGNETOM Aera, Siemens Healthineers, Germany) with retrospective fetal cardiac gating (Northh medical GmbH, Germany). Images were reconstructed using BART v0.8.00 (Uecker et al., ISMRM23:2486) and Matlab R2022b (The MathWorks, USA), on a 16-core computing server. Images were reconstructed from the same acquired data both with the standard singular-value decomposition (SVD) and the ROVir methods.

Figure 1



Example images comparing the two image reconstruction methods. The top row shows images of a healthy fetus reconstructed using the SVD (left) and ROVir (right) methods in presence of maternal obesity. The bottom row shows images reconstructed using the SVD (left) and ROVir (right) methods in a fetus with suspicion of slight ventricular asymmetry. Note the improved image quality when using the ROVir method compared with the standard reconstruction using SVD.

Diagnostic quality with regards to tissue contrast between blood pool and myocardium, function assessment, and presence of artefacts was graded 1–4; 4='high diagnostic quality', 3='acceptable diagnostic quality', 2='low diagnostic quality', and 1=insufficient diagnostic quality'.
Results: Seven fetuses were included (gestational week 32–38). The ROVir reconstruction doubled temporal resolution from 66 ms by SVD versus 33 ms by ROVir (i.e. 8 versus 16 time-frames for one cardiac cycle). Figure 1 shows image quality for the standard SVD and ROVir reconstructions. Diagnostic quality was higher for ROVir vs SVD reconstructions (3 [2.5–3] vs 2 [1–2.5]; p=0.03).
Conclusions: Diagnostic quality and temporal resolution increased using the ROVir reconstruction method. The improved image contrast may be related to reduction of noise-like artefacts which in turn also increases the perceived contrast-to-noise ratio. This allows for improved appreciation of fetal cardiac function and improved diagnostic quality.

Keywords: Prenatal diagnosis, CHD, 3D cine fetal CMR.

PP-083
Developing a new echo based 3-D-haemodynamic-simulation software - first results determining the accuracy of simulated QP:QS in Asd patients

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Background and Aim: With our vision of a simple, echo-based method to a more personalised, but still time- and cost-efficient digital solution for paediatric cardiology, we developed a virtual haemodynamic 3-D heart model (Figure 1). Measuring the shunt volume (Qp:Qs) in transthoracic echocardiography (TTE) has shown to overestimate Qp:Qs. We aim for better anatomical visualization of the defect and haemodynamic assessment of cardiac load via Qp:Qs calculation using a virtual 3-D model. This can improve the understanding of the pathology for patients, parents and medical professionals using.
Method: A single centre retrospective study was conducted. ASD patients planned for interventional closure via cardiac catheterisation at our institution were included (n=10, mean age: 16.26 (3.36 - 57.92), mean weight: 33.53 kg (13.4kg – 70kg). Qp:Qs was determined using echocardiography by measuring left and right ventricular outflow tract diameters and the left and right subvalvular velocity time integral. A 3-D-heamodynamic-heart model was created for every patient. The Qp:Qs calculated by the 3-D-model was then compared to the Qp:Qs from the echocardiography.
Results: The analysis has shown a good agreement between the measured shunt volume in echocardiography (mean 1.87 ± 0.49 with a median value of 1.74) and the calculated shunt volume of the 3-D-model (mean 1.67 ± 0.4 and a median value of 1.67) with a mean deviation of -8.2%.
Conclusions: The first "real-world" results of our echocardiography based 3-D-haemodynamic-simulationsoftware demonstrates encouraging preliminary results for calculating valid Qp:Qs values in ASD patients. Inclusion of more patients is ongoing to prove the statistical significance of the calculated shunt volumes. After further fine-tuning of the model, bigger multicentre studies are planned for clinical validation. The individual illustration of the pathology and the surrounding structures can improve the visualization thus the understanding of the pathology for physicians, students and

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patients. It is an innovative, fast, non-invasive, and inexpensive method, which can be used routinely as a multifunctional assistant for TTE.

Keywords: 3-D Haemodynamic Simulation, Echocardiography simulator, echocardiography 3-D simulation software, digital cardiac model, digital twin

Figure 1&2

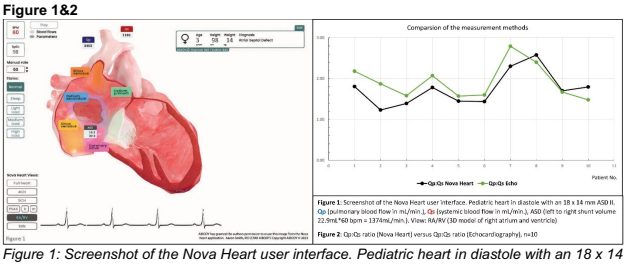

*Figure 1: Screenshot of the Nova Heart user interface. Pediatric heart in diastole with an 18 x 14 mm ASD II. Qp (pulmonary blood flow in mL/min.), Qs (systemic blood flow in mL/min.), ASD (left to right shunt volume 22.9mL*60 bpm = 1374mL/min.). RA/RV (3D simulation of right atrium and ventricle) Figure 2: Line graph: Qp:Qs ratio - Nova Heart versus Qp:Qs ratio – Echocardiography of 10 patients*

PP-084

A unique case of vascular ring with subclavian coarctation

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Background and Aim: This case study is intended to share how subclavian coarctation may occur simultaneously with a vascular ring. This patient was originally referred as an outpatient for difficulty taking solid feeds with echocardiogram results suggestive of a right aortic arch from a prior evaluation. The child had been temporarily transitioned to pureed foods because of the choking with solid foods. She did also have a congenital scoliosis secondary to a spinal abnormality which was being treated via bracing, through pediatric orthopedics. Given speech delays, she was also referred to genetics, whose evaluation was pending as of the time of writing of this article. Physical examination was unrevealing. Echocardiogram and cardiac CT were performed.
Method: Anytime a right aortic arch is found on echocardiography with unclear assessment of the remainder of the head vessel branching, congenital heart lesions and vascular rings should be considered, especially with the history of repeated difficulty taking solid foods. Right aortic arch with aberrant left subclavian, double aortic arch, Tetralogy of Fallot, and other complete vascular rings are highest in the list of considerations. EKG was unrevealing. Echocardiogram was performed, which was suggestive of a right aortic arch however not all branches of the transverse aorta could be clearly visualized.
Results: She was referred for a cardiac CT to give definitive anatomic analysis of the aortic arch. The CT revealed an aberrant left subclavian, R aortic arch, left subclavian coarctation versus isolation, and a diminished caliber of the entire left subclavian.
Conclusions: The patient was referred for surgical release of the vascular ring. While it is typical for the right aortic arch and aberrant left subclavian to be a vascular ring in the presence of a left sided patent arterial duct, it is less common for there to be an outright subclavian coarctation with diminished LSCA caliber. This diagnosis should be considered if the ligamentum arteriosum is tight enough. The left subclavian did receive sufficient collateral flow from the vertebral vessels, and so the cardiology team did not recommend reimplantation given no functional limitation of the left arm.

Keywords: Vascular ring, subclavian coarctation

Right aortic arch, aberrant left subclavian, with subclavian coarctation

Right aortic arch, aberrant left subclavian, with subclavian coarctation

PP-085

Impaired cardiopulmonary exercise capacity in paediatric fontan circulation - relation to pulmonary CMR 4D flow

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Background and Aim: Patients with Fontan circulation experience a number of complications and comorbidities across a broad range of end-organ systems. They show a restricted exercise ability, and complications are closely linked to hemodynamic regulation of the pulmonary blood flow and congestion of the venous system. The aim of this study was to assess whether the pulmonary circulation in children with Fontan type palliation, as measured by CMR 4D flow, is associated with maximal oxygen uptake (VO2max) as measured by cardiopulmonary exercise test (CPET).
Method: Twenty-one paediatric patients with Fontan circulation admitted for annual cardiac follow up, were prospectively included. Cardiac MR (CMR) including cardiac function, 4D flow with total assessment of the Fontan circuit and CPET were performed. Non-parametric correlations between flow, cardiac function and exercise capacity were determined.
Results: Demographics and results are presented in Table 1. Thirteen out of 21 patients performed both CPET and CMR. VO2max was low as compared to normal reference standard, but as expected for this patient group.

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The pulmonary circulation as measured by 4D flow Qpv:Qs, was associated to VO2max (R 0.7, p=0.02). Other specific 4D-flow measurements, such as in the Fontan-tunnel, were not associated with VO2max. However, single ventricle enddiastolic volume (EDVI) was strongly associated with VO2max (R 0.7, p=0.03).
Conclusions: This small study show low oxygen uptake in children with Fontan circulation. CMR 4D flow enabling multiflow assessment, revealed a low Qpv:Qs as a marker of imbalance in the pulmonary flow, which was associated with decreased oxygen uptake, as was low EDVI. These findings indicate the importance of both pulmonary flow balance and ventricular volumes for exercise capacity in these patients.

Keywords: CMR, Fontan, CPET, 4D flow

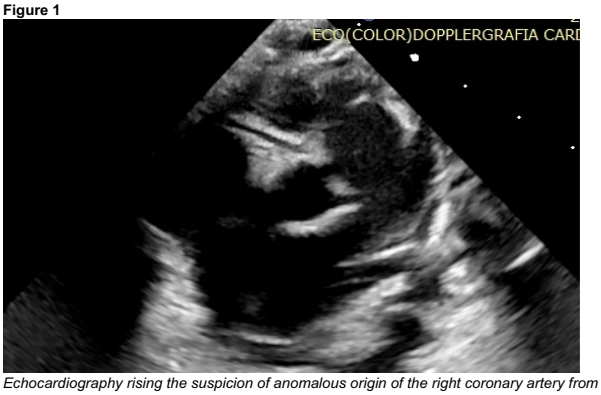
Table 1

Table 1	
Demographics (n =21)	Mean ± SD (range)
Age (years)	13.9 ± 4.7 (3-18)
Height (cm)	152±32
Weight (kg)	48±22
BSA m^2	1.42±0.46
Time since Fontan (days)	3683±1700 (148-5664)
CPET (n=13)	
VO2max (ml/kg/min)	26±5 (19-33)
Load max (W/kg)	2.1±0.6 (1.3-3)
RER max	1.10±0.06 (10.3-23.0)
HR max (beats/min)	164±21 (125-199)
VE/VCO2-slope	37±8 (29-53)
Cardiac MRI (n=18)	
Single Ventricle EDVIndex (ml/m^2)	92 ±23
Single Ventricle ESVIndex (ml/m^2)	48±18
Single Ventricle SVIndex (ml/m^2)	48±14
EF (%)	52±9
Flow aorta (L/min)	3.8±0.3
Flow superior vena cava (L/min)	1.4±0.4
Flow Tunnel (L/min)	2.1±1.1
Right pulmonary artery	1.5±0.7
Left pulmonary artery	1.2±0.6
Flow Pulmonary veins (L/min)	3.6±1.3
Opv:Qs	0.96±1.3

examination revealed dry and dehydrated skin, tachypneea (respiratory rate=50/min), along with an systolic cardiac murmur (V/VI) at the left sternal border. Point-of-care analysis showed increased increased NTproBNP 11.000pg/ml, negative Troponine, while central laborabory tests revealed anaemia (Hb=9g/dl) and no inflamatory syndrome. The electrocardiography was unremarkable with synus tachycardia. The transthoracic echocardiography showed aorto-pulmonary window, small atrial septal defect (4 mm), diastolic flow in the interventricular septum and dilation of the left coronary artery (3 mm). Additionally, suspicion of anomalous origin of the right coronary artery from the pulmonary trunk was emerged (Figure 1).
Results: Patient was referred to cardiac surgery clinic, and the diagnostic of ARCAPA was confirmed intraoperatory. The aorto-pulmonary window was closed along with the right coronary artery reimplantion in the anterior wall of the aorta.
Conclusions: This case is raising the importance of making a full echocardiographyc study, by evaluating also the coronary arteries when there is association with other cardiac deffects, such as aorto-pulmonary window. Other important echocardiographic sign in ARCAPA can be the presence of diastolic flow in the interventricular septum (which can explain collateral coronary vessel circulation).

Keywords: ARCAPA, aortopulmonary window, diastolic flow

Figure 1



Echocardiography rising the suspicion of anomalous origin of the right coronary artery from the pulmonary trunk

PP-086

A case of anomalous origin of the right coronary artery originating from the pulmonary trunk in the presence of aorto-pulmonary window

Georgiana Maria Militaru[1], Eliza Cinteza[2], Georgiana Nicolae[2], Gabriela Duica[2], Simona Pascu[1], Andreea Botez[1], Amelia Aria[1] and Alin Nicolescu[1]
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Background and Aim: Anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA) is a rare condition, with prevalence reported less than 0.002%. ARCAPA diagnosis requires expertise, as frequently the diagnosis pathway is challenging, in presence of low burden of symptoms.
Method: We present the case of a twenty days old newborn, referred to our clinic for feeding and breathing difficulties with progressive onset started from day 7. The general clinical

PP-087

A case report of mitral valve apparatus calcification in a childhood cancer patient treated with chemotherapy

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Background and Aim: Cardiovascular disease following cancer treatment with radiation and chemotherapy, in childhood, is a well recognised complication. These complications may not manifest for many years, especially after receiving radiation in the chest area. Manifestations such as heart failure and valvular disease may be seen earlier with chemotherapy.
Method: We describe a case of mitral valve (MV) apparatus calcification sparing the leaflets, in a patient previously treated for acute lymphoblastic leukaemia (ALL) with chemotherapy.
Results: A 2-year-old girl was diagnosed with B-cell ALL. Echocardiography before treatment showed a structurally normal heart with good biventricular function. The mitral valve (MV) apparatus was normal with no evidence of stenosis or regurgitation.

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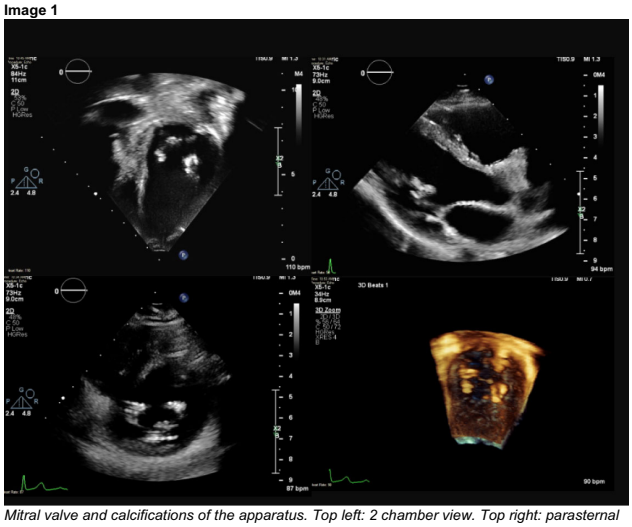
She was treated with dexamethasone, vincristine, daunorubicin, pegaspargase and intrathecal methotrexate.
Her echocardiogram was repeated 4 months later and showed multiple small, calcified areas on the chordae and tips of the papillary muscles (PMs) of the MV with normal function of the valve, no stenosis or regurgitation. Cardiac function remained normal.
The patient had frequent repeat echocardiograms which showed similar appearances, with an otherwise well-functioning MV. Four years later, echocardiographic findings remain unchanged, with calcifications persisting on the chords and PMs but no compromise of the valve function. Cardiac function has remained normal.
Conclusions: Valvular disease post chemotherapy seems to affect the mitral valve more frequently, however there are very few reported cases in children. We have identified two case reports of extensive cardiac calcifications, including the MV, in a background of ALL, both being at the acute phase and during induction therapy; one patient had sepsis.
In our case, the patient developed isolated MV apparatus calcifications but no other complications and continues to be asymptomatic with no haemodynamic compromise on follow up.

Keywords: mitral valve calcification, chemotherapy, ALL

Image 1



Mitral valve and calcifications of the apparatus. Top left: 2 chamber view. Top right: parasternal long axis view. Bottom left: parasternal short axis view. Bottom right: 3D view

PP-088
Comparing 3D cardiovascular mr angiography with 3D bssfp whole heart imaging in congenital heart diseases: A react study

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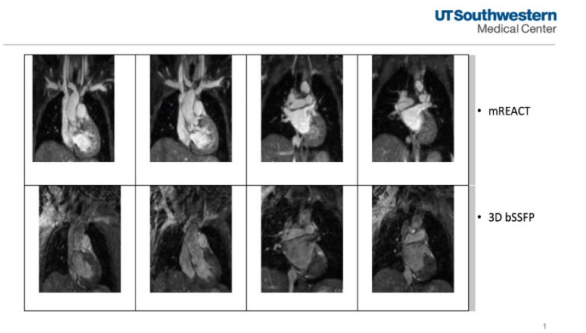
Background and Aim: 3-Dimensional Whole Heart balanced Steady-State-Free-Precession (3D bSSFP) Magnetic Resonance (MR) imaging is reliable for congenital heart disease (CHD) assessment, but field inhomogeneity-induced banding artifacts limit its utility. This study assesses a 3D Whole Heart approach utilizing a modified REACT (Relaxation Enhanced Angiography without Contrast and Triggering) technology to enhance image quality (IQ) for cardiac cardiovascular anatomy compared to conventional 3D bSSFP imaging.
Method: All scans were performed on a clinical whole-body 1.5 T CMR system (Ingenia Philips Healthcare, Best, Netherlands). The protocol included a modified REACT sequence and a standard 3D SSFP sequence. The REACT sequence is modified with EKG triggering in mid-diastole and respiratory navigator gating. The 3D single-phase SSFP sequence was respiratory-gated and ECG-triggered at diastole's end. The study involved 11 children with CHD, IQ and contrast to noise ratios (CNR) for cardiac structures and vessels. Pediatric cardiologists independently graded IQ using a five-point system, with consensus grades. Cross-sectional measurements cardiac structures and vessels were compared between sequences using the Wilcoxon test for categorical data and t-test for normally distributed variables.
Results: Mean CNR values significantly varied between the REACT sequence and the 3D bSSFP Whole heart sequence for different cardiac structures. Specifically, the REACT sequence had higher CNR ratios in the right upper pulmonary vein (4.7 vs. 1.9), left lower pulmonary vein (7.8 vs. 4.1), and left atrium (11.4 vs. 9.3), all with $p<0.05$. Conversely, the 3D whole heart pulse sequence showed significantly greater CNR in the right ventricle (14.9 vs. 11.8, $p=0.03$). Regarding image quality, 3D whole heart imaging outperformed the REACT sequence in several areas, including the right atrium, right ventricle, main pulmonary artery, left pulmonary artery, left atrium, and left coronary artery, all with highly significant differences ($p<0.05$). In contrast, the REACT sequence demonstrated superior IQ compared to 3D WH for the aortic valve ($p=0.04$). Although the REACT sequence also showed better IQ at pulmonary veins and the ascending aorta, these differences were not statistically significant. All cross-sectional measurements remained consistent between the two sequences.
Conclusions: Modified REACT improves cardiovascular imaging in CHD particularly of the pulmonary veins and the aorta compared to traditional 3D bSSFP imaging.

Keywords: Magnetic resonance, image quality, REACT, 3D bSSFP.

Ascending aorta and pulmonary veins comparison between REACT and 3D b SSFP



REACT-CMRA provides better image quality particularly for the pulmonary veins. Although 3D bSSFP whole-heart imaging can visualize the thoracic vasculature, enhanced visibility of the ascending aorta can be attained using REACT.

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PP-089

Large tricuspid tumor in a child: Blood CYST

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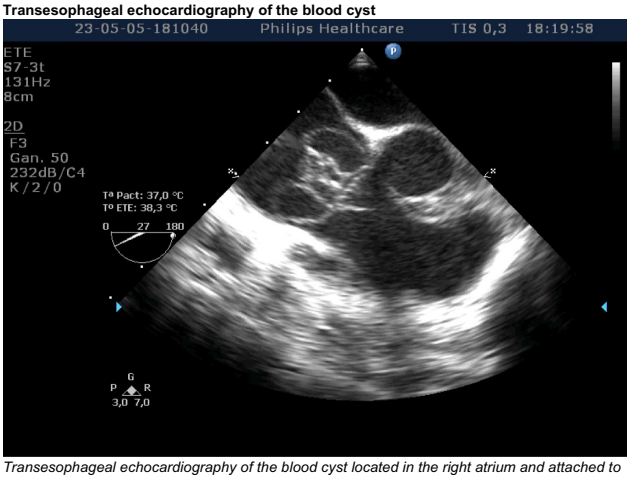
Background and Aim: Blood cysts of the heart are rare in adults. They are relatively common in newborns, but in the majority of cases they disappear spontaneously before the age of 6 months. They are usually found in the heart valves. Most often blood cysts are asymptomatic and are discovered incidentally on echocardiography performed for other reasons. Occasionally, they may cause valve dysfunction, and rarely embolization has been reported.
Method: We report a case of a 5-year-old male patient, asymptomatic, that attended consultations to rule out cardiac disease due to pectus excavatum.The patient had already attended at one month of age due to a murmur, with normal echocardiography. In the transthoracic ultrasound, a multicystic hypermobile mass was observed in the right atrium adhered to the tricuspid valve with a dimension of 2.5 cm, which generated mild tricuspid insufficiency. Transesophageal echocardiography was performed to better identify the mass in the right atrium, showing its adhesion to the septal leaflet of the tricuspid valve, occupying almost the entire valve orifice, with no associated lesions except for mild tricuspid insufficiency.
Results: Given the size of the mass and the possibility of embolization, surgical resection was performed, which passed without incident. A multilobulated pedunculated tumor with a cystic appearance with blood content and implantation in the septal leaflet of the tricuspid of about 3x2cm was resected, with no residual lesion in the tricuspid valve. Given the suspicion of a blood cyst, a sample is sent to pathology that confirms the diagnosis.
Conclusions: Blood cysts of cardiac valves are un unusual finding but may cause life-threatening complications, based in its location and size. Because this is a rare finding, management should be individualized based on presence of symptoms and complications associated with the intracardiac cyst.

Keywords: Echocardiography, Tricuspid valve blood cyst

Transesophageal echocardiography of the blood cyst



Transesophageal echocardiography of the blood cyst located in the right atrium and attached to the tricuspid valve.

PP-090

The utility of three-dimensional strain echocardiography in the assessment of left ventricular function in children with benign arrhythmia

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Background and Aim: Strain echocardiography is sensitive method of functional cardiac assessment, however the data on three-dimensional (3D) strain utility in children are still scarse. Paroxysmal non sustained supraventricular tachycardia and asymptomatic single ventricular extrasystoly in children are considered benign arrhythmias not affecting left ventricular function. The aim of the study was to determine utility and sensitivity of left ventricular function assessment in 3D-strain echocardiograpy in patients with benign arrhythmia in comparison with healthy population.
Method: Prospective study enrolled consecutive children with structurally healthy hearts and history of non sustained supraventricular tachycardia (nsSVT) in the past (no preexcitation in ECG) or with asymptomatic ventricular extrasystoly (VEx, monomorphic single beats, <5% in 24 hours). In all patients 3D-echocardiographic records were obtained and left ventricular function was assessed using 4D LV-Analysis software (Philips Medical Systems, USA) to calculate 3D average global longitudinal, circumferential and radial strain (LV-GLS, -GCS and -GRS). The results were compared with healthy children matched by age and sex.
Results: The studied group consisted of 20 children with nsSVT (aged 7 -17 years, mean 12+4.6 years old) and 20 children with VEx (aged 6 -15 years, mean 11+4.5 years old). All patients had normal left ventricular ejection fraction (LVEF) in standard echocardiography. Also in all children 3D LV-GLS, -GCS and -GRS were within normal limits, however in comparison with healthy patients the subgroup with nsSVT showed significantly lower values of LV-GLS (-21.7+4.6% vs -24.8+3.8%, p=0.007) and LV-GRS (-40.5+5.4% vs -45.4+5.9%, p=0.010) while children with VEx had lower LV-GLS (-20.7+6.1%, p=0.030), LV-GRS (-40.0 +7.3%, p=0.015) and LV-GCS (-25.1+4.4% vs -28.2+4.8%, p=0.019).
Conclusions: Three-dimensional strain echocardiography is effective and sensitive method for the assessment of left ventricular function. Even in benign arrhythmias in children with structurally healthy hearts subclinical dysfunction of left ventricle may be detected.

Keywords: Three-dimensional strain echocardiography, arrhythmia, supraventricular tachycardia, ventricular extrasystole

PP-091

The impact of percutaneous transcatheter closure of atrial septal defect on echocardiographic right ventricular strain indices in children

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Background and Aim: Speckle-tracking echocardiography is used for the assessment of right ventricular (RV) function. Echocardiographic indices can be influenced by changes in loading condition. Atrial septal defect (ASD) is associated with right-sided chronic volume overload.The aim of the study is to assess the relationship between volume overload and echocardiographic strain indices of RV function in children with ostium secundum ASD and the impact of the percutaneous transcatheter closure of ASD on the echocardiographic indices.
Method: We performed a prospective study by assessing 17 consecutive children with ASD who underwent percutaneous transcatheter closure and 17 sex and age matched controls. Conventional and speckle-tracking echocardiography was performed in ASD children prior to catheterization, and then 24 hours and one month after ASD closure. 2D RV free wall (RVFWLS) and RV global longitudinal strain (RVGLS) were performed by autostrain method. The same parameters were assessed in control group.
Results: The mean age of the patients was 11.64±3.58 years (5.07 – 16.75), and the mean diameter of the occlusive devices was 17.70 +/-4.48 mm (10.5 – 27). TAPSE z score, right atrial area z score, RV tissue Doppler peak systolic velocity z score was significantly higher in ASD group in comparison with control group. RVFWLS and RVGLS were significantly higher in absolute value in ASD group compared to control group (-33.57±3.24 vs -28.25 ± 2.89, p=0.03; respectively -28.56±2.50 vs -23.30 ±2.66, p=0.008). We noticed a progressive decrease of the strain parameters at 24 hours and at one month after ASD closure to a level comparable to the control group. RVFWLS and RVGLS decreased significantly at one month after ASD closure compared to preclosure status (-33.57 ± 3.24 vs -28.29 ± 5.93, p=0.044 respectively -28.25 ±2.89 vs -23.82 ± 5.15, p=0.036).
Conclusions: The echocardiographic strain indices of RV function can be enhanced by right-sided chronic volume overload. RVFWLS and RVGLS can assess the response of the RV to correction of volume overload after transcatheter closure of ASD in children.
Aknowledgement: This work was supported by the University of Medicine, Pharmacy, Science and Technology "George Emil Palade" of Târgu Mureş, Research Grant number 511/5/17.01.2022.

Keywords: speckle-tracking echocardiography, right ventricle, atrial septal defect, percutaneous transcatheter closure, children

PP-093
3D reconstruction and rapid prototyping for treatment planning in congenital heart disease

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Background and Aim: One-third of children with congenital heart defects (CHD) require immediate post-birth open-heart surgery, facing challenges due to the vast variability in anatomical features and the heterogeneous nature of CHD. Personalized treatment strategies, heavily reliant on medical imaging for accurate anatomical assessment, are critical for successful diagnosis and surgical planning. Recent advancements in 3D reconstruction of patient-specific anatomy have shown promise in enhancing these processes. These reconstructions allow for detailed visualization using computer graphics, virtual reality, and life-size 3D-printed models. Despite its potential, clinical application of this approach faces hurdles, primarily due to the lack of standardized reconstruction workflows. Our study aims to establish standard operating procedures (SOPs) for this purpose, focusing on Double Outlet Right Ventricle (DORV) patients as a case study.
Method: The study begins with structured interviews with paediatric cardiologists and cardiac surgeons to identify prevailing challenges and requirements for 3D CHD reconstruction and visualization. Simultaneously, existing evidence for the use of 3D reconstructions for diagnosis therapy planning, but also improving patient-caregiver interaction will be collected via a systematic literature review. Following this, CT, or MRI datasets from DORV patients are collected from clinical repositories, evaluated for image quality, and lesion complexity. We will reconstruct patient-specific anatomies from these datasets, documenting required time and procedural complexity. This will lead to the development of preliminary SOPs for reconstruction, visualization, and 3D printing of patient-specific anatomies. The SOPs' efficacy will be tested by having users of varying experience levels perform reconstructions following the SOPs. This will also allow to assess inter- and intra-observer variability to quantify the robustness of reconstructions.
Results: The study has not yet started. Therefore no results are available yet.
Conclusions: Successful validation of these SOPs, with manageable operator biases, will pave the way for a prospective, randomized trial to explore the benefits of 3D reconstruction, visualization, and printing in the management and treatment of DORV patients.

Keywords: double outlet right ventricle, congenital heart disease, 3D printing, therapy planning, diagnosis

PP-094
Echocardiographic evidence of early lv dysfunction progression in duchenne muscular dystrophy– case control study

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Background and Aim: Duchenne muscular dystrophy (DMD) is a progressive rare genetic disorder that leads to a lack of dystrophin, causing (among other morbidities) cardiomyopathy and overt heart failure in early adulthood, but sometimes in the teenagerhood. Yearly heart ultrasounds are advised from diagnosis, though their predictive value remains unclear. This study aimed to assess echocardiography results in DMD children.
Method: For measurements, a standard heart ultrasound technique was employed, incorporating B-mode and M-mode (Teichholtz) imaging, along with Doppler (to rule out comorbidities). As most ultrasound indices align best with body surface area (BSA), BSA-matched healthy controls were chosen for comparison. Thresholds of BSA 0.88m2 and 1.30m2 were used to result in 3 equally seized strata (subgroups: Early, Intermediate, Late). BSA strata was also

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considered as proxy for age and thus disease progression. Statistical analysis was performed using Wizard 2.0.16, applying both parametric and non-parametric tests based on the data type. Repeated studies on the same patient were treated as individual data points. Results are presented as average ± standard deviation (p-value).
Results: Ninety-three DMD patients having 324 echocardiographic studies (105,113,106 per subgroup) and 278 controls with 278 (103,87,88) studies were included. Patients were 9.7 ±4.1 and controls 9.4 ±4.2 years old, p=0.030 and BSA 1.13 ±0.36 vs 0.95 ±0.24 (p=0.884) overall, as expected by matching. The detailed results are given in Table 1. IVSd was not found to be different between any subgroups while LVPWd and IVSs were lower in the Late subgroup but LVPWs was lower in all subgroups. LVDd and LVEDV were higher in the Early and Intermediate subgroup while lower in the Late. LVDs and LVESV were higher in all subgroups. Interestingly LVSV was lower only in the Late subgroup. EF and SF were lower in all comparisons.
Conclusions: Several differences from healthy controls are evident even from the earliest studies including higher LVDd, LVDs, LVEDV, LVESV, LVM, LVMi and lower LVPWs, EF, SF suggesting early occult progression of the disease. The prognostic value of these findings remains to be studied.

Keywords: Duchenne muscular dystrophy, cardiomyopathy, heart failure, echocardiography

Table 1.

BSA strata	<0.88		≥0.88 and <1.30		≥1.30	
Group	CTRL	DMD	CTRL	DMD	CTRL	DMD
Numer.	103	105	87	113	88	106
BSA [m2]	0.75 ±0.10 0.590	0.74 ±0.09	1.07 ±0.12 0.492	1.08 ±0.12	1.61 ±0.20 0.051	1.55 ±0.18
IVSd [mm]	5.83 ±1.00 0.810	5.80 ±0.92	6.78 ±1.32 0.315	6.95 ±1.02	8.50 ±1.61 0.221	8.25 ±1.20
LVPWd [mm]	5.40 ±0.73 0.104	5.23 ±0.80	6.36 ±0.96 0.904	6.34 ±0.88	7.74 ±1.53 0.035	7.33 ±1.05 L
IVSs [mm]	8.52 ±1.58 0.322	8.71 ±1.19	10.07 ±1.28 0.251	9.86 ±1.33	12.31 ±2.11 <0.001	11.36 ±1.47 L
LVPWs [mm]	8.73 ±1.59 0.008	8.23 ±1.08 L	10.46 ±1.76 0.001	9.73 ±1.15 L	12.83 ±2.57 <0.001	10.99 ±1.80 L
LVDd [mm]	34.52 ±3.19 0.003	35.97 ±3.64 H	39.47 ±3.30 0.010	40.69 ±3.33 H	46.65 ±3.88 0.005	44.95 ±4.52 L
LVEDV [ml]	49.78 ±10.75 0.001	55.10 ±12.84 H	68.51 ±13.63 0.011	73.64 ±14.35 H	101.55 ±20.41 0.012	93.52 ±23.56 L
LVDs [mm]	21.24 ±3.03 <0.001	23.22 ±3.13 H	24.60 ±2.47 <0.001	26.88 ±2.94 H	28.81 ±3.27 0.002	30.51 ±4.35 H
LVESV [ml]	15.21 ±4.28 <0.001	19.06 ±6.30 H	21.74 ±5.55 <0.001	27.29 ±7.73 H	32.38 ±9.29 0.003	37.69 ±14.82 H
LVSV [ml]	34.56 ±8.53 0.198	36.04 ±7.93	46.77 ±10.39 0.756	46.35 ±8.35	69.17 ±14.09 <0.001	55.83 ±11.75 L
EF Teich. [%]	68.87 ±4.64 <0.001	65.79 ±5.46 L	68.07 ±5.20 <0.001	63.31 ±4.69 L	68.39 ±5.58 <0.001	60.42 ±6.48 L
SF Teich. [%]	38.40 ±7.70 0.001	35.60 ±4.20 L	37.60 ±4.30 <0.001	34.00 ±3.40 L	38.30 ±4.40 <0.001	32.30 ±4.40 L

Results by BSA strata and patient Group. Within each cell: the upper row: Mean ±SD, the lower row p-value and the flag: H – significantly higher mean in the DMD group, L – significantly lower mean in the DMD group, empty – the difference is statistically not significant.

PP-095

Mechanisms of aortic remodelling post-fontan are associated with early-stage haemodynamic efficiency in the reconstructed aorta: A 4D flow MRI study

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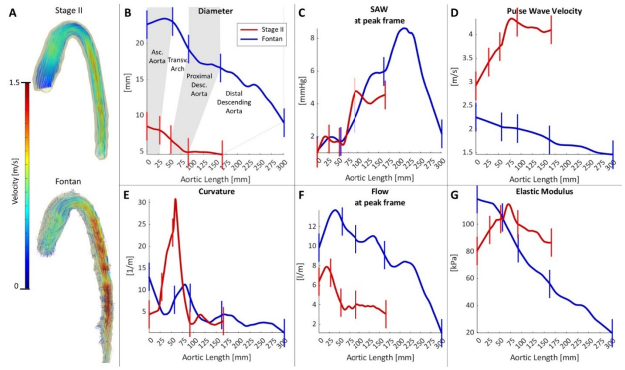
Background and Aim: In single right ventricle pathologies, such as Hypoplastic Left Heart Syndrome (HLHS), 4D Flow MRI allows characterizing conduit and reservoir function in the reconstructed aorta. At an early stage, such assessment can have prognostic value for cardiovascular remodelling after Fontan completion. We present a case study to provide initial evidence that MRI-based analysis of conduit and reservoir function in the reconstructed aorta after stage II can predict future remodelling after Fontan (stage III).
Method: Longitudinal cardiac MR (CMR) data, including 4D Flow MRI, were acquired in one HLHS patient before and after Fontan completion at 3 and 12.4 years-old, respectively (Body Surface Area, BSA, of 0.6m^2 and 1.4m^2). Aortic haemodynamic efficiency (conduit and reservoir functions) was computed from 4D Flow data and anatomical metrics were measured on 3D angiography. All metrics were indexed by BSA and compared to identify remodelling patterns.
Results: After Stage II, this patient exhibited a sharp curvature at the transverse arch and an abrupt change in diameter and pressure drop at the proximal descending aorta. The longitudinal analysis showed that the aorta had remodelled after Fontan to remove the sharp changes in curvature and diameter (decrease from 38m^{-1} to 11m^{-1} in peak curvature and cross-sectional tapering from 33% to 11%). This anatomical adaptation resulted in a less abrupt pressure drop at the proximal descending aorta, which indicated a more efficient conduit function compared to Stage II (from 3.5mmHg to 2.5mmHg). Furthermore, the descending aorta after Fontan was less stiff compared to Stage II (Elastic modulus reduction from 101kPa to 12kPa) and exhibited an increase in arterial strain (from 16.88% to 69.10%). Finally, an increase in peak pressure drop at the distal descending aorta was also observed after Fontan, potentially indicating abnormal haemodynamics in this region.
Conclusions: This longitudinal study shows a case where the reconstructed aorta positively remodelled, removing localized high curvature, reducing the abrupt diameter change through the arch and minimizing the subsequent loss of conduit function. This remodelling was associated with a compensatory increase in compliance along the descending aorta.
These results provide a framework for more comprehensive studies on patient-specific single-ventricle arterial remodelling.

Keywords: HLHS, 4D Flow MRI, haemodynamics, Aorta, Fontan

Geometric and haemodynamic remodelling of Fontan in HLHS case



A: Anatomy and streamlines of the HLHS patient considered at HLHS pre-Fontan stage II and post-Fontan stage III; B-G: Patient-specific traces along the aortic centreline for the HLHS patient at stage 2 and after Fontan (stage 3), where five vertical lines at each trace delimit the four subdivisions of the aorta: ascending (Asc.) aorta, transverse (Trans.) arch, descending (Desc.) aorta proximal and distal. The metrics considered are divided into geometry (B: Radius and E: curvature), conduit function (C: simplified Advective work-energy relative pressure - SAW and F: flow rate), and reservoir function (D: pulse wave velocity - PWV, and G: Elastic modulus).

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PP-096

Early experience with virtual reality modelling for planning of percutaneous, surgical and hybrid procedures in patients with congenital heart defect

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Background and Aim: Advanced imaging techniques play a crucial role in paediatric cardiology, with traditional methods now being complemented by virtual reality (VR) tools. We present our initial experience in converting standard computed tomography (CT) scans into three-dimensional (3D) VR models to assess their suitability for planning of percutaneous, surgical and hybrid procedures.
Method: Between January and August 2023, VR models were created in 60 patients with various congenital heart defects. In all cases CT scans performed previously for clinical indications were utilized. VR modelling was selectively used when advanced imaging promised additional benefits.
Results: All 60 CT scans were successfully converted into VR models, enhancing visualization, spatial depth, anatomical assessment, precise measurements and facilitating procedural simulation.
We most commonly created models for pulmonary artery interventions (19) or prior to percutaneous pulmonary valve implantation (12). Detailed VR analysis was conducted in 6 newborns undergoing ductus arteriosus (DA) stenting in duct-dependent pulmonary circulation. Virtual reality models were prepared in 6 cases of aortic coarctation and in 4 patients with hypoplastic left heart syndrome after Norwood operation, including 2 with partial anomalous pulmonary venous return (PAPVR). Extensive VR evaluation was performed in 3 patients with multiple aorto-pulmonary collaterals prior to unifocalization procedures, 3 patients with heterotaxy syndrome at various stages of palliation, and in 2 patients with severe aortic dissection. Single models were prepared in patients with Bland-White-Garland syndrome, pulmonary embolism, neuroendocrine heart tumor, infective endocarditis, and sinus venosus atrial septal defect (SVASD).
VR imaging improved procedural planning for trans-catheter closure of SVASD, aortic coarctation with pseudoaneurysms and an additional structure dividing the arch, and a newborn with heterotaxy as well as PAPVR. Detailed VR simulations facilitated effective interventions on an extremely narrow pulmonary artery in a premature infant, in cases of pulmonary valve implantation and in DA stenting with a high risk of airway compression.
Conclusions: In our early experience successful VR modelling was possible with all routinely performed CT scans. This advanced imaging modality provided interventional cardiologists and surgeons with additional information facilitating procedural planning in various congenital heart defects. Further studies evaluating objective benefits of this new imaging modality are warranted.

Keywords: virtual reality, three-dimensional modelling, congenital heart defect, interventional cardiology, multimodality imaging,

PP-097

Three-dimensional left ventricle strain assessment in children treated for acute lymphoblastic leukemia

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Background and Aim: Acute lymphoblastic leukemia (ALL) is the most common childhood malignancy. ALL treatment is effective but involves the use of cardiotoxic anthracyclines. Thus, early detection of cardiotoxicity before the onset of heart failure symptoms is imperative. Strain echocardiography has been reported as a novel and sensitive method of cardiac function assessment. However, it is still rarely used in the follow-up of children treated for ALL and no data on three-dimensional (3D) strains' utility in this population had been published. To assess the left ventricular function in 3D strain echocardiography in children treated with anthracyclines for ALL and compare with healthy individuals.
Method: For this prospective study consecutive children who completed at least one ALL treatment protocol involving the use of doxorubicin and/or daunorubicin were enrolled. 3D echocardiographic records were analyzed offline with the use of 4D LV-Analysis software (Philips Medical System, USA) assessing left ventricle ejection fraction (LVEF), end diastolic volume (LVEDV), end diastolic volume indexed (LVEDVI), global longitudinal strain (LV-GLS), global circumferential strain (LV-GCS) and radial strain (LV-GRS). The results were compared with healthy individuals matched by age and sex. Two echocardiographers evaluated data for interobserver variability.
Results: The study group consisted of 24 children (14 males and 10 females) aged 3-17 years (mean 9.8, SD=3.8 years) treated for ALL. The median time from the first anthracycline dose was 2(IQR1=0.63, IQR3=6.75) years with median cumulative dose of 120(IQR1=104, IQR3=180)mg/m2 for body surface area. Only 2 children received cumulative anthracycline dose of above 300mg/m2. Statistically significant differences between study and control group were observed in terms of LVEF (53.8%; SD=5,5 vs 60.45%; SD=6.1, p<0,001), LV-GCS (-22.9; SD=3.45 vs -28.2; SD=4.8, p<0,001) and LV-GRS (-38.6; SD=5,9 vs -45.4; SD=5.9, p<0,001). The differences in terms of LV-GLS, LVEDV and LVEDVI were not statistically significant. The strain values were within literature norms for sex and age.
Conclusions: 3D LV-GCS and LV-GRS may be sensitive indicators of subclinical cardiac function impairment in children treated for ALL with anthracyclines.

Keywords: three-dimensional strain (3D strain), left ventricular function (LV function), anthracyclines, cardiotoxicity, acute lymphoblastic leukemia (ALL)

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PP-098

Total anomalous pulmonary venous connection in combination with an anomalous drainage of the inferior vena cava in a newborn with heterotaxy syndrome

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Background and Aim: Anomalous pulmonary vein connections are frequent components in heterotaxy syndromes. It is well recognized that abnormalities of the systemic venous system can be associated. However, they are frequently underdiagnosed.
Method: A full-term newborn girl was transferred to our clinic with suspected supracardiac total anomalous pulmonary venous connection (TAPVC) without obstruction. An atrial septum defect and a ventricular septum defect were present. On admission, there were no signs of heart failure or respiratory distress. All four pulmonary veins (PV) drained via a collecting vein into a vertical vein to the superior vena cava. Abdominal ultrasound demonstrated a midline liver, a right sided stomach and asplenia. Thus, heterotaxy syndrome, specifically right atrial isomerism, was diagnosed.
She underwent surgical correction of the intracardiac defects and surgical anastomosis of the PV to the left atrium (LA). The presence of anomalous subdiaphragmatic veins was suspected but difficult to visualize during surgery. Postoperatively, oxygen saturations ranged between 88 and 96% without the presence of respiratory symptoms. CT and MRI imaging were performed. These revealed a drainage of the inferior vena cava (IVC) into the LA, which was confirmed on angiography, also a drainage of the right and middle hepatic vein directly into the right atrium (RA), and the left hepatic vein drained via the IVC to the LA. In addition, stenosis of the surgical anastomosis of the PV with the LA developed 5 weeks after surgery. Thus, revision of the surgical anastomosis of the PVs to the LA and anastomosis of the IVC to the RA was performed.
Results: The patient recovered quickly and could be discharged with normal oxygen saturations on the 7th postoperative day.
Conclusions: To the best of our knowledge, only one case of anomalous drainage of the IVC to the LA with TAPVC is described in literature. Patients with right isomerism with TAPVC can present with unusual variations of the systemic vein connections, which are challenging to detect by echocardiography. A low threshold for additional imaging such as CT or MRI is suggested in these cases. Cardiologists and surgeons should have a high awareness for rare systemic venous abnormalities in patients with heterotaxy syndrome.

Keywords: total anomalous pulmonary vein connection, heterotaxy

PP-099

Three-dimensional arch geometry and blood flow in neonates after surgical repair for aortic coarctation

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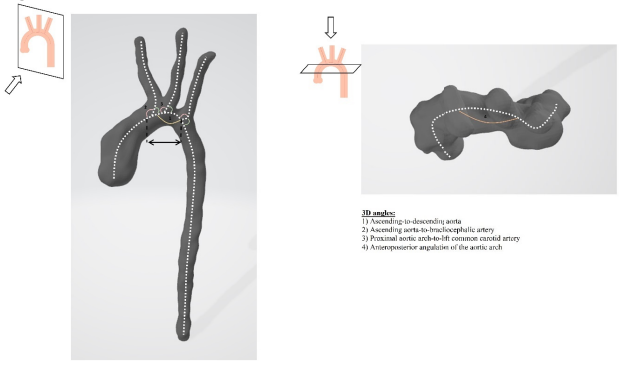
Background and Aim: Re-coarctation of the aorta (re-CoA) is a known, although not fully understood complication after surgical repair, typically occurring in 10–20% of cases within months after discharge. AIMS: to 1) characterize geometry of the aortic arch and blood flow from pre-discharge four-dimensional (4D) magnetic resonance imaging (MRI) in neonates after CoA repair, and 2) compare these measures between patients developing re-CoA within 12 months after repair and patients who did not.
Method: Neonates needing CoA repair, without associated major congenital heart defects, were included. Transthoracic echocardiography (echo) and 4D phase-contrast MRI (1.5T MAGNETOM Aera, Siemens Healthineers, Germany) were performed prior to discharge after CoA repair to assess 3D arch geometry, flow velocity and flow pattern in the distal aortic arch corresponding to the area at risk for re-CoA. Arch geometry was assessed by measuring angles of the aortic arch and its branches (Figure 1) using 3D patient-specific geometries segmented from MRI. Continuous data are presented as median and interquartile range.
Results: The age at CoA surgery was 9 [11] days. Four out of the included 28 patients (14%) developed re-CoA within the first 12 months after surgery. Re-CoA was associated with repair technique (lateral thoracotomy 100%, p=0.02), higher postoperative isthmic flow velocity by echo (1.9 [0.89] m/s vs 1.25 [0.5] m/s, p=0.04) and postoperative crenel aortic arch geometry (100% vs. 21%, p=0.007). A trend to a smaller angle between the ascending aorta and brachiocephalic artery (91 [38]° vs 122 [37]°, p=0.06) and smaller angle between proximal aortic arch and left carotid artery (75° vs 97 [37]°, p=0.04) were seen in re-CoA patients. Patients that developed re-CoA showed more left-handed helical

Figure 1



Aortic arch geometry

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flow in systole (p=0.045), more right-handed helical flow (p=0.02) in diastole and less vortical flow (p=0.05).
Conclusions: Changes in arch geometry and flow pattern after neonatal CoA repair may contribute to the risk for early re-CoA.
Acknowledgment: We thank Ning Jin, Ph.D. at Siemens Medical Solutions USA Inc., Cleveland, Ohio, USA, for providing the 4D phase-contrast research sequence.

Keywords: re-coarctation, neonates, MRI, 4D flow, arch geometry, 3D angles

Cardiovascular Morphology

PP-100
VSD restriction in patients with double-inlet left ventricle during stages of single ventricle

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Background and Aim: Patients with double-inlet left ventricle with ventriculoarterial discordance (DILV-VAD) are predisposed to systemic outflow tract obstruction necessitating systemic outflow relief operations. In this study we evaluate our patients with DILV-VAD with VSD restriction and their outcomes postoperatively.
Method: Between 2010-2023, total of 90 DILV patients were followed, 5 patients were excluded due to missing data. Out of 85, 36 patients (22 males) median age 1.5 months (1day-37years); median 5kg (2.5-57kg); median saturation 88% (55-100%) with DILV-VAD were analyzed retrospectively and 10 patients with VSD restriction were detected. The demographic features of patients, echocardiographic and hemodynamic measurements, invasive/surgical procedures and their outcomes were reviewed.
Results: Median follow up was 68 months (2-146 months). Out of 36 patients 3 patients were lost follow up, 3 patients died. 24 had pulmonary hypertension with pulmonary stenosis/atresia in 9 patients. Arch obstruction was demonstrated in 12, VSD was found diminutive in 11 patients at echocardiography (VSD/Aortic anulus ratio was <1 in 10 patients/among these ≤0.5 in 5 patient where VSD area indexes were <2 cm2/m in all).
Surgery was performed in 33 patients. 21 patients underwent initial pulmonary artery banding (PAB) procedure (9 with arch reconstruction+/-VSD enlargement) and 12 patients underwent arch reconstruction (3 isolated), VSD restriction was developed in 10/36 patients (27%),VSD noted as dimunitive in 9 of them initially. Median age at operation for VSD restriction was 8 months (0.5-111months).
Initial VSD enlargement performed in 4 out of 10 with VSD restriction; (during PAB and arch reconstruction in 4 patients). 1 patient whose VSD nonrestrictive initially, developed outflow obstruction after bilateral banding and Norwood operation performed. DKS operation+Glenn was performed a patient 6 months after PAB+arch reconstruction. VSD enlarged in 2 patients during Glenn/Fontan, in 2 patients after Glenn/Fontan procedures. Pacemaker implanted in 1 patient after VSD enlargement. Bidirectional Glenn shunt performed in 27 patients and 16 patients ended up with Fontan totally.
Conclusions: DILV–VAD patients are candidates for systemic outflow restriction and need to be carefully evaluated for this aspect. Although the primary treatment strategy displays a great variability, VSD enlargement or Norwood type operations in unsuitable patients can be performed initially in DILV–VAD patients with outflow restriction.

Keywords: Double Inlet Left Ventricule, ventriculoarterial discordance, Ventriculer septal defect restriction

PP-101
Congenital corrected transposition of great arteries with situs inversus: Clinical features and long term outcomes

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Background and Aim: ccTGA represents a rare congenital cardiac anomaly marked by discordance in both atrioventricular and ventriculoarterial connections. Although detailed reports exist for adult ccTGA patients, limited literature addresses pediatric cases with situs inversus (IDD-ccTGA). This study aims to assess the characteristics and follow-up results of pediatric patients with IDD-ccTGA.
Method: This retrospective analysis examined 25 pediatric patients with IDD-ccTGA treated at pediatric cardiology clinic from January, 2010, to November, 2023. IDD-ccTGA characteristics included inverted atria, D-loop ventricles, and D-TGA. VSD, pulmonary stenosis or atresia, AV valve regurgitation and ventricular hypoplasia were recorded as associated lesions. The study excluded heterotaxy, double-inlet atrioventricular connection, superior-inferior ventricles, and criss-cross hearts. Documented outcomes encompassed clinical, electrocardiographic, echocardiographic, and angiographic findings, providing a comprehensive exploration of pediatric IDD-ccTGA characteristics and outcomes.
Results: Out of 145 patients diagnosed with congenitally corrected transposition of the great arteries (ccTGA), 25 individuals (17.5%) exhibited IDD-ccTGA. The gender distribution comprised 44% males and 56% females. The median age at diagnosis was 5 months, ranging from 3 days to 25 years, with a median follow-up duration of 92 months (IQR: 65,8-113,5). Patients with isolated ccTGA, having associated lesions of minimal or no clinical significance was 8, and none of these patients had surgery. The median age at the time of the initial operation was 5.5 months (IQR: 0.77-6.5). Among the remaining 17 ccTGA cases, 7 were identified with concurrent pulmonary atresia and had single ventricle repair. Two patients with complex ccTGA without pulmonary atresia had total correction as biventricle. Following biventricular repair, postoperative complete AV block occurred in two patients. A cardiac resynchronization therapy (CRT) was performed in a patient nine years after the initial permanent pacemaker implantation due to biventricular dysfunction resulting from pacing-related dyssynchrony. A patient, who has been followed-up since the neonatal period, was initially diagnosed with congenital complete AV block, and at the age of three months, an epicardial single-chamber pacemaker was implanted.

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Conclusions: This study sheds light on the distinctive features and outcomes of IDD-ccTGA. Surgical interventions varied based on associated lesions, with notable instances of postoperative complications, such as complete AV block, requiring innovative approaches like cardiac resynchronization therapy.

Keywords: cctga, situs inversus, congenital heart disease

Adult Congenital Heart Disease

PP-102
The role of ppvi in adult patients operated for right ventricular outflow tract pathologies with multiple valve insufficiency

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Background and Aim: Right and left ventricular dysfunction may develop in patients with long-term free pulmonary insufficiency (PR) or pulmonary stenosis (PS) who underwent right ventricular outflow tract (RVOT) surgery. Although there is no anatomical problem of the valves in these patients, severe insufficiencies may occur, often secondary to dilatation and cardiac surgery can lead to significant complications and mortality in patients with impaired cardiac functions. Transcatheter interventions for PS/PR reduces volume and pressure load, improves cardiac functions, as well as reduction in other valve insufficiencies and can postpone the need for intervention in these patients.
Method: Patients over 18 years with severe PR or PS with insufficiency in one or more other valves and who underwent primarily transcatheter pulmonary valve intervention (PPVI) were reviewed retrospectively.
Results: 7 patients with free/severe PR or PS as well as multiple valve insufficiency (tricuspit regurgitation (TR), aortic regurgitation (AR), mitral regurgitation (MR), moderate and more) were included. Mean age was 47.9 years (median 47.5 years, range 27- 67.5 years). 5 patients were female, 4 patients had ≥1 RVOT surgery with the diagnosis of TOF, 3 patients had RVOT surgery for other reasons. 6 patients were evaluated as NYHA III/IV. 6 patients had RV dysfunction and right heart failure, and additionally 3 patients had decreased LV functions. Surgery had been planned for 3 patients previously due to multiple valve insufficiency. As the pulmonary pathologies relieved after the PPVI procedure, other valve insufficiencies resolved or subsided in 6 patients, and improvement in ventricular functions and NYHA were observed in 5 patients. Although left or right ventricular dysfunction continued in some degree in some of them, their symptoms subsided and were followed up with medical treatment. No patient underwent surgery after PPVI.
Conclusions: Chronic PR/PS developed after surgery can lead to right heart failure, arrhythmias and even sudden cardiac death as a result of RV dilatation, and may progress rapidly in case of multiple valve insufficiency and also affect LV functions. In appropriate cases, where the patient's condition poses a high risk for surgery, especially when ventricular dysfunction is present, it would be appropriate to perform PPVI first and then re-evaluate the patient in terms of intervention for other valve insufficiencies.

Keywords: PPVI, RVOT surgery, valve insufficiency, RVOT pathologies, adult congenital heart disease

Adult patients operated for right ventricular outflow tract pathologies with multiple valve insufficiency

	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6	Case 7
Age- years	67.5	53.5	47.5	27	56	37	48
Preintervention NYHA	II	III	IV	IV	III	III	III
Pulmonary pathology	Free insufficiency	Free insufficiency	Free insufficiency	Free insufficiency Severe stenosis	Free insufficiency	Free insufficiency	Free insufficiency
Preintervention valve insufficiency	TR(mild) AR(mild) MR(mild)	TR(severe) MR(mod-severe)	AR(severe)	TR(mod) AR(mod)	TR(mild) AR(mod) MR(mild)	TR(mod) AR(mod) MY(mild)	TR(severe) AR(mild-mod) MY(severe)
Preintervention LV dysfunction	no	yes (mild)	yes (mild)	yes	yes (mild)	no	yes(mild)
Preintervention RV dysfunction	yes (mild)	yes	yes	yes	yes	yes (mild)	yes
PP valve- mm	30.5mm	32mm	30.5mm	24.5mm	27.5mm	30.5mm	
Postintervention NYHA	I-II	II	II-III	II	II	II	II-III
Postintervention valve insufficiency	TR(mild) AR(mild) MR(mild)	TR(mod) MR(mod)	AR(mod-severe)	TR(mild) AR(mod)	AR(mild)	TR(mild) AR(mild-mod) MY(mild)	TR(mod-severe) AR(mild) MY(mod-severe)
Postintervention complication	no	Atrial fibrillation	Plevral effusion	no	no	no	no
Postintervention LV dysfunction	no	no	no	subsided	no	no	subsided
Postintervention RV dysfunction	no	subsided	subsided	subsided	subsided	no	subsided

General Cardiology

PP-103
Steps to a spesific insight: Results of a newly establishing fontan clinic from a tertiary center

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Background and Aim: Our aim is to provide data of our specific Fontan clinic which embraces diverse age groups after Fontan surgery and emphasis on routine follow up protocols for early recognition of circulation specific problems on the path to the better quality of life.
Method: All patients who had Fontan surgery and follow-up in Fontan clinic which was initiated as once per week setting since February 2022, were enrolled in the study group.
Results: Out of 69 patients aged between 5 to 49 years, 37 were male. The most common initial diagnoses were "double inlet left ventricle" (n:16). Fifty-nine patients had extracardiac, 6 had intra-extracardiac conduit, 3 had atrio-pulmonary and 1 had lateral tunnel type of Fontan surgery. Ten patients had fenestration initially at the time of operation. Eight patients had a history of prolonged length of hospital stay. The mean oxygen saturation was 94% at last visit. Four patients have active complaints under investigation which can be attributed to Fontan failure (shortness of breath and persistent cough: 3, diarrhea: 1). Pro-BNP levels were elevated in 33 patients. Six of them proved to have decreased systolic ventricular functions in MRI. GGT levels were slightly elevated in the cohort (mean: 44, range: 17-135IU/L). No patient has current albumin level below 3.5 gr/dl. PLE was detected in 3-patients, arrhythmia in 4, thromboembolism in 4, plastic bronchitis in 2. Random stool concentration of alpha-1 antitrypsin was elevated

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in 25 patients. Twenty three patients had abnormal 24-hour tape results with most common changes being 1-2. degree AVblock. Hepatobiliary USG demonstrated increased heterogeneity in 4, hepatosteatosis in 3, hypertrophy of caudate lobe in 3, nodular change in 1. Eighteen patients were evaluated for lymphatic grading system (grade 1 in 10, grade 2 in 5, grade 3 in 2 and grade 4 in 1). Fifteen patients undergone cardiac catheterisation (3-fenestration occlusion, 1-transcatheter AV-valve closure, 1- fenestration opening, 2-LPA stenting, 2-MAPCA closure, 1-ablation for IART. No patient is currently in need for asist device or listed for transplant.
Conclusions: Institutions dealing with this population should be familiar with the unique aspects of Fontan circulation and implementing proper screening, along with effective communication and collaboration among different specialties, could ultimately improve outcomes for this promising young adults.

Keywords: complication, Fontan, single ventricle

Interventional Cardiology

PP-104
A nightmare of an embolised pulmonary valve during a percutaneous pulmonary valve implantation

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Background and Aim: In this report, we present a 15 years-old female patient with post operative tetralogy of Fallot and severe pulmonary regurgitation whose procedure was complicated by pulmonary valve embolization. We describe the successful reimplantation of the semi-opened pulmonary valve into the right ventricular outflow tract.
Method: Under general anesthesia, after intubation, sheaths were percutaneously placed in the right femoral vein, left femoral artery. Contrast material was injected into the RVOT in several positions. Measurements in the right oblique and lateral positions showed a proximal MPA diameter of 27mm, middle section 29mm, distal section 30mm, and a length of 38mm. The 11F sheath in the femoral vein was replaced with an 18F Mullins long sheath and advanced to RPA via the Backup-Meier guidewire. A 43mm Andra XXL stent, loaded onto a Z-med 33x40 mm balloon, was delivered to the MPA. The Mullins sheath was replaced by a 14F Python sheath, and a 32 mm Meril's Myval pulmonary valve and delivery system were advanced to the right ventricular outflow tract. During inflation, the balloon shifted over the valve, moved proximally, and initially opened at the distal end. The partially opened valve and stent were displaced towards the RVOT. Then, the balloon was fully deflated. Through manipulations with the guidewire and the deflated balloon, the partially opened valve and stent were moved back to the RVOT. The distal part of the valve and stent were gently opened by inflating the balloon, and in this state, they were advanced towards the MPA. The valve's own balloon was removed, and it was replaced with a Z-med balloon that can be inflated to a higher atmospheric pressure The Z-med 33x40 mm balloon was passed over the guidewire and through the valve. The balloon was inflated four times from the proximal to the distal end, ensuring the complete expansion of the stent and centering of the valve.
Results: The final inflation confirmed the proper position of the valve.
Conclusions: In transcatheter procedures, repositioning a displaced pulmonary valve can be extremely challenging and dangerous if embolization occurs. Utilizing higher-pressure balloons to reposition and implant the valve may be a viable option.

Keywords: transcathater pulmonary valve implantation, valve embolization

Cardiovascular Morphology

PP-105
Hypoplastic left heart syndrome, severe form, with pulmonary valve stenosis, an extremely rare association

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Background and Aim: The association between hypoplastic left heart syndrome (HLHS) and pulmonary valvular stenosis (PVS) is extremely rare, with a reported incidence of 0.4% in the setting of complex hypoplasia of the aortic tract. This condition is the effect of premature narrowing during embryogenesis of the patent foramen ovale. Clinical manifestations occur most commonly in the first 3 days of life and include respiratory distress with tachypnea, cyanosis or/and shock. While constituting only 1.4-3.8% of congenital heart disease cases, HLHS is responsible for 23% of all cardiac-related deaths within the first week of life. Opinions on optimal therapy range from medical care to surgical options of staged palliation or cardiac transplantation. There is great variability in HLHS cardiac morphology, being the major determinant of successful surgery.
Method: We present the case of an 11-day-old male newborn from a partially investigated pregnancy, gestational age=39 weeks, birth weight= 3550g, with unfavorable postnatal evolution. Shortly after birth, grade IV/VI systolic murmur and central cyanosis are detected. At the regional hospital, an echocardiography (EC) screening is performed by the attending physician and the suspicion of a complex cardiac malformation is raised, which is why he is transferred to our clinic. On admission, he presents a serious general condition, cyanosis, breathes spontaneously with oxygen on the mask, SpO2=78%, normal blood pressure and ventricular allure. EC is performed which diagnoses hypoplastic left ventricle, with moderate systolic dysfunction, hypoplastic mitral valve, with severely limited opening, aortic valve pseudoatresia, severe ascending aorta hypoplasia, dysplastic pulmonary valve, with moderate stenosis, wide patent ductus arteriosus, restrictive atrial septal defect.
Results: After surgical evaluation only palliative care was chosen, requiring intubation, mechanical ventilation and continuous administration of Alprostadil, Epinephrine and Furosemide.

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Surgical correction was considered to be inappropriate in the context of PVS association.
Conclusions: The particularity of the case is represented by the extremely rare association of HLHS with PVS, the only therapeutic option being heart transplantation. Isolated cases of the Norwood palliative surgical procedure have been reported as having been performed in America, Japan and Taiwan. The risks being very high, palliative medical care is preferred.

Keywords: hypoplastic left heart syndrome, pulmonary valvular stenosis, extremely rare association, optimal therapy

PP-106
Prominent eustachian valve – A case series in a level III NICU
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Background and Aim: The Eustachian valve is an embryologic remnant of the inferior vena cava (IVC) that during fetal life directs oxygenated blood from the IVC towards the foramen ovale (PFO) to the systemic circulation. Usually it regresses after birth and has no pathologic significance. When prominent, it may cause veno-arterial shunting in the neonatal period through the PFO and result in hypoxemia and cyanosis. The echocardiographic appearance is variable. Rarely, it can cause symptoms associated with obstruction to the right heart or arrhythmia. We aimed to described the newborns (NB) with hypoxemia that were diagnosed with this entity.
Method: This is a retrospective and descriptive, single-site study (a level III NICU) between 2013 and 2023. Data was collected from clinical files.
Results: The study includes four male NB; three were admitted after a positive Pulse oximetry screening and one with mild respiratory distress and transient hypoxemic spells. Median GA 39.5 weeks, median BW 3880g and median Apgar scores at 1st/ 5th min 9 and 10. All presented with hypoxemia (84-92%). Oxygen support improved oxygenation. Echocardiogram exhibited a prominent EV, with right-to-left (R-L) shunting across the PFO. No other cause for hypoxemia was found. Median duration of supplementary O2 was 3.5 days and NICU stay 6 days. They were discharged without oxygen and no hypoxemia. No R-L shunting was present at 2-month follow-up appointment.
Conclusions: In the neonatal period cyanosis or desaturations caused by a prominent EV may be misdiagnosed as a respiratory disorder because in both there is improvement with oxygen therapy. This remnant should be differentiated from Chiari network or cor triatriatum dexter; it may mimic a cardiac mass or be associated to R-L shunting through the PFO or atrial septal defect. The recognition of a prominent EV is also important in planning for various procedures that involve instrumentation of the right atrium. Pediatricians should be aware of the variable presentations and clinical implications.

Keywords: Prominent eustachian valve, Neonate, hypoxemia, critical congenital heart disease screening

PP-107
Ventricular septal defect combined with anomalous origin of right coronary artery in an asymptomatic infant
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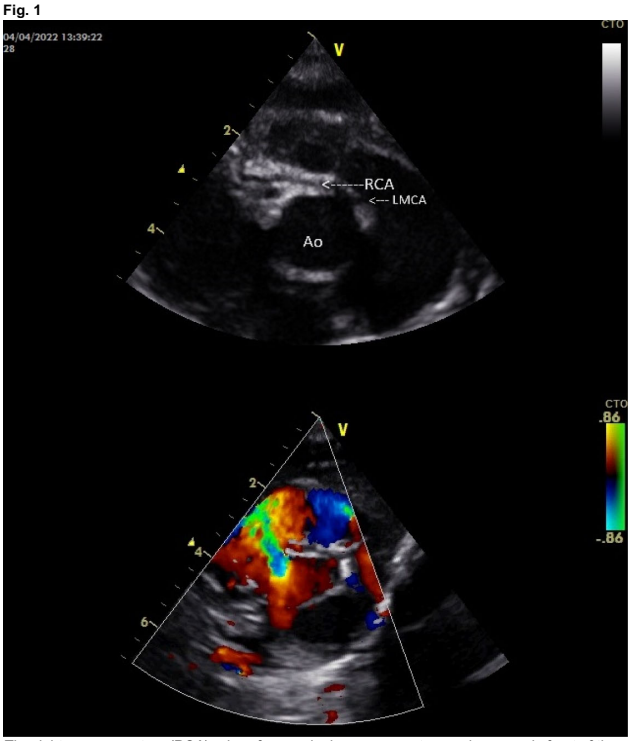
Background and Aim: Anomalous origin of the coronaries are uncommon anatomical abnormalities and usually present without clinical symptoms but as incidental findings. The association of such conditions with ventricular septal defects are rarely diagnosed and reported in infancy. This report details the case of an asymptomatic 2-month-old non-syndromic infant with an anomalous origin of right coronary artery along with a small ventricular septal defect.
Method: Chest radiograph showed cardiomegaly and the transthoracic echocardiogram depicted a small membranous ventricular septal defect of 4mm diameter and anomalous origin of the right coronary artery. Cardiac catheterization with coronary angiography was performed.
Results: The intracardiac pressures were normal. The coronary angiography confirmed the initial diagnosis made by the echocardiogram (Fig. 1) and revealed that the right coronary artery originated from a single coronary artery and had a course in front of the aortic root and behind the main pulmonary artery. After diagnosis, the patient remains asymptomatic and is under cardiology follow-up for potential surgical correction of both malformations. A CT coronary angiography will be scheduled before surgery.

Fig. 1



The right coronary artery (RCA) arises from a single coronary artery and courses in front of the aortic root and behind the main pulmonary artery.

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Conclusions: The occurrence of congenital heart defects concurrent with anomalous origin of any of the coronary branches is sporadic and rarely reported in the literature. Despite its rarity, the recognition of such combined anomalies is pivotal for determining individual patient outcomes, particularly in cases of correctable defects through surgical intervention. To the best of our knowledge, this the first report of an anatomical anomaly involving the right coronary artery arising from a common coronary artery in association with a ventricular septal defect in an infant. Early identification of these anatomical abnormalities poses a challenge for physicians given the potential complication of coronary compression resulting from anomalous origin of coronary arteries.

Keywords: coronary anomalies, right coronary artery (RCA), ventricular septal defect (VSD)

PP-108
Biventricular parachute AV valves in complete AVSD rastelli type B, D-TGA, pulmonary atresia and heterotaxy

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Background and Aim: In complete AVSD, the view from the right atrium shows – apart from the malalignment between the atrial septum secundum and the ventricular septum – the right component of the common AV-valve with the anterior-superior bridging leaflet (ABL) and the posterior-inferior bridging leaflet (PBL).
For best surgical results the decisive anatomic characteristics of the right and left ventricular AV-valve support apparatus in relation to the ventricular and arterial structures need to be demonstrated by the preoperative echo.
Method: The morphologic analysis of 82 heart specimens (DHM 1974-92) demonstrated 11 cases with AVSD (1 partial, and 10 complete). We reanalyzed all and focused on the anatomy of the AV-valves, the papillary muscles and their chordal attachment in relation to the ventricular structures and the great arteries. Furthermore, the systemic and pulmonary venous anatomy and their connections were assessed.
Results: We found the specimen of a 7 weeks-old male heterotaxy patient with AVSD Rastelli type B, D-TGA and valvular pulmonary atresia. The complex anatomical findings in the LV consisted of a parachute AV-valve with a single papillary muscle. All its chords are attached to the threefoliate AV-valve (i.e. free floating ABL, fixed PBL and left ventricular posterior AV leaflet).
The associated valvular pulmonary atresia is secondary due to the extension of the inner heart curvature, muscle of Moulaert, that is obstructing the subpulmonary outflow tract.
The anatomy of the RV is similar: The solitary papillary muscle is the combination of the medial papillary muscle Lancisi and the anterior papillary muscle with all chords attached to the threefoliate right ventricular component of the common AV-valve (ABL, PBL and the right ventricular posterior AV-leaflet).
Conclusions: Each three apposing AV leaflets represent the ventricular entrances, they must be distinguished from the usual cleft. Surgical closure can lead to relevant AV - inflow obstruction.

Keywords: CAVSD Rastelli Type B, Morphology

Congenital Heart Surgery

PP-109
Vascular ring morphology, age at presentation and surgical outcomes – A single centre review

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Background and Aim: Congenital anomalies of the aortic arch may cause formation of vascular rings, which compresses on the upper respiratory and gastrointestinal tracts. Various morphologies of vascular ring have been described, including double aortic arch and right sided aortic arch with aberrant left subclavian artery. In this single centre review, we aim to investigate the antenatal diagnosis, age at presentation and surgical outcomes of the various vascular ring morphologies.
Method: A 10-year retrospective study of patients who had undergone vascular ring surgery at Alder Hey Children's Hospital in Liverpool, United Kingdom. This project was registered with hospital audit committee and data was collected using Electronic Patient Records.
Results: Over a 10-year period (2012-2022), 62 patients underwent vascular ring surgery. Breaking down the types of morphology of vascular ring, 2 were left sided aortic arch with aberrant right subclavian arteries (LAA ARSCA), 33 were double aortic arches (DAA) variants of which 3 were associated with aberrant left subclavian artery (ALSCA), and 27 were right aortic arch (RAA) variants, with 20 being associated with aberrant left subclavian artery (ALSCA) and a further 2 associated with both ALSCA and Kommerell diverticulum. The majority (51/62) patients were symptomatic, with the commonest age at presentation of symptoms being under 12 months old (48/51) with the remaining 3 presenting between 12-24 months old, all of which were RAA variants. Almost half (23/51) of symptomatic patients were diagnosed antenatally, of which 15/23 were RAA variants and 8/23 were DAA variants. Post-operative complications were seen in only 6/62 patients, of which 2 were DAA and 4 were RAA variants. Complications reported were chylothoraces (3/62), re-operation (2/62) and 1 postoperative cardiac arrest requiring ECMO, in a patient later found to have CPVT. No mortalities were reported at time of review.
Conclusions: Double aortic arch variants, followed by right sided aortic arch variants are the most common vascular ring morphologies in our study. Most patients were symptomatic and present in the first year of life, of which almost half were diagnosed antenatally. There is a low rate of complications and no mortality in this retrospective review.

Keywords: Vascular ring, aortic arch anomalies, cardiac morphology, surgical outcomes

PP-111
Mitral valve replacement in small infants: Venturing into the unknown

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Background and Aim: Mitral valve replacement (MVR) in infants and children is still a challenging procedure. Despite improvements in surgical mitral valve (MV) repair techniques, MVR is sometimes unavoidable. In growing children, the fixed diameter of the valve is a disadvantage requiring replacement as the child develops. Also, the anticoagulation necessary after MVR presents a unique challenge in young patients and can cause significant complications.
Method: We present the case of a baby boy, born at 28 weeks of gestation, ELBW. Subsequently an echocardiography was performed demonstrating severe mitral stenosis. Pharmacological treatment was initiated with unsatisfactory evolution requiring mechanical ventilation.
Results: At 2 months of age, the baby was referred to our center, where echocardiography revealed the aforementioned severe MV stenosis alongside severe MV regurgitation and a severely dilated left atrium. After 2 months of life (corrected age 36 weeks, weight=3750g), a MVR with an Edwards'Sapien'3 Ultra (20 mm) was performed in mitral position and an ASD was surgically created. Anticoagulation therapy was started. The degree of residual mitral stenosis has progressed post-operatory, resulting in a high transprothetic gradient and evidence of severe pulmonary hypertension. After 1 week, the patient presented bradycardia and hypotension, followed by resuscitated cardiorespiratory arrest subsequently needing V-A ECMO support. The echocardiography examination revealed an organized clot completely fixing one leaflet and severely restricting the other. Because of this early thrombosis of the valve the reimplantation of a 20 mm Edwards'Sapien'3 valve was needed. The initial evolution after reimplantation was favorable. However, serial echocardiographic assessments over the course of the following two months revealed two severe paravalvular regurgitations due to paravalvular leaks associated with a severe left atrium dilatation and a EEG demonstrating sever neurologic impairment. An interventional paravalvular leaks closure procedure with balloon dilatation is scheduled.
Conclusions: Considering the limited experience in treating this pathology in newborns and small infants, as well as the complicated course demonstrated by our case we advocate for extensive research for new innovative treatment methods in order to improve the morbidity and mortality of these patients.

Keywords: Mitral valve Stenosis, Mitral valve replacement, Edwards Sapien, Paravalvular regurgitation

PP-112
A rare combination of cardiac hydatid cyst and pulmonary hydatidosis in a child

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Background and Aim: Hydatid disease is a common health problem in the sheep-farming countries of the Mediterranean, caused by infection with Echinococcus Granulosis. The common sites are liver and lings. Cardiac hydatidosis is rare.
Method: Herein, we report a case of interventricular septum hydatid cyst with lung involvement in a 4-year-old child.
Results: On one year follow-up, she was asymptomatic without any evidence of recurrence of the hydatid cysts.
Conclusions: Cardiac hydatid cysts should be resected surgically even in asymptomatic patients since delayed diagnosis and treatment can lead to fatal outcomes. In spite of its rarity, the combination of cardiac and pulmonary hydatidosis can be managed with single operation.

Keywords: Hydatid cyst, Hydatidosis, Ventricular septum hydatid cyst, pulmonary hydatidosis

Figure 1

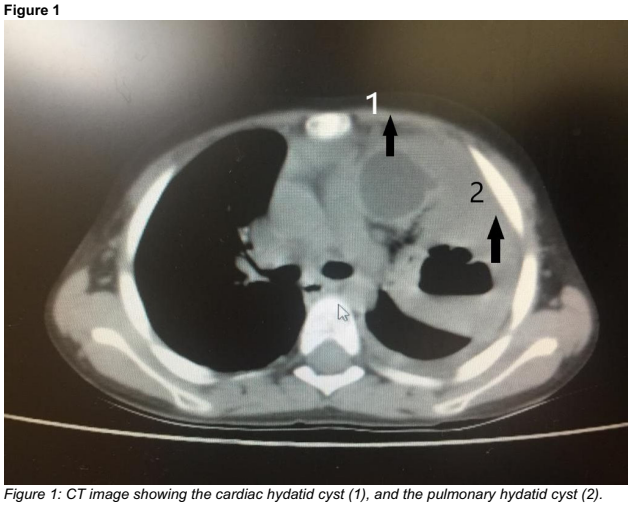

Figure 1: CT image showing the cardiac hydatid cyst (1), and the pulmonary hydatid cyst (2).

PP-113
Delayed sternal closure following complex cardiac surgery in neonates

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Background and Aim: Delayed sternal closure (DSC) is a well-known strategy for management of neonates following complex cardiac procedures requiring prolonged cardiopulmonary bypass (CPB) times and/or deep hypothermic circulatory arrest, which

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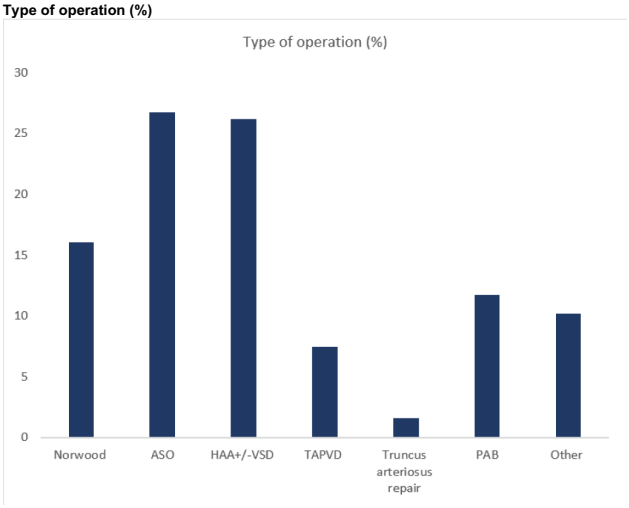
predispose to myocardial oedema, low cardiac output and haemodynamic instability. The purpose of this study is to evaluate clinical outcomes and morbidity associated with DSC in this group of patients.
Method: Retrospective review of neonates who underwent DSC after complex congenital cardiac surgery in a single centre from 2015 to 2021. Out of 357 neonates who had cardiac surgery via median sternotomy, we identified 187 cases who had DSC.
Results: Mean age and weight were 12.8 ± 6.8 days and 3.3 ± 0.5 Kg, respectively. Mean RACHS-1 Score was 4 ± 1. Arterial Switch operation for either simple or complex TGA, hypoplastic aortic arch repair associated or not with VSD and Norwood operation were the most frequent performed procedures. Mean aortic cross clamp time and CPB time were 102.8 ± 52.5 and 183 ± 81 minutes. Mean days of opened chest were 3.8 ± 5.8 days. Mean ICU and hospital stay were 12.8 ± 16.6 and 25.9 ± 36.9 days. Intra-operative ECMO was required in 6 cases (3.2%), while 21 patients (11.2%) needed post-operative ECMO for ECPR. 30-day hospital mortality was 4.8% (N = 9). 8 out of 27 patients (29.6%) who required ECMO died. Only 2 patients (1.07%) needed sternal wound debridement for deep wound infection, while 19 patients (10.2%) had superficial wound infection that was managed conservatively. Univariate analysis showed that DSC days (p 0.011), ECMO (p 0.0001), aortic cross clamp time (p 0.007) and CPB time (p 0.006) associated with 30-day mortality, while in multivariate analysis, only ECMO was significant (p 0.002) for mortality. However, only RACHS-1 score was an independent risk factor for sternal wound infection in both univariate (p 0.019) and multivariate analysis (p 0.05).
Conclusions: Delayed sternal closure (DSC) is a safe and effective therapeutic option following complex cardiac surgery in neonates. DSC might also speed up establishing ECMO during ECPR which nonetheless remains a risk factor for mortality. Moreover, higher RACHS-1 score is associated with sternal wound infection.

Keywords: Delayed sternal closure, Neonates, ECMO, Congenital Cardiac Surgery.

Type of operation (%)



PP-114

Anomalous right coronary artery from pulmonary artery: To do or not to do?

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Background and Aim: Anomalous right coronary artery from the pulmonary artery (ARCAPA) is a rare congenital anomaly, with a reported incidence of 0,002%. Usually asymptomatic in infancy and childhood, it is often incidentally discovered during the assessment of other medical issues. Understanding normal and variant coronary anatomies is crucial for accurate diagnosis and potential treatment planning.
Method: A 2-month-old male infant was referred following a prenatal diagnosis of persistent left superior vena cava (PLSVC). There were no significant cardiovascular findings on physical examination. Initial echocardiography confirmed the presence of PLSVC and showed normal biventricular and valvular function, revealing a prominent left main coronary artery (LMCA) originating normally from the aorta and a suspected anomalous origin of the right coronary artery (RCA) from the main pulmonary artery, with a visible flow at this site. Electrocardiography indicated a normal sinus rhythm with no signs of ischemia. Further diagnostic procedures (coronariography and CT-angiography) confirmed the diagnosis of ARCAPA, revealing an increased LMCA diameter and retrograde filling of the RCA from the left (Figure 1). The patient underwent successful surgical repair at 9 months, with reimplantation of the RCA into the ascending aorta. The postoperative period was uneventful, and the child was discharged under antiplatelet therapy.
Results: At the two-year follow-up, the patient remained asymptomatic, with normal biventricular size and function while maintaining a dilated LMCA.

Figure 1

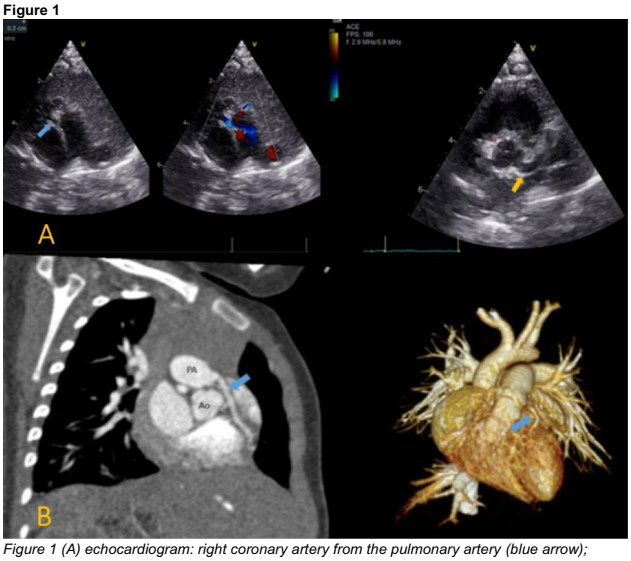

Figure 1 (A) echocardiogram: right coronary artery from the pulmonary artery (blue arrow); increased left main coronary artery diameter (yellow arrow); (B) CT-angiography: right coronary artery from the pulmonary artery (blue arrow); PA: pulmonary artery; Ao: aorta

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Conclusions: ARCAPA is a rare and mainly incidental diagnosis. Despite its usual asymptomatic nature, the association with cardiac symptoms and sudden death, along with significant advances in cardiothoracic and microvascular surgery, supports surgical correction even in asymptomatic cases. This aims to prevent myocardial ischemia and sudden cardiac death by restoring a more natural two-coronary system and abolishing the "coronary steal" phenomenon. Thorough investigation of coronary artery anatomy should be pursued, particularly when associated with other cardiac lesions, as initial echocardiography may be misleading. Early recognition and correction are critical to mitigate risks associated with compromised coronary circulation. Operated infants require close monitoring until risks of coronary reimplantation decrease with growth. Despite positive short and medium-term outcomes, potential late postoperative complications emphasize the necessity of long-term patient follow-up in order to improve management.

Keywords: ARCAPA, coronary anomaly, anomalous origin, right coronary artery, congenital heart disease

PP-115

Aortic coarctation in the neonatal intensive care unit: A ten-year case series

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Background and Aim: Coarctation of the aorta (CoA), often associated with transverse aortic hypoplasia or other malformations, can lead to severe neonatal cardiac failure. Advances in intensive care have enhanced perioperative care. This study aims to describe clinical variables during the perioperative period and assess the results of surgical CoA treatment in neonates.
Method: A retrospective clinical review was conducted on neonates with CoA admitted to the Neonatal Intensive Care Unit (NICU) undergoing surgery from 2013 to 2023. Exclusion criteria included hypoplastic left heart syndrome and interrupted aortic arch. Perioperative characteristics were extracted from our institutional database.
Results: Among the 41 patients (61% male), the median gestational age at birth and age and weight at surgery were 38.7 weeks (IQR 37.4-39.1), 11 days (IQR 7.5-17.3) and 3190g (IQR 2807-3462), respectively. Hypoplastic aortic arch (transverse aortic arch z-score <-2) and bicuspid aortic valve occurred in 43.9% and ventricular septal defect in 26.8%. Prenatal diagnosis existed in 48.8%, without significantly affecting surgery timing compared to postnatal diagnosis (p=0.123). Left lateral thoracotomy was performed in 73.2%, and extended end-to-side anastomosis in 65.9%. Preoperative prostaglandin was used in 73.2%, with 36.6% presenting clinical symptoms of severe CoA. The 30-day mortality was 2.4% (1/41). Extubation occurred at a median of 3.5 days (IQR 2-5) post-operation, and median NICU stay was 12 days (IQR 8.3-20.8). Relevant predictors for NICU length of stay were intubation duration (p=0.009), significantly correlating with chylothorax (p=0.026). Chylothorax occurred in 9.8% and showed no association with surgical approach or technique. The cohort exhibited a 14.6% rate of feeding dysautonomia, correlating with a longer stay (p<0.001). Vocal cord paralysis (41.5%) occurred more in the lateral approach but lacked statistical significance (p=0.303). Cross-clamping time showed no correlation with neurological complications, intubation days or postoperative aminergic support. Aortic arch reinterventions occurred in four patients, with balloon angioplasty performed at a median of 17.5 months (IQR 4.5-52.8) after surgery.
Conclusions: This study provides insights into perioperative characteristics and outcomes of neonatal CoA surgical treatment. While mortality rates are low, attention to early complications impacting hospitalization duration is crucial for overall patient care optimization. Ultimately, neonatal CoA repair demonstrates an overall favorable outcome.

Keywords: Coarctation of the aorta, perioperative care

PP-116

Personalised exernal aortic root support (PEARS) to prevent autograft dilatation for the ross procedure in paediatric population

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Evelina London Children's Hospital

Background and Aim: Ross procedure is the preferred option for aortic valve replacement in children and young adults, offering excellent results at long term. However, one of the main disadvantages of the free-standing Ross is potential autograft dilatation and valve leaflet coaptive failure. Despite several techniques have been developed, none have been the definitive solution in terms of ease of deployment and complete support of the autograft and ascending aorta. We have used the PEARS graft, custom made and personalised, to support the autograft ("Ross PEARS operation") in both adult and paediatric population with adult sized great vessels.
Method: We present a single surgeon experience with the Ross PEARS operation since 2016 (83 cases of which 14 are paediatric) The PEARS graft in this case is made from composite measurements, obtained from the pulmonary phase of a CT angiogram to produce the pulmonary sinuses and the main pulmonary artery shape, coupled with the aortic annular dimensions. We included all patients aged 18 or under. Post-operative follow-up includes annual echocardiograms and cross-sectional imaging.
Results: We have performed 14 Ross-PEARS operations in paediatric patients. Youngest patient was 9 years old (mean age 15.71). 4 patients had previous cardiac surgery, 3 of them had an aortic valve repair (including one Ozaki procedure). Almost 65% had bicuspid aortic valve, with aortic regurgitation being the main lesion (64.29%). One patient required a Konno type annular enlargement. 6 patients had a reduction ascending aortoplasty to match the distal ascending aorta with the pulmonary artery. Mean bypass and cross-clamp time were 197.4 and 151.5 minutes respectively. All patients had at most mild neo-aortic valve regurgitation at discharge, which didn't increase during the follow-up. No autograft dilatation or dysfunction have been observed.
Conclusions: The application of PEARS for supporting the Ross autograft can be a game changer for the Ross operation which is also applicable in the paediatric population. Our cohort has a large number of cases with severe aortic insufficiency and almost half of them had dilated aortopathy. This is proving to be an excellent option providing personalized technology to allow a reproducible technique that avoids autograft failure and subsequent reinterventions.

Keywords: Ross, autograft, PEARS, aorta

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PP-117

Enhancing preoperative evaluation through virtual reality for successful pairing of norwood and glenn procedures in a premature male patient with HLHS

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Background and Aim: Preoperative evaluation is pivotal in determining eligibility and success in surgical interventions for patients with complex congenital heart diseases. Virtual Reality (VR) technology presents a promising tool to enhance surgical decision-making by providing immersive visualization and interactive assessment of cardiac anatomy. This case report outlines the utilization of VR in the preoperative evaluation and successful pairing of the Norwood and Glenn procedures in a premature male patient with hypoplastic left heart syndrome (HLHS).
Method: A male patient, prematurely born with HLHS at 34 weeks, weighing 2830 g, underwent hybrid stage 1 palliation with pulmonary arterial banding and stent implantation in the patent ductus arteriosus at 8 days of age. VR technology, utilizing patient-specific data, was employed for a detailed assessment of anatomical suitability for the pairing of Norwood and Glenn procedures.
Results: Utilizing patient-specific data, a VR-assisted assessment was conducted to evaluate the anatomical suitability for the Norwood and Glenn procedures. The VR simulation provided an in-depth visualization of the patient's cardiac structures, facilitating the determination of procedural feasibility and compatibility. Subsequently, based on this assessment, the surgical team successfully combined the Norwood and Glenn procedures when the child was 4 months and 19 days old.
Conclusions: This case report demonstrates the efficacy of VR technology in the preoperative evaluation and successful pairing of the Norwood and Glenn procedures in a premature male patient with HLHS. VR-assisted visualization of complex cardiac anatomy aided in surgical decision-making and contributed to the optimal procedural outcomes. VR has the potential to enhance the assessment process and improve outcomes in patients with complex congenital heart diseases. Further research and exploration in this field are necessary to validate these findings and expand the application of VR in surgical decision-making for congenital heart disease interventions.

Keywords: virtual reality, hypoplastic left heart syndrome, Norwood and Glenn procedures pairing

PP-118

Interstage hypoplastic left heart syndrome follow-up: Surgical versus hybrid approach. A single-center experience

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Background and Aim: Newborns with diagnosed hypoplastic left heart syndrome (HLHS) require three-stage surgical treatment and often supportive interventional therapy. While each stage has a risk of surgical mortality and morbidity, this becomes a crucial clinical issue between stages 1 and 2. This retrospective study aimed to present a single-center experience of interstage HLHS infants follow-up by comparing two treatment strategies for the first stage: surgical versus hybrid procedure (bilateral pulmonary artery or pulmonary trunk bands and a ductal stent).
Method: Seventeen patients with HLHS hospitalized from January 2022 to June 2023 were included. Twelve of the patients underwent surgical treatment (Group S); among them, four had Norwood operation and eight Sano modifications. The remaining five patients were treated with the hybrid procedure (Group H). Length of ICU hospitalization after the first and second stages, duration of mechanical ventilation after surgeries, weight gain, NTproBNP values, necessity for additional interventions (planned and urgent), and degree of regurgitation of tricuspid valve in echocardiography were reviewed.
Results: The overall survival was 88% (Group S: 91%; Group H:80%; p=0. 749). Patients in group S had nominally shorter hospitalization time in the ICU after the first stage (25,2 days vs 26,6 days, p=0,712) and the second stage (14,16 days vs 30,2 days, p=0,671), shorter mechanical ventilation time after the first stage (9,3 days vs 16,2 days, p=0,711) and the second stage (6 days vs 12 days, p=0,662). Hybrid procedure was linked with nominally weaker interstage weight gain (17g/day vs 18g/day, p=0,268) as well as higher NTproBNP concentration (3080,4pg/ml vs 2820,5 pg/ml, p=0,225). All differences were not statistically significant. 83,3% of patients in group S and 60% in group H needed additional interventions before the second stage. All patients in group H vs 2 in the group S needed urgent catheterization after Glenn operation. 40% of patients in group H and 58,3% in group S had significant tricuspid regurgitation after Glenn operation.
Conclusions: The results may indicate the advantage of surgical treatment over hybrid procedure as the first stage of treatment of HLHS in the analyzed aspects. Further studies on the larger group of patients are required.

Keywords: hypoplastic left heart syndrome, Norwood operation, hybrid procedure

PP-119

Clinical case of percutaneous treatment of massive thrombosis of the right pulmonary artery in a two-month old child

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Background and Aim: Massive venous thrombosis in infants is extremely rare. Such patients require thrombolytic therapy, or if it isn`t effective - surgical or transcatheter thrombectomy (TT). We present case of TT of massive thrombosis of the right pulmonary artery (RPA) in a 2-month old child.
Method: After a car accident, a child required ventriculoperitoneal shunt with central venous catheter placement postoperatively for one month. One month after the intervention, a routine Computed Tomography (CT) in an asymptomatic patient (a level of saturation - 93%, no symptoms of respiratory and heart failure and normal right ventricular function) revealed a massive thrombosis of the right branch of the pulmonary artery. After an ineffective 10-day thrombolytic therapy, TT was performed.

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Results: Recanalization of RPA was performed through the right femoral vein access, utilizing the 4F C4 angiographic catheter with a Runthrough Intermediate 0.014 coronary guidewire. Balloon angioplasty of peripheral branches was performed with coronary balloons from 1,25 to 3.75 mm in diameter. The middle third part of the RPA (thrombosed zone) was dilatated with a mini Tyshak II 6x20 mm. In control angiography, the thrombosed zone of the RPA was expanded to 3 mm; segmental branches of the RPA were well filled. In postprocedural ICU stay for 2.5 days systemic heparinization 25-30 units/kg/h (target aPTT level of 90-100 seconds) was done. On the 2nd postoperative day aspirin 5 mg/kg daily and enoxaparin at a dose of 1 mg/kg twice per day (for 5 days), warfarin - 0.2 mg/kg daily target INR level (2.5-3.5) were added. The child was discharged from the hospital on aspirin 5 mg/kg daily for 6 months. CT (6 months after the intervention) confirmed that the size of both branches were same and the pulmonary arteries were patent well. Antiaggregant therapy was discontinued.
Conclusions: In case of ineffective thrombolytic treatment of massive pulmonary artery thrombosis in infants, the interventional method in combination with anticoagulant therapy can be a good approach to open surgical thrombectomy.

Keywords: massive thrombosis of the pulmonary artery, infants, transcatheter thrombectomy, anticoagulant therapy

PP-121

Determinants of peak VO2 and VE/VCO2 slope in patients with repaired tetralogy of fallot

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Background and Aim: Cardiopulmonary exercise test (CPET) is a well-established method to assess cardiopulmonary function in repaired Tetralogy of Fallot (rTOF) patients. In addition, peak VO2 and VE/VCO2 slope have been linked to adverse cardiac events in this population. However, the determinants of peak VO2 and VE/VCO2 slope are not clear
Method: All rTOF patients who performed a CPET and CMR exam within one-year interval were enrolled. Demographic and surgical data were collected. A multivariate linear regression was performed to identify predictors of peak VO2 and VE/VCO2 slope and variables which remained significant at univariate analysis were included in the final model.
Results: Four-hundred thirty-eight exams tests were completed in 342 patients (56% males; mean age at CPET 22.3 ±7.9 years). The mean age at total repair was 12.9±19.7 months. The mean right end-diastolic volume indexed (RVEDVi) was 120 ± 24 ml/m2, and the mean end-systolic volume indexed (RVESVi) was 56 ± 15 ml/m2. Right ventricle (RV) and/or left ventricle (LV) ejection fraction (EF) mean were respectively 54± 7 and 58 ± 6%. CPET revealed a decreased peak VO2 (26.5±6.4 mL/min/kg) and predicted peak VO2 (73±16%) and fair VE/VCO2 slope (29.8±4.7). At multivariate regression, adjusted for age at CPET, sex and BMI, peak VO2 was predicted by RVEDVi (p=0.04), age at surgery (p=0.01); NYHA class (p=0.04) and sex (p=0.001). Surprisingly, RV and/or LV EF as well as pulmonary regurgitation (PR) did not affect peak VO2. LV EF, age at surgery and gender influenced VE/VCO2 slope (all p=0.01).
Conclusions: In our study, older age at total repair affects both was peak VO2 and VE/VCO2 slope, suggesting that it could be one of the predictors of quality of life in this population. While RV size and NYHA class were the most important determinants on the decreased peak VO2 in rTOF patients, only LV function was significantly linked to VE/VCO2 slope.

Keywords: cardiopulmonary exercise stress test, peak VO2, Tetralogy of Fallot

PP-122

Concurrent repair of pectus exacavatum and open heart surgery: How and when?

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Background and Aim: Pectus excavatum can be associated with connective tissue diseases and congenital cardiac disorders that also require surgical repair. The ideal surgical technique and best timing for pectus excavatum repair in these cases are still controversial. We present our surgical approach for concomitant open heart surgery and pectus repair.
Method: In 2023 two patients have undergone concomitant open heart surgery and repair of pectus. We analysed preoperative and intraoperative characteristics. Surgical technique for the pectus repair was the Nuss procedure. We describe our surgical strategy and postoperative outcomes.
Results: Patient 1 (17 years old), female, had the Loeys-Dietz syndrome and had already undergone one pectus excavatum surgery at age 7. Subsequently, she developed aortic root aneurysm (4.1 cm, Z-score +4.7) and inferior vena cava compression due to chest progressive deformation. Patient 2 (26 years old), male, suffered from aortic root aneurysm (4.7 cm) and severe mitral insufficiency due to a prolapse, and also severe pectus excavatum. An heterozygous mutation in FBN1 gene was diagnosed in October 2022. Open heart procedures were aortic root replacement in patient 1 and mitral valve replacement with a mechanical prosthesis combined with a Bentall-Bono procedure (aortic valve, root and ascending aorta replacement with a graft) in patient 2. In both cases, minimally invasive repair of the pectus excavatum was performed by modified Nuss technique 24 hours later, through a median sternotomy, with no incidences. Intubation time was 28 and 72 hours, respectively. The length of stay in the intensive care unit was 8 and 10 days, and the length of hospital stay 23 and 34 days, respectively. Postoperative pain control was managed with intravenous patient-controlled analgesia with opioids and posteriorly oral treatment with gabapentine and benzodiazepines for 4 months. Postoperative early complications: possible infective endocarditis in patient 2 (2 major Duke standards) resolved by antibiotherapy. Follow-up: both patients in NYHA I, good cosmetic results and no chest deformities.
Conclusions: Concomitant open heart surgery and minimally invasive pectus repair within 24 hours appear to be feasible and safe,

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eliminating the higher risks of staged procedures. This initial experience can set a brand-new horizon in the treatment of these patients.

Keywords: congenital heart surgery, pectus excavatum, concomitant surgery

PP-123

Peripheral lymphatic flow during non-invasive negative intrathoracic pressure with biphasic cuirass ventilation

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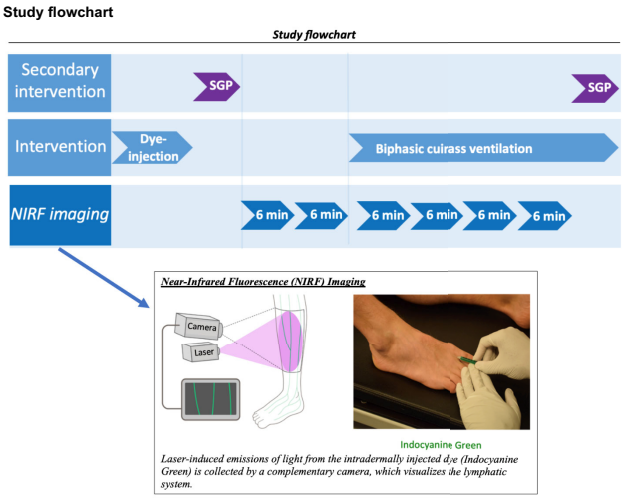
Background and Aim: Single ventricle heart defects have an increased central venous pressure and a congested and vulnerable lymphatic system. The central venous pressure is sensitive to the intrathoracic pressure and decreases during inspiration aiding both venous return to the heart and lymphatic return to the venous system. In contrast to positive pressure ventilation, biphasic cuirass ventilation (BCV) mimics pressure changes of natural breathing and further decreases the intrathoracic negative pressure. To improve the knowledge and understanding of the interplay between lymphatic function and intrathoracic pressure, we plan to evaluate the impact of non-invasive negative biphasic pressure ventilation on the peripheral lymphatic system.
Method: Cardiopulmonary healthy individuals (N=20) between the ages of ≥ 18 and ≤ 40 years and with a BMI ≤ 30 are to be included. The non-invasive negative intrathoracic pressure is achieved by biphasic cuirass ventilation (BCV). A cuirass placed on the chest, actively controls inspiration and expiration by controlling the pressure environment inside the shell. Lymphatic function is examined using Near-Infrared Fluorescence (NIRF) imaging. NIRF-imaging allows for measurements of contraction frequency and contraction velocity. The microvascular fluid filtration is measured through strain gauge plethysmography. A venous congestion protocol is carried out, where the increase in calf volume is recorded by the strain gauge each time the cuff pressure is increased. This enables us to estimate capillary filtration rate. Fluid filtration and lymphatic function is measured during spontaneous respiration and negative pressure biphasic cuirass ventilation (inspiratory pressure: -30 mmHg; expiratory pressure: +2 mmHg; respiration frequency: 10-14).
Results: 5 study subjects are included at the time of abstract submission. Inclusion concludes January 2024.
Primary endpoints:
• Changes in lymphatic vascular contraction frequency and velocity of lymphatic transport during BCV.
Secondary endpoint:
• Changes in SGP-estimated capillary filtration rate during BCV.
Conclusions: Normal lymphatic fluid transport is essential for maintaining a healthy fluid balance and for avoiding edema formation. This study aims to describe the interplay between intrathoracic pressure and lymphatic fluid transport from the periphery. In the future, BCV may serve as a viable alternative to positive pressure ventilation in individuals with a vulnerable lymphatic vasculature prone to lymphatic complications.

Keywords: congenital heart defects, lymphatic dysfunction, biphasic cuirass ventilation, near-infrared fluorescence imaging



Flowchart of the study procedure and Near-Infrared Fluorescence (NIRF) imaging illustration.

PP-124

Perioperative antibiotic prophylaxis in paediatric cardiac surgery – is one day sufficient to prevent surgical site infections?

Christoph Zuern[1], Kai Beckschulte[1], Philipp Mueller[1], Martina Beiderbeck[1], Luisa Paul[2], Sebastian Uhl[2], René Hoehn[1], Johannes Kroll[3], Philippe Grieshaber[4], Tsvetomir Loukanov[4], Brigitte Stiller[1], Matthias Gorenflo[2] and Victoria Ziesenitz[2]
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Background and Aim: Perioperative antibiotic prophylaxis (PAbP) is a standard practice in paediatric cardiac surgery to prevent surgical site infections (SSI). Current prophylactic regimens in Germany include cefuroxime (second-generation cephalosporin). The duration of its administration is debated, due to the lack of randomised controlled trials. The target of antibiotic stewardship is to effectively treat infections, prevent antibiotic resistance and to protect patients from unnecessary harm. This study aims at comparing the efficacy of PAbP with cefuroxime during one versus five days perioperatively in children undergoing congenital heart surgery.
Method: This retrospective, bi-centre study included patients <18 years of age undergoing congenital cardiac surgery with cardiopulmonary bypass (CBP) and primary chest closure from 2015 to 2019. Patients with preoperative antibiotic treatment or signs of infection and cases of prior surgeries with CBP during the same admission were excluded. PAbP consisted of cefuroxime, which was administered according to two different regimens based on local guidelines: one day (centre 1) versus five days (centre 2). Patients were stratified according to age and procedural risk (STAT score). The endpoints were the duration of invasive ventilation and the duration of postoperative hospitalisation.

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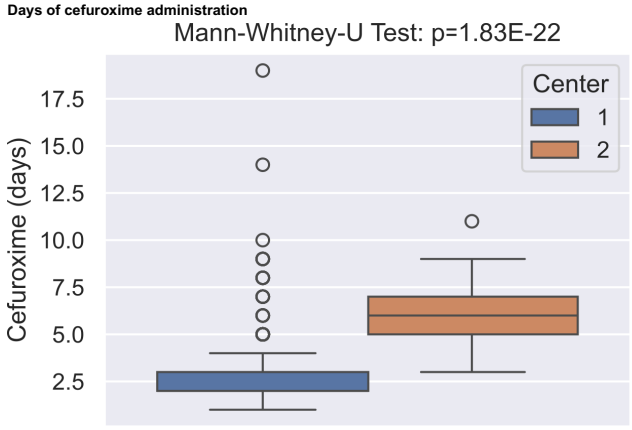
Results: A total number of 305 patients could be included in this analysis so far. Bodyweight (mean 14,0 vs. 16,7 kg, p = 0.074) and STAT score (median 2 vs. 2, p = 0.14) were similar, but the patients were older in centre 2 (median 1,3 vs. 2,5 years, p = 0.04) and mechanical ventilation was longer in center 1 (mean 1,8 vs. 1,7 days, p < 0,01). Mean duration of cefuroxime administration was 3,1 days in centre 1, and 6,3 days in centre 2 (p < 0.01). These results show that the shorter use of cefuroxime is non-inferior and does not cause a significant difference in the duration of postoperative hospitalisation (median 10 vs. 10 days, p = 0.1414). or mechanical ventilation (mean 1,8 vs. 1,7 days, p < 0.01).
Conclusions: According to this retrospective bi-centre study, a one-day PAbP regimen is sufficient to prevent SSI around congenital cardiac surgical procedures. Prospective data should be obtained in a multicentre study to confirm these results.

Keywords: Perioperative antibiotic prophylaxis, Antibiotic stewardship, Postoperative treatment in congenital heart surgery



PP-125
Using a text messaging platform to support patients and families after cardiac surgery

Christopher Buckland[1], Astrid De Souza[2], Erin Bleker[1], Cindy Sha[3], Mohammed Al Aklabi[4], Andrew Campbell[4], James Potts[2] and Kathryn Armstrong[2]
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Background and Aim: Four-to-six weeks post-cardiac surgery is a time when patients and their families have many questions for their healthcare team. In our centre, the surgical nurse practitioner telephones the family once in this time frame to follow-up. Using SMS text messaging between patients and healthcare providers (HCP) can increase communication during this period and better support families following cardiac surgery. Our aim is to implement a SMS text messaging platform (WelTel) to support cardiac surgery patients.
Method: Patients with a scheduled cardiac surgery between January 1 and October 31, 2023 were offered WelTel and consented at either their pre-admission visit or immediately post-surgery. Parents or adolescents (who were capable of managing their own care) were enrolled. Automated messages were sent every Monday asking, "How are you?" Participants could respond with how they were feeling or express any non-emergency health-related concern. All HCP responses occurred within 48 business hours and participants were asked to contact the on-call team for urgent inquiries. Participants received a feedback survey after 4-6 weeks and were discharged from WelTel. Withdrawn participants were not offered a survey. Text messages and care conversations (>2 text messages exchanged) were tabulated.
Results: One-hundred and four participants (101 adults, 3 adolescents) were consented. Eighty-seven participants completed the study and were discharged, 11 are actively participating and 6 withdrew. Forty-nine of 87 (56%) feedback surveys were returned, all parental: 41/49 (84%) parents found it helpful to text their child's HCPs; 37/49 (76%) felt better connected to their child's HCPs; and 32/49 (65%) thought it would be helpful to continue messaging their child's HCPs. The response rate to Monday weekly check-in messages was 63% (383/610) with 309 care conversations between HCPs and participants: 157/309 (51%) were families expressing no concerns, 96/309 (31%) were health-related concerns with 36/96 about surgical wound healing, 17/309 (6%) were about medications and prescription refills and 11/309 (4%) were about appointment scheduling.
Conclusions: A text messaging platform was found to be helpful and increased feelings of support from the healthcare team. WelTel allows families to access supplementary care post-surgery in an easy and timely way.

Keywords: texting, cardiac surgery, follow-up care, mobile communication

PP-127
Optimization of follow-up of children with coarctation of the aorta in the postoperative period

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Background and Aim: To study the features of the clinical status of children with aortic coarctation in the preoperative period; to evaluate the circadian profile of blood pressure in children with aortic coarctation in the postoperative period; determine the level of nitric oxide in children with hypertension, which persists after surgical correction of aortic coarctation; to evaluate the histological features of the aortic structure wall in intraoperative biopsies.
Method: Laboratory, instrumental, statistical, and histological.
Results: The follow-up of 87 patients with aortic coarctation in the preoperative period was analyzed. Daily monitoring of blood pressure was performed in 44 patients. 61 children (the main group consisted of 46 children with high blood pressure: 17 children with stable arterial hypertension, 29 – with labile hypertension and 15 healthy children aged 12 to 17 years) studied the level of nitrite as the final product of nitric oxide, as a marker of endothelial dysfunction. Identified and studied 15 histological features of the structure of the aortic wall in the area of narrowing
Conclusions: Proven practical significance of long-term registration of blood pressure in preclinical diagnosis of hypertension in children after surgical correction of aortic coarctation, detection of

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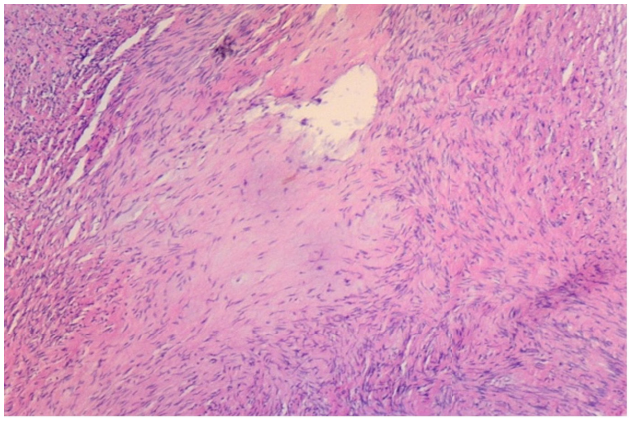
individual features of circadian blood pressure profile, nitric oxide level, which is important for planning preventive and curative rehabilitation measures.

Keywords: children, aortic coarctation, arterial hypertension, nitric oxide, endothelial dysfunction, prognosis.

Locus of aortic coarctation



PP-128

Predicting in-hospital complications in pediatric patients with repaired total anomalous pulmonary venous connection

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Background and Aim: This study sought to evaluate the performance of logistic regression (LR), least absolute shrinkage and selection operator (LASSO), and machine learning (ML) models and to establish the best model for predicting in-hospital complications in children after total anomalous pulmonary venous connection (TAPVC) repair.
Method: A total of 908 pediatric patients who underwent TAPVC surgery were retrospectively included and divided into derivation and validation cohorts at a ratio of 7.5:2.5 based on time. We established eight models using perioperative data to predict in-hospital complications. In-hospital complications were defined as death, low cardiac output syndrome, delayed sternal closure and need for mechanical circulatory support. Through a comprehensive evaluation, the best performing model was used to predict and stratify patients.
Results: The LR model demonstrated excellent discrimination (AUROC of 0.752, AUPRC of 0.519), outperforming the other ML algorithms in the validation cohort. Patients with a higher LR score were found to have a significantly increased risk of in-hospital complications (adjusted odds ratio (OR) 6.609; P <0.001). CPB duration, body surface area (BSA), preoperative intubation, emergence surgery, ventricular septal defect (VSD), vertical vein (VV) processing, anatomic type and index left atrium diameter were important factors for predicting adverse outcomes.
Conclusions: An LR-based risk stratification tool is a reliable prediction model for in-hospital complications in children with repaired TAPVC and may serve as a potential tool to promote personalized treatment for TAPVC.

Keywords: Total anomalous pulmonary venous connection, Predictive model

Analysis flow for the development and evaluation of models

PP-129

Outcome after surgical correction of isolated bland-white-garland syndrome

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Background and Aim: Patients with isolated Bland-White-Garland syndrome (BWG) usually present with severely impaired cardiac function and sometimes significant MV incompetence during the first year of life. Corrective surgery including redirection of coronary artery flow from the aorta is crucial for survival.
Method: After diagnosis patients were urgently scheduled for surgical connection of the left coronary artery to the aorta. The operation was performed predominantly by a modified Takeuchi procedure using a patch of autologous pericardium to tunnel the coronary artery blood flow from a fenestration between the ascending aorta and the pulmonary artery to the aberrant coronary ostium. In cases of proximity of the ostium a direct coronary artery anastomosis to the aorta was undertaken. 24 Patients with an

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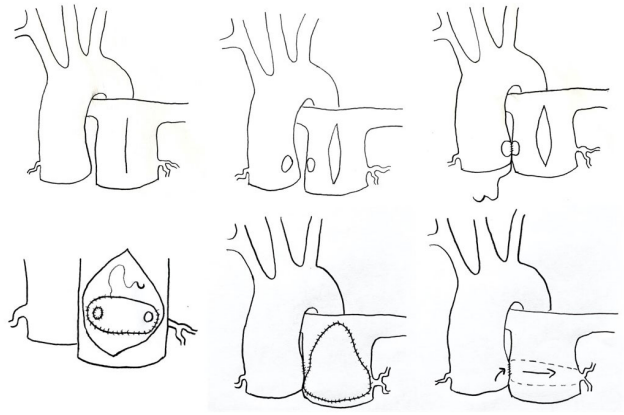
isolated BWG syndrome underwent surgery at the study site between 01.01.1993 and 01.03.2023. Median age at surgery was 138 days [105;296]. The objective of this retrospective single center study was to assess the long-term outcome regarding our surgical technique, reoperations and recovery of LV-function.
Results: 3 patients required intensive care therapy prior to surgery because of congestive cardiac failure.16 pts. received a Takeuchi procedure and 8 a coronary transfer. Mean aortic cross clamp time was 64 min (range 33 – 138 min). Median ICU stay was 8 days [7;17], median hospital stay 20 days [15;38]. All patients were alive after a median follow up of 10,5 years [3,8;15,9]. Preoperatively severely depressed left ventricular function improved dramatically, showing no or mild ventricular dysfunction at the last follow up examination in 22 pts. 6 patients underwent cardiac reoperations during FU: 2 patients had mitral valve surgery (leading to mechanical mitral valve replacement after 2 attempts of reconstruction in 1 pt.); 4 patients had pulmonary artery stenosis after median 8 years. There was no sign of baffle stenosis or leak during FU.
Conclusions: The modified Takeuchi procedure offers excellent operative results with a good perspective on recovery of the depressed left ventricular function during long term follow up. Reoperations at the pulmonary artery were seen and have to be prevented by a generous patch in the pulmonary artery. Associated mitral valve incompetence can make additional procedures necessary.

Keywords: Bland-White-Garland syndrome, ALCAPA, Takeuchi procedure

Modified Takeuchi procedure



Pericardial Baffle from aberrant coronary ostium to fenestration between aorta and pulmonary artery

PP-130
Severe dysplasia and regurgitation of the tricuspid valve combined with a giant right atrium: Prenatal diagnosis and consequent surgical repair

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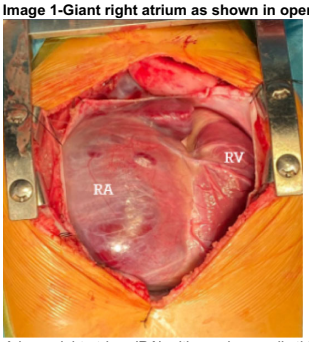
Background and Aim: The most common congenital malformation of the tricuspid valve (TV) is Ebstein's anomaly, while tricuspid dysplasia is a less common cause of tricuspid valve regurgitation (TVR). Differentiation of the two pathologies is carried out through recognition of a normal delamination of the leaflets and a normal septal leaflet's hinge point.
Method: We report a case of a full-term neonate transferred to our hospital because of prenatal diagnosis of severe TVR and a giant right atrium (RA).
Results: The patient was hospitalized in the neonatal ICU and treated with diuretics and ACE inhibitors. Since being asymptomatic, we decided a watchful waiting monitoring. At the age of 2 years old, he underwent elective surgery. Transesophageal echocardiogram (TOE) in theatre confirmed known findings of a dysplastic, non-Ebstein TV with severe eccentric regurgitation and a giant RA, measured 66mm in one diameter. After median sternotomy was performed a huge RA with an abnormally thin (transparent in some areas) wall was found displacing the RV to the left. On cardiopulmonary bypass and the heart still, a RAtriotomy was done. TV was severely dysplastic: the septal leaflet (SL) was extremely thin with its motion limited by a short corda tendinae; the anterior leaflet (AL) was connected to the RA free wall with a "spider net"-like fibrous tissue; and both the anterior and posterior leaflets (PL) had a couple of tiny perforations. TV repair included: cut of the anormal corda tendinae limiting AL's motion, resection of the "spider net"-like tissue and freeing the AL, fine suture of the perforations of the PL and AL and, finally, a De Vega tricuspid annuloplasty. The RA was generously reduced in size by excision of a large strip of the most abnormal RA wall. Post repair TOE showed a mild TVR and a RA diameter 36mm. Postoperative course was uneventfull. Patient was discharged home asymptomatic and free of medication. At follow-up at 4 years the patient is free of medication, has a stable mild TVR and the RA remains within normal limits.
Conclusions: Congenital non-Ebstein TVR with a giant RA is a rare entity and individualized surgical repair contribute to an optimum surgical result.

Keywords: giant right atrium, dysplastic tricuspid valve, congenital, surgical repair

Image 1-Giant right atrium as shown in operation theatre



A huge right atrium (RA) with an abnormally thin and transparent in some areas wall displacing right ventricle (RV) to the left, as shown through median sternotomy.

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PP-131

MID-term outcome with a standardized repair strategy for redo left atrioventricular valve repair after atrioventricular septal defect repair

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Background and Aim: Recurrent left atrioventricular valve (LAVV) regurgitation after valve repair is a challenging technical problem. The aim of this study was to introduce a standardized LAVV repair strategy and to evaluate the results.

Method: The LAVV was exposed by means of stay sutures placed at both trigones and posterior part of the annulus. Direct and systematic inspection of the LAVV apparatus (ie, papillary muscles, chordae, leaflets, and annulus) using nerve hooks allowed us to confirm the echocardiographic findings and completely assess the valve pathologic condition.

Results: A total of 64 patients met the including criteria and were included. With regard to the AVSD morphology, there were partial AVSD in 28, and complete AVSD in 36. Age at the first reoperation was 61.0+15.6 months. Age at the primary AVSD repair was 18.6+29.9 months and the median time to the reoperation was 43.4 (0.41-70) months. The techniques were used in combinations: mal-connected chordae resection in 31 patients, papillary muscle splitting in 10, cleft closure in 54, leaflet plication in 9, leaflet augmentation in 18, and annuloplasty in 56. Forty-six patients were grouped in non-augmentation group while 18 patients were categorized in leaflet augmentation group. The Median follow-up after the reoperation was 36.5 months (2-78.3 months). Two patients died during the follow-up period. Valve replacement was required in 4 patients. Freedom from second reoperation at 1 year, 3 years and 5 years was 100%, 88.5%, 88.5%, respectively. During follow-ups, severe LAVVR was documented in 6 patients. Estimated freedom from severe LAVVR at 1 year, 3 years and 5 years was 96.4%, 87.7%, and 83.7%, respectively. Freedom from reoperation and estimated freedom from severe LAVVR were similar between augmentation and non-augmentation group at 1 year, 3 years and 5 years.

Conclusions: Surgical results for the redo LAVV repair had good outcomes using the Standardized-Oriented strategy. Leaflet augmentation is a reliable and reproducible technique in the selected cases.

Keywords: Congenital heart disease, Atrioventricular septal defect, AVSD repair

PP-135

To correct or not to correct? A meta-analysis on mitral surgery in patients undergoing surgery for anomalous left coronary artery

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Background and Aim: The present study is the first meta-analysis comparing long-term outcomes in patients undergoing correction for anomalous left coronary artery (ALCAPA), in regards to concomitant mitral valve surgery.

Method: A systematic review of the literature was conducted to identify all relevant studies with comparative data on mitral valve surgery performed during surgery for ALCAPA correction. Predefined primary endpoints included mortality and mitral valve reoperation.

Results: Fifty-three relevant retrospective studies were identified from the existing literature. The study population included 3,851 patients. Mean age at time of surgery was 6 months [0-99months]. Mean follow-up after surgery was 22.7years [6.3-37.2yrs]. Echocardiographic data were available in 41 studies, including 2,118 patients (55% of total sample). Among patients with available echocardiographic data at time of ALCAPA correction, 283 had no mitral regurgitation (13%), 642 had mild mitral regurgitation (30%), 756 had moderate mitral regurgitation (36%), and 437 had severe mitral regurgitation (21%). A subgroup of patients who underwent mitral valve surgery (yMVS; n=753; 19.5% of total population) was identified. Criteria for concomitant mitral surgery included: age at diagnosis (4 studies; including 146 patients), severity of mitral valve regurgitation (23 studies; including 471 patients), or evidence of structural mitral degeneration regardless of MR grade (16 studies; including 127 patients). A total of 228 patients died during follow-up (5.9%). Among the 1,536 patients with available follow-up echocardiographic data, 519 developed no mitral regurgitation (34%), 663 developed mild mitral regurgitation (43%), 268 developed moderate mitral regurgitation (17%), and 86 developed severe mitral regurgitation (6%). Overall, during follow-up, 91 patients underwent mitral valve surgery (first MVS for 20pts and re-intervention for 71pts, representing a 10% re-intervention rate). In regression analysis, we found no association between mean age at first surgery and need for reoperation (p=0.458).

Conclusions: Mitral valve surgery is performed in about 20% of patients undergoing ALCAPA correction. Indication for MVS vary among centers, although severity of MV regurgitation was the most common criteria (more than 50% of the yMVS group). Of yMVS patients, approximately 10% underwent a second MVS during follow-up, regardless of age at first surgery. Overall, long-term mortality in these patients is over 5%.

Keywords: anomalous origin coronary artery, mitral valve, mitral valve repair

PP-136

A rare case of severe aortic valve regurgitation associated with HLA- B27 arthritis in a 8 year old boy

Giridhar Reddy Soda

Dr Giridhar Soda, Alderhey Childrens Hospital, Liverpool

Background and Aim: We present a 8 year old boy referred with a murmur. He was under Rheumatology team with significant arthritis and Enthesitis. He was managed with Methotrexate and intra articular steroids suggesting severe joint involvement. He was HLA B27 antigen positive and has been under Rheumatology review for few years.

Method: He was reviewed in the cardiology clinic and was noted to have a soft systolic murmur and a rumbling diastolic murmur. He was progressively getting breathless and struggling to keep up with his peers over the past few months. He had classic signs of severe

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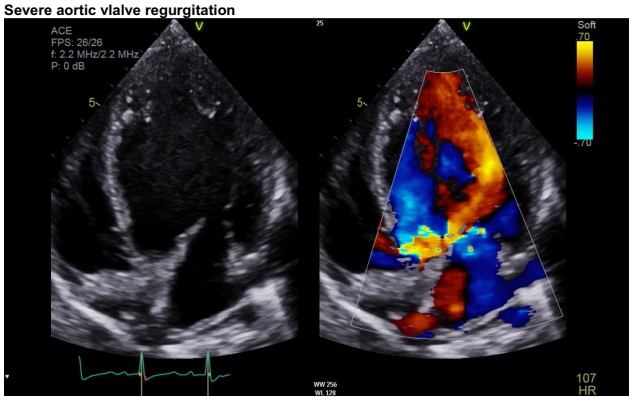
aortic valve regurgitation with collapsing pulse and wide pulse pressure of 110/50 m of Hg.He had Hepatomegaly.
Results: His Ecg was in sinus with left axis deviation.His Echocardiogram revealed severe aortic valve regurgitation with flow reversal noted in the abdominal aorta.He had severely dilated left ventricle with least ventricular diastolic dimension of 6.2 cm.He had a pressures half time of 200 milliseconds.The aortic valve was Trileaflet, with central cooptation defect resulting in severe aortic valve regurgitation.The left ventricle was severely volume loaded and was associated with mild Mitral valve regurgitation.He had a cardiac MRI which revealed severe aortic valve regurgitation with a regurgitant fraction of 48%.The indexed left ventricular end diastolic volume was noted to be 210 millilitres.He has a prosthetic aortic valve replacement and is on long term anticoagulation with Warfarin.His ventricular function has improved and the left ventricular end diastolic dimension has improved over the next few years after surgery.
Conclusions: There is well known association of HLAB27 antigen positive ankylosing spondylitis with Aortic valve regurgitation and conduction defects in adult population.There are very few reported case studies of severe Aortic valve regurgitation requiring valve replacement in paediatric age group.It is important to screen children with HLAB27 antigen for evidence of Aortic valve involvement to prevent progressive deterioration.The severity of joint involvement might also be a surrogate marker for Aortic valve regurgitation.

Keywords: Aortic regurgitation, HLA B27

Severe aortic vlalve regurgitation



PP-137
Risk factors for recoarctation of the aorta in a cohort of 195 paediatric patients after surgery for aortic decoarctation

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Background and Aim: Aortic coarctation is the fifth most common congenital heart disease, accounting for 5 to 8% of all congenital heart defects. Over the years, the survival of paediatric patients who have undergone corrective surgery has improved considerably. However, recoarctation after surgery is still a major problem. Considering this aspects, we have rewieved our experience at IRCCS Policlinico San Donato over the last 19 years.
Method: Demographic and clinical data were collected on all patients who underwent cardiac surgery for aortic decoarctation at the IRCCS Policlinico San Donato, from August 2004 to March 2023. A total of 195 patients aged 0-14 years were recorded and subsequently analyzed.
Results: The incidence of recoarctation in our population was 20% (39/195). At the time of surgery, in the total study population the median age was 30 days and the median weight was 3.8 kg; in the group of patients who presented recoarctation, the median age was 10 days and the median weight was 3.3 kg. Among the parameters considered, younger age ($p = 0.00016$) and lower weight ($p = 0.00033$) at the time of surgery were found to be risk factors for recoarctation. The other considered factors (sex, time of diagnosis, presence of associated defects, choice of surgical technique, type of surgical approach, development of arterial hypertension after surgery, presence of arterial pulses in the lower extremities and signs of ventricular hypertrophy on electrocardiogram) showed no significant differences between the total study population and the group of patients who presented recoarctation.
Conclusions: Our data are in line with the current litterature showing that age and weight at the time of surgery appear to be key factors in determining the likelihood of recoarctation in patients undergoing cardiac decoarctation surgery in childhood. This study underline that a close follow up will be necessary in all patients but a closer follow up should be envised for ones at highest risk, like younger and smaller patients. In the future, it is advisable to identify other possible risk factors by analysing larger cohorts of patients with long-term follow-up.

Keywords: Coarctation of the aorta,Recoarctation

PP-138
Early and mid-term outcomes of the modified senning procedure in the double switch operation: application of the triangular double-door technique

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Background and Aim: We have developed a modified Senning procedure using the triangular double-door technique to preserve the right atrium function as much as possible and report our early and mid-term outcomes of the modified procedure in the double switch operation (DSO) for the patients with congenitally corrected transposition of the great arteries (ccTGA).
Method: A retrospective chart review was performed for patients who had undergone the DSO using our modified Senning procedure in the anatomical repair of ccTGA since July 2018.
Results: Five consecutive patients underwent the DSO for an anatomical repair of ccTGA using our modified Senning procedure. Median age and weight at the operation were 37 months (range, 28-40 months) and 13.1 kg (range, 12.2-14.6 kg). All five patients had pulmonary artery (PA) banding at mean age 2.6±1.3 months (range, 1-5 months), and period between PA banding and DSO was 32.3±5.4 months (range, 26-39 months). There was no operative mortality. All patients show no significant systemic and pulmonary venous pathway obstruction and normal sinus rhythm during median 8.1 months (range, 1-59 months) follow-up period. All patients also show good ventricular function at last follow-up echocardiography.

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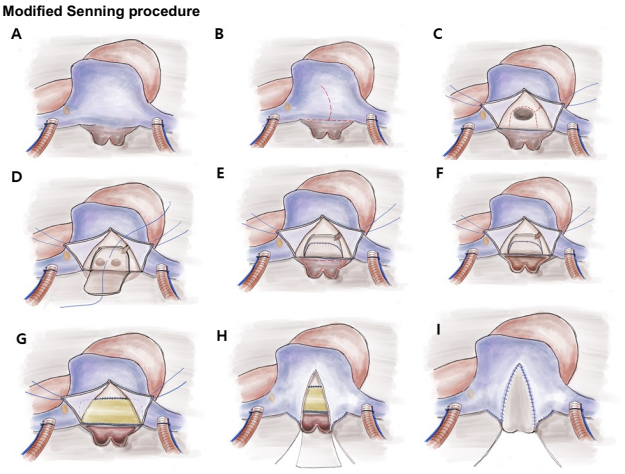
Conclusions: Our modified Senning procedure using triangular double-door technique could be part of the safe and effective surgical option for the DSO of ccTGA in low-volume centers. However, long-term follow-up in more patients is mandatory.

Keywords: double switch operation, Senning procedure, congenitally corrected transposition of the great arteries



A schematic drawing of the modified Senning procedure using triangular double-door technique

PP-140

Efficacy and speed of effect after the first dose of aspirin in children with congenital heart disease

Irene E. Regan[1], Sean T. Kelleher[2], Dermot Cox[3] and Colin J. Mcmahon[4]

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Background and Aim: [2]Many studies have reported that patients must be on stable aspirin or taking it for a minimum of 5 days to achieve adequate response. This is not always practical, especially in critical settings. Prospective identification of patients that are unresponsive to anti-platelet agents such as aspirin could potentially prevent thrombotic events by identifying these patients sooner.

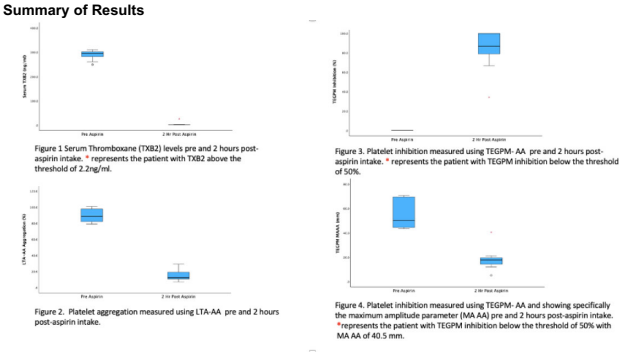
The aim of this study was to investigate prospectively, for the first time, if the first dose of aspirin is effective in decreasing various measures of platelet aggregation, and thromboxane formation and if this can be measured after 2 hours in paediatric cardiology patients and be used as an indicator of responsiveness. A secondary aim was to identify a cut-off for a Thromboelastography with Platelet mapping (TEGPM) parameter, the maximum amplitude with arachidonic acid, (MA AA) as a marker for aspirin response dramatically reducing the blood volume required. Thirdly, we aimed to prospectively identify responsive patients by spiking their blood ex vivo with aspirin.

Method: Recruitment included patients < 18y with congenital heart disease scheduled for interventional cardiac catheterisation and due to receive their first dose of aspirin post-procedure; stent implantation, device closure of atrial or ventricular septal defects and requiring aspirin therapy for 3–6months. Patients were prescribed aspirin at an empirical concentration of 1-5mg/kg/day up to a maximum of 75mg/day. Baseline samples were taken pre-procedure and 2 hours after aspirin initiation. A sub-group had samples taken after their cardiac catheterisation procedure, before aspirin administration to determine surgical effects. Healthy age-matched children attending innocent heart murmur clinics, confirmed with normal cardiovascular status served as a control group.

Results: The majority of patients were responsive (92.3%), and this can be measured at 2 hours post aspirin dosing. Non-response or inadequate response (7.7%) can also be identified at 2 hours after taking the first dose of aspirin. Additionally, we have shown a novel way to reduce blood sample volume requirements by measuring the MA AA as a marker of response, particularly for monitoring.

Conclusions: These findings of rapid efficacy in the majority of patients offer assurance in a sound, practical way to attending clinicians, the patient and their families.

Keywords: congenital heart disease, thrombosis, first dose aspirin



Summary of Results

PP-141

The effects of aspirin dose in children with congenital and acquired heart disease. Results from the paed-m study

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Background and Aim: The optimal dose of aspirin required in children with congenital and acquired heart disease is not known. The primary aim of this prospective observational study was evaluate the effects of aspirin dose on platelet inhibition. The secondary aim was to determine the prevalence and clinical predictors of

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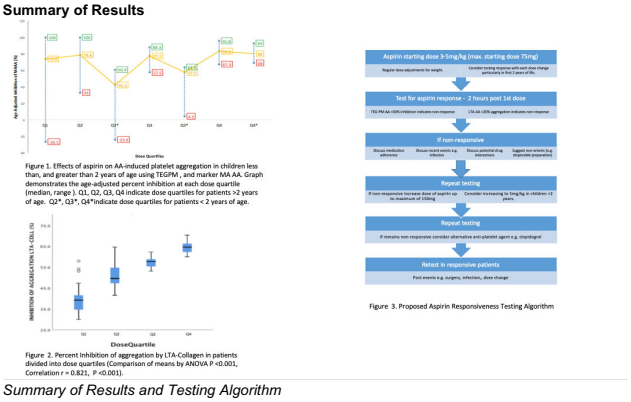
aspirin non-responsiveness. Measurements were by TEGPM only in children less than 2 years (y) of age and using both TEGPM, and light transmission aggregometry (LTA) in children greater than 2y.
Method: We prospectively studied 101 patients with congenital and acquired cardiac disease who were receiving empirical doses of aspirin for a minimum of 4 weeks but no other antiplatelet agents. Patients were stratified according to dose concentration and age. Due to ethical considerations in stopping treatment to get baseline results and because of assumed age-related differences in platelet parameters a healthy age-matched cohort was used to calculate the approximate inhibitory effect of aspirin. This cohort was also used to determine typically normal results using the different platelet assays of patients not taking aspirin. Healthy age-matched children attending the innocent heart murmur clinics, confirmed with normal cardiovascular status served as a control group. This control group stratified into six age categories with twenty patients in each (n=120) was not taking any drugs known to affect platelet function.
Results: There was a trend toward lower age in patients with no response or semi-response to aspirin. All patients were considered responsive to aspirin in the higher dose quartile (Q4) with a median dose of 4.72 (4.18 – 6.05) mg/kg/day suggesting that patients in this age group may require 5mg/kg/day as an empirical dose. In children >2y there was no significant difference in inhibition found in patients dosed at higher doses in Q3 versus Q4 suggesting that patients in this cohort are responsive with 3mg/kg/day dose.
Conclusions: The current practices may lead to reduced platelet inhibition in some children due to under-dosing, or over-dosing in others.
In conclusion younger children require higher doses of aspirin. Laboratory assessment is warranted in this population to mitigate against under and over dosing.

Keywords: aspirin dose, congenital heart disease, thrombosis, resistance



Summary of Results and Testing Algorithm

PP-142

Pediatric health related quality of life of pre- and post-operative patients with congenital heart disease: A single center study in Tanzania

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Background and Aim: Congenital heart disease (CHD) impacts health related quality of life (HRQOL), but data originating from low- and lower-middle income countries are scarce. This study addresses the knowledge gap in HRQOL in children with CHD in Tanzania and compared pre-operative patients with post-operative patients. The objectives were to obtain the general HRQOL reported by patients (5-18) and parents (2-18), clinical (diagnosis, age at diagnosis, cardiac medication, etc.), and socio-demographic variables (region, socioeconomic status), collected from health records and interviews.
Method: This cross-sectional study included patients with CHD at the out-patient department of a national specialized hospital and referral center in Tanzania. Patients were either pre- (n=104) or post-operative (n=100). Patients (n=104) and parents (n=204) answered the HRQOL questionnaires (PedsQL 4.0 SF-15) during interviews. Clinical and socio-demographic variables were collected either during the interviews or through medical health records. Simple descriptive statistics compared the clinical and socio-demographic variables, while t-tests were used to compare HRQOL between groups. Generalized linear models identified significant predictors of HRQOL.
Results: Mean age was 6.3±3.7 years (female n=110, 53.9%), with post-operative patients having a higher age (7.6±4.1 vs. 5.1±2.7), socioeconomic status (0.71±0.14 vs. 0.66±0.20), and more frequent early diagnosis (<1 year; 67.8% vs 47.1%) compared to pre-operative patients. The complexity of cardiac diagnosis was similar between groups. Post-operative patients had significantly higher parent reported HRQOL scores in all domains, with the biggest difference in physical score (effect size d=-0.813). Significant predictors for better HRQOL were post-operative status and higher socioeconomic status. HRQOL significantly decreased with higher Ross classification.
Conclusions: HRQOL is significantly higher in post-operative patients compared to pre-operative patients. Being operated, SES and the Ross classification is significantly influencing HRQOL. Thus, operated children with CHD seem to have a better HRQOL.

Keywords: Congenital Heart Disease, Health Related Quality of Life

PP-143

Cardiac transplantation and 22Q11.2 deletion

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Background and Aim: Congenital heart disease (CHD), neurodevelopmental delay, particular phenotype and immunologic abnormalities. Most patients show only mild immunological disorders. Heart transplantation is rarely needed but should be addressed considering additional abnormalities.
Method: We describe the evolution of a 4 year old girl diagnosed with pulmonary atresia, ventricular septal defect and major aorto-pulmonary collateral arteries (MAPCAs) who underwent CHD surgery and later developed severe biventricular failure that led to cardiac transplantation.

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Results: Our patient was prenatally diagnosed with pulmonary atresia, ventricular septal defect and MAPCAs. Genetic study disclosed 22q11 deletion. She developed heart failure in the first month of life and a surgical procedure was undertaken for unifocalization of MAPCAs and placement of a 6F goretex conduit from the right ventricle to pulmonary arteries. Follow up cath at a year of age revealed conduit stenosis and right pulmonary artery stenosis. She needed several dilatation procedures, a 2nd surgical procedure with fenestrated ventricular septal closure and replacement for a12F RV-PA conduit. A third procedure included closure of septal fenestration and replacement of RV-PA conduit with a 16F Hancock. The patient remained stable with moderate pulmonary insufficiency. She did not show significant infectious complications. Nine months after last surgery the patient showed signs of severe heart failure due to biventricular dysfunction that needed intensive heart failure treatment and heart transplantation was discussed. Pretransplant study: blood group B, good vaccine history. Measles and chicken pox titres were low and she received boosters. She had low Anti A isohemaglutinine titres, despite multiple surgeries and blood transfusions. Inmunologists disclosed mild T cell function tests abnormalities. She was included in the ABO incompatible heart transplant list. A blood A group donor was offered and accepted shortly afterwards. Follow up after transplantation (7 months) has been good, without rejection or significant infectious episodes. Inmunosupressive therapy included induction with basiliximab and standard therapy with tacrolimus, mycophenolate mofetil and steroids.
Conclusions: Heart transplantation for patients with DiGeorge syndrome and mild immunologic abnormalities can be undertaken with standard inmunosupressive therapy. Isohemaglutinine levels should be studied even after infancy in order to consider ABO incompatible heart transplantation.

Keywords: transplant, 22q11.2, DiGeorge, inmunology

PP-144
Secondary subaortic stenosis following ventricular septal defect closure in children: a 13-year experience

Jie Dong, Shun Liu, Shuo Dong, Chuhao Du, Yangxue Sun, Haitao Xu, Jiashu Sun, Qiang Wang, Shoujun Li, Keming Yang and Jun Yan
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Background and Aim: Secondary subaortic stenosis (SSS) can occur after ventricular septal defect (VSD) closure resulting in a secondary surgical intervention. Few case reports were published concerning this phenomenon. Our study aims to summarize the patient and surgical characteristics and highlight the risk factors for SSS after VSD surgeries.
Method: We retrospectively collected young patients (0 to 18 yrs), who underwent a primary open-heart VSD surgical repair followed with a surgical intervention for secondary subaortic stenosis from January 2008 to May 2020 at Fuwai Hospital. Patients who diagnosed with other complex malformation at the first surgery were excluded. Patient information, surgical data and echocardiography variables from the primary VSD surgery to the most recent follow-up (January 1st, 2023) were reviewed by two separate surgeons and received consensus.
Results: In total, forty-one patients were included. For patient and surgical characteristics, a high rate of medium or large VSD was noted in this patient group (38 pts, 98%). The mean size of VSD in this cohort is 10.58mm (IQR:8,13 mm). Patients with a large VSD diagnosed LVOT earlier than those with medium sizes (HR 2.1, $p<0.05$). Most patients (38pts, 98%) in this cohort used patches for VSD closure. Seven patients (17.1%) had subaortic membrane growing along with the VSD patch. Twenty-seven patients (65.8%) also diagnosed with aortic valve regurgitation (AR) when diagnosed with SSS, and 11 patients (26.8%) still have aortic valve regurgitation up to the latest follow-up. Two patients (4.8%) had pacemaker implantation after the SSS. LVOT gradient significantly increased from SSS diagnosis (Median: 53, IQR: 27-100.5mmHg), to SSS surgery (Median: 71.2, IQR: 31-130 mmHg). Patients tend to wait long until underwent secondary surgery to relief left ventricular outflow tract obstruction (Median: 1 year, IQR: 31 days, 3.54 years). Up to the latest follow-up (median: 5.6 years), the mean LVOT gradient rises to 24.7mmHg, which indicates there is still a high risk for LVOTO recurrence in this group. Three patients (7%) had a recurrent LVOTO after successful SSS surgical repair and require a third operation.
Conclusions: A medium or large VSD with patch closure tend to increase the risk of SSS, early surgery of SSS is recommended soon after the diagnosis of pos-top LVOTO.

Keywords: ventricular septal defect, congenital heart disease, subaortic stenosis

PP-145
Risk factors for recurrent left ventricular outflow tract obstruction after surgical repair for subaortic stenosis

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Background and Aim: Subaortic stenosis (SAS) is a progressive secondary cardiac malformation. Surgical procedure is the common treatment for SAS however the recurrence rate of postoperative left ventricular outflow tract obstruction (LVOTO) is high (0-55%). Our study aims to identify the risk factors for recurrent LVOTO following SAS surgery.
Method: We retrospectively included patients (0-18 years-old) who underwent open heart SAS surgery at Fuwai Hospital from Jan 2009-Jan 2020. Children with hypertrophic obstructive cardiomyopathy or unsuccessful primary surgeries for SAS were excluded. Detailed surgical, echocardiography data from the first SAS surgery to the most recent follow-up (January 1st, 2023) were reviewed by two separate surgeons and have received consensus. Recurrent SAS was defined as left ventricular outflow tract gradient higher or equal to 40 mmHg at least 1 month after SAS surgery. Freedom from recurrence was evaluated using K-M method. Cox regression was used for uni/multi-variable analysis. The statistical significance was set at $p<0.05$.
Results: A total of 256 children were included in our study. The medium age of children at the time of SAS surgery was 4.6 years (3 months-17.8 years). After a median follow-up of 6.1 years (IQR, 4.19, 8.47 years), a total of 37 patients developed recurrent LVOTO, with a recurrence rate of 14.5%, however only 18 patients (7.1%) underwent a second surgery. The main reason for stalling the secondary surgery include anxiety for reoperation (21.2%), asymptomatic or symptoms not severe (14.1%), and hesitation from parents (9.2%). Being compared with the non-recurrent group, children in recurrent group were younger (<2.5yrs) at the time of surgery ($p=0.047$) and have a longer length of stay ($p=0.038$). In Cox analysis, when only considering preoperative

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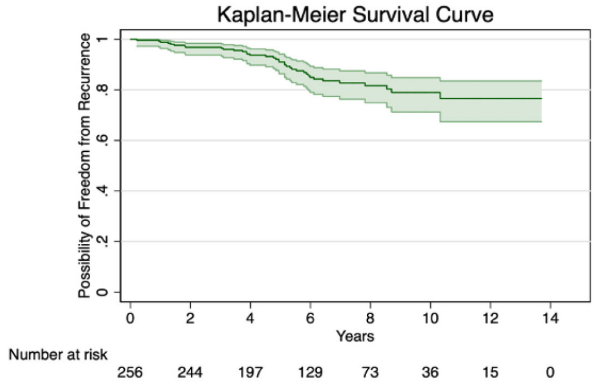
variables, the independent risk factor for LVOTO recurrence were a peak left ventricular outflow tract gradient higher than 60 mm Hg (HR 2.41, p=0.004), and length of SAS longer than 5 mm (HR 3.54, p<0.001). When both preoperative and intra-operative variables were considered, shorter distance between AoV and SAS (<4mm) (HR 2.62, p=0.047) and peeling from the aortic valve (HR 3.23, p=0.010) were independent risk factors for postoperative recurrence.
Conclusions: Immediate surgical intervention for LVOTO recurrence after diagnosis is recommended for SAS.

Keywords: Subaortic stenosis, congenital heart disease, cardiac surgery

Kaplan-Meier Survival Curve for the Study Cohort



PP-146
Subxiphoidal approach: A highly effective alternative for closing atrial septal defects in paediatric patients

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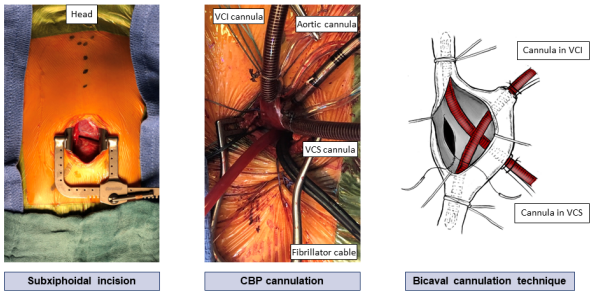
Background and Aim: The traditional approach for surgical closure of atrial septal defects (ASD) has shown excellent results with minimal risks. However, minimal invasive techniques like the subxiphoidal approach have gained popularity due to potential advantages such as reduced postoperative pain and improved cosmetic outcomes. This study aims to provide a comprehensive review of the operative technique and demonstrate the feasibility and outcomes of using the subxiphoidal approach for ASD closure.
Method: A retrospective analysis was conducted on all paediatric patients who underwent surgical ASD closure with the subxiphoidal approach or traditional sternotomy with minimal skin incision at our institution between January 2019 and October 2023. Subxiphoidal procedures involved bicaval and aortic cannulation and were performed without aortic cross-clamping on a fibrillating heart (Fig. 1). Clinical, procedural, and postoperative data were collected and analysed.
Results: Nineteen patients underwent ASD closure using the subxiphoidal approach, with a mean age of 4 years (range: 1-10 years). None of the procedures required conversion to a full sternotomy. Additionally, sixteen patients underwent ASD closure using sternotomy, with a mean age of 2.9 years (range: 0.4-8.8 years). In the subxiphoidal group, the median surgery duration and cardiopulmonary bypass time were 122 minutes (IQR, 41) and 34 minutes (IQR, 15), respectively. In the sternotomy group, these durations were 108 minutes (IQR, 39) and 28 minutes (IQR, 12). ASD size was similar between both groups. Neither group experienced early or late deaths, pericardial effusions, bleeding, or wound complications. The median hospital stay for both groups was 4 days. Follow-up assessments with echocardiography revealed one small, hemodynamically insignificant residual shunt in a patient from the sternotomy group. The cosmetic results following the subxiphoidal approach were considered excellent.
Conclusions: The subxiphoidal approach is a highly effective alternative to traditional sternotomy for closing ASDs in paediatric patients. This approach offers unique advantages, such as excellent cosmetic outcomes, preservation of the thoracic cage by avoiding sternotomy, and the ability to perform the procedure without cardioplegic arrest. By utilizing the subxiphoidal approach, patients can benefit from reduced surgical trauma, faster recovery, and improved cosmetic outcomes.

Keywords: secundum atrial septal defect, supxiphoidal approach, surgical treatment, minimal invasive procedure, atrial septal defect

Figure 1



An intraoperative photograph was taken following subxiphoidal incision and cannulation, along with a schematic illustration showcasing the bicaval cannulation technique in the subxiphoidal approach.

PP-147
A novel autologous heart valve for children and adults: a pre-clinical case report

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Background and Aim: Standard materials for transcatheter heart valve replacements (THVR) are currently limited to heterologous tissue. To overcome persistent constraints (e.g. limited lifetime,

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lack of adaptive growth), tissue-engineered THVR are subjects of ongoing research. The GrOwnValve project of the German Heart Center Charité Berlin is currently developing a novel procedure to manufacture an autologous THVR which should overcome these limitations and offer the first solution for pediatric and an improved solution for adult patients. As a prerequisite to an Investigator Initiated Trial (IIT), a pre-clinical trial with a porcine model has been performed.
Method: A pulmonary heart valve was shaped intra-operatively from autologous tissue and implanted in a minipig during a one stage intervention. Preceding the intervention, a CT-based model of the heart has been developed utilizing a 4D straightened segmentation approach to identify the prosthesis' landing zone and prevent a prosthesis-patient mismatch. Using this data and 3D printing technology, a patient specific mould for the intra-operatively shaped valve is subsequently produced. During the single stage intervention, a thoracotomy is performed in order to harvest pericardial tissue for valve manufacturing. Using the mould and a crosslinking agent for collagen stabilization of the autologous material, the tissue is shaped and sutured into a balloon-expandable stent. The valve replacement is crimped onto a balloon catheter and implanted via the femoral vein using a 22 French introducer sheath. After dilation of the stent in the landing zone and retrieval of the delivery system, transthoracic echocardiography, angiography and invasive pressure measurements were performed to assess the position, morphology, function, and dimension of the THVR.
Results: Post-operatively, the animal was left alive for another 2 hours showing good results regarding, paravalvular leakage, insufficiency, and migration of the THVR.
Conclusions: The first proof-of-concept in vivo study demonstrated the feasibility and safety of the GrOwnValve heart valve replacement. The method showed great functionality. The procedure will now be performed in a first-in-human IIT study, which is designed as a prospective, nonrandomized, single-centre trial in seven adult study subjects. In the future, the procedure can also be utilized for manufacturing autologous aortic heart valves.

Keywords: Heart Valve Replacement, Autologous Tissue, Minimal Invasive, Transcatheter

Education/Training

PP-148
International educational initiative: inaugural "fundamentals of inherited arrhythmias" symposium in zagreb, croatia

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Background and Aim: Multi-disciplinary Inherited Arrhythmia (IA) programs are the standard model of care in evaluating and managing patients and families with IAs. The aim of this symposium was to expand the fund of knowledge in IAs with the long-term goal of improving the quality of life and survival of children and adults at risk for sudden death in Croatia, where currently no formal IA program exists.
Method: An educational collaboration was established between the Pediatric Cardiology Divisions at SickKids and University Hospital Centre Zagreb to plan and execute a one-day educational symposium targeting pediatric and adult cardiologists and electrophysiologists, and cardiac surgeons. Itinerary included fundamentals of long and short QT syndrome, arrhythmogenic right ventricular cardiomyopathy, catecholaminergic polymorphic ventricular tachycardia, and Brugada syndrome. Invited faculty speakers were local and internationally recognized experts in their fields from Canada, USA and Europe. Success was measured by registrant feedback on learning outcomes and financial feasibility.
Results: There were 83 attendees at the symposium, of which 32 were registrants, 29 invited medical students, 15 organizing committee and faculty speakers, 3 industry representatives, and 4 invited guests. Of the 32 registrants, 28 (88%) were from Croatia, 2 (6%) from BiH, 1 (3%) from Slovenia and 1 (3%) from Sweden. Croatian registrants were practitioners from diverse geographical areas representing urban and rural areas. The attendees were predominantly female (24/32 registrants, 75%; 22/29 medical students, 76%; 7/8 invited speakers, 88%). Feedback on learning outcomes (scale 1-5) was received from 39 attendees, with an average score of 4.87 on the symposium meeting their expectations; 4.77 on improving their understanding of IAs; and 4.87 learning something new. The symposium was funded by industry sponsors at a total cost of €15,704.
Conclusions: The "Fundamentals of Inherited Arrhythmias" symposium was the first educational meeting on IAs and arrhythmogenic cardiomyopathy in Croatia and was successfully executed in a transparent and financially feasible manner, achieved its learning objectives, and served a geographically diverse underserved area. Gender diversity was evident and highlighted in the symposium. Ongoing educational initiatives will be imperative to solidify and build upon the information learned and hopefully expand to virtual case rounds and annual symposiums.

Keywords: education, collaboration, inherited arrhythmias, cardiomyopathy

Cardiac Dysrhythmias and Electrophysiology

PP-149
A case of severe qt prolongation caused by NAA10 P.ILE72THR, a rare, X-linked missense pathogenic variant

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Background and Aim: Hypertrophic cardiomyopathy (HCM) may prolong the QT interval, the degree of which typically corresponds to the degree of ventricular hypertrophy and outflow obstruction. NAA10-related neurodevelopmental syndrome (Ogden Syndrome) is a rare, X-linked condition caused by pathogenic variants in the NAA10-encoded catalytic subunit of the NatA N-acetyltransferase complex and characterized by developmental delay, dysmorphisms, hypotonia, prolonged QT, and cardiomyopathy.
Method: Not applicable.
Results: Case Presentation: A 6-year-old White male presented with cyclic vomiting. He was the term product of a naturally conceived, uncomplicated pregnancy and had an uncomplicated infancy. Global developmental delay and mild intellectual

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ECG of Case Presentation

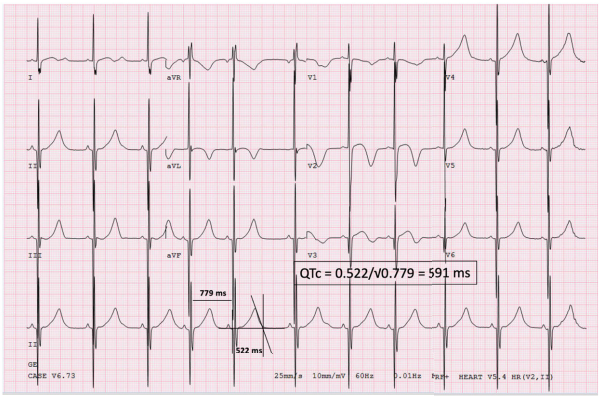

Figure. ECG of case presentation demonstrating sinus rhythm and severely prolonged QTc interval of 591 msec and abnormal broad-based T waves.

ECG of case presentation demonstrating sinus rhythm and severely prolonged QTc interval of 591 msec and abnormal broad-based T waves.

disability were diagnosed in early childhood. At age 10, he was evaluated for palpitations. Echocardiography revealed asymmetric hypertrophy of the interventricular septum (IVSd 0.67 cm, z score +0.6) with normal biventricular systolic function and no outflow obstruction. MRI demonstrated mild, mid-septal late gadolinium enhancement. ECGs showed sinus rhythm with broad-based, biphasic T waves with a prolonged QTc (523-591 msec), which persisted during exercise and recovery. Dysmorphic features were noted, including narrow nasal ridge, thin upper lip vermillion, hypotelorism, broad square chin, and left-hand broad base of digits 3-5. Parents are non-consanguineous. His brother has HCM and mild cerebral palsy and a maternal second cousin had sudden unexplained death. β-blocker therapy was initiated. Gene panels for HCM, arrhythmia, and cardiomyopathy did not identify a causative variant. Trio whole exome sequencing identified a pathogenic, hemizygous missense variant in NAA10 [NM_003491.4, Xp28]: c.215T>C, p.Ile72Thr, associated with X-linked Ogden Syndrome. The variant was inherited from his healthy mother. No other reportable or clinically significant variants were identified.

Conclusions: This is the first detailed description of severe QT prolongation caused by the NAA10 p.Ile72Thr variant. While 10-15% of HCM patients will have a prolonged QT interval, when the degree of QT prolongation is out of proportion to HCM severity and multiple organ systems are involved, rare genetic causes should be considered. Careful phenotyping to identify a unifying diagnosis is potentially life-saving, as both males and females with NAA10 variants can have QT prolongation. More research is needed to quantify the risk of sudden cardiac death in Ogden Syndrome and to determine optimal management.

Keywords: cardiomyopathy, arrhythmia, genetics

Education/Training

PP-150
looking through the crystal ball - feasibility of tele-echocardiography using smart glasses in neonates

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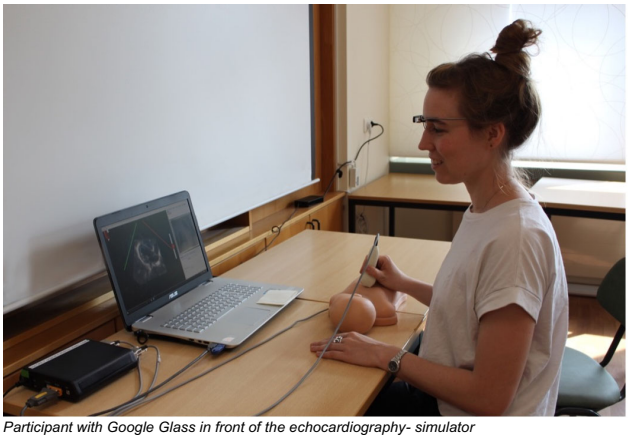
Background and Aim: In recent years, the importance of telemedicine has increased significantly. Virtual Reality glasses are a promising new tool in this context. Especially in the field of echocardiography, they offer the possibility of real-time data transmission without restrictions in the examination process. In particular, the care of critically ill newborns with suspected congenital heart disease might be improved by allowing a specialised paediatric cardiologist to remotely guide an echocardiographic examination. The current study aims to prove whether novices, under Google Glass guidance by an experienced pediatric cardiologist, can perform an appropriate neonatal echocardiography.
Method: The current study is a prospective monocentric single-blinded pilot study. As smart glasses, we used Google Glass. Participants were supposed to perform two test runs: The first test run as "unguided" and second test run instructed via Google Glass as "Google Glass guided". A validated training simulator for neonatal echocardiography "EchocomNeo, Echocom GmbH" was used. The study took place at the Leipzig Heart Center, Department of Pediatric Cardiology from April 2022 to November 2022.
Results: A total of 21 medical students were enrolled. In total 252 views (126 views in the first test run and 126 views in the second test run) were recorded. The overall performance was significantly higher in the Google Glass guided test run compared to "unguided" (structure score: 77.6% vs. 63.2%. p<.001 and quality score: 58.7% vs. 47.2%, p<.001). Also the time was significantly lower in the Google Glass guided test run (mean 40.1 seconds) than in the unguided test run (mean 47.7 seconds), p=.014.
Conclusions: Google Glass guidance by a pediatric cardiologist could optimize the performance of novices in echocardiography using a standardized neonatal echo- simulator with structural normal cardiac anatomy.

Keywords: telemedicine, pediatric cardiology, tele-echocardiography, smart glasses, digital health, neonatal functional echocardiography



Participant with Google Glass in front of the echocardiography- simulator

PP-151
Pen plus: supporting decentralization of pediatric cardiovascular care to rural communities in Africa

Charles Chale[1], Wellington Chirimaunga[1], Yolanda Sabino[2], Neusa Bay[3], Emily Wroe[4], Sam Patel[3], António Ribeiro[5], Gene Bukhman[4], Ana Mocumbi[6] and Ana Mocumbi[3]

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Background and Aim: In countries most people living in rural settings accessibility to quality care for paediatric cardiology (PC) is poor. PEN-Plus introduces essential health system interventions that focus on bridging critical gaps in training, referral pathways, and mentorship for diagnosis and management of these complex cardiovascular conditions children and adolescents. We describe the initial experience of establishing detection, referral and post-surgical management in rural Mozambique.
Method: Cascade training was used to train health providers in Hospital Rural de Nhamatanda (HRN) to screen, diagnosis and transfer/manage rheumatic heart disease (RHD) and congenital heart disease (CHD). Two general practitioners and eight mid-level providers (medical officer and nurses) were trained with the support of cardiologists from Mozambique, USA and Brazil, who customized the PEN-Plus training module and used digital platforms for tele-education/-mentoring. Tailored clinical protocols (incorporating physical examination, chest x-ray, electrocardiography and abbreviated ultrasound) were used for screening/diagnosis, management and referral for surgery. A six-month on-site coaching and mentoring program was established, including community-based activities to promote personalized medicine and retention in care. Data of the first 12 months is described using descriptive analysis.
Results: We have screened 44 children. Of the 50 among those with suspicion of either RHD (35) - including two with signs of RF - or CHD (15), 4 and 2 had confirmed diagnosis, respectively. The mean age of initial diagnosis was 13 years for RHD and 5 months for CHD; 100% in NYHA functional class II/IV; 4 had signs of malnutrition). The mean distance from home to the health facility was 17 km. 4 patients were referred for surgery in the capital; 1 was operated (mitral repair) and 3 are awaiting surgery (one diagnosed with RF).
Conclusions: Point-of-care devices and digital technology can bridge major gaps in PC care in rural settings, by improving access, enhancing disease detection, promoting efficient referrals and ensuring post-surgical follow-up. This decentralization of PC care allowed early detection and better management of RHD and CHD, improving their outcomes and supporting efforts towards global health equity.

Keywords: Decentralization, Taskshifting, Cascade Training, Paediatric Cardiology

PP-152
What do adolescents with a congenital heart disease know about their condition?

Isabel Gijón Criado, Natalia Fernández Suárez, Maria Teresa Viadero Ubierna, Jesús Garde Basas, Maria Celina Díaz Pérez and Maria Del Rosario Tazón Abascal
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Background and Aim: Congenital heart disease (CHD) has an estimated incidence of 5-8 per 1,000 live births. Treatment for CHD has improved dramatically in recent years, and 85% of children born with CHD, which was considered to be incompatible with life only a few years ago, now reach adulthood. They will need lifelong monitoring and sometimes reinterventions, so it is very important that they have adequate knowledge of their disease at the time of transition to adult cardiac care.
Method: The degree of knowledge of 39 patients between the ages of 11 and 20 with CHD who are followed up in our pediatric cardiology unit is assessed. Using a pre-formatted survey, their knowledge of their heart disease, interventions, medication, specific risks in relation to sport, contraception, pregnancy, etc. will be assessed.
Results: The sample consisted of 39 patients (53.8% male, 46.2% female) aged from 11 to 20 years, with a mean and median age of 14 +/- SD. CHD can be divided into simple (53.9% of the sample), including septal defects, bicuspid aortic valve, aortic coarctation, etc; and complex (46.1%), such as transposition of the great arteries, Fallot's tetralogy, hypoplastic left heart syndrome, etc. Is noted that 23% of the sample do not know that they have a cardiopathy, grouped in younger ages and in simple CHD. 77% know that they have a congenital cardiopathy and their level of knowledge is greater the simpler. In order to determine the dependency of "degree of knowledge" on other variables, a Fisher test is performed. All the tests performed have p-values >0.05, but gender is the less influenced variable (p-value = 0.054). With this in mind, the contingency coefficient (CC) and Cramer's V are calculated for the variable "knowledge", taking into account "CHD" and "age", showing a moderate significant association between these variables and child illness awareness (V = 0.56 and CC = 0.62).
Conclusions: Adolescents with CHD are usually a vulnerable population and do not have sufficient knowledge about their disease and specific risks. Transition programs from pediatric cardiology units to adult cardiology care are essential to provide them with information and health education.

Keywords: Congenital heart disease, transition programs, pediatric cardiology units, knowledge surveys

Figures

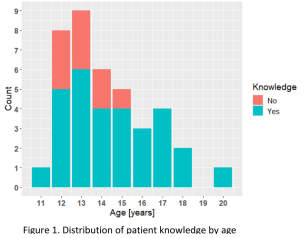

Figure 1. Distribution of patient knowledge by age

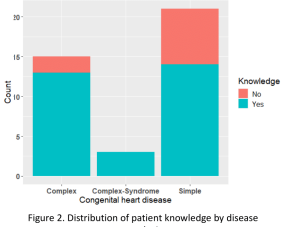

Figure 2. Distribution of patient knowledge by disease complexity

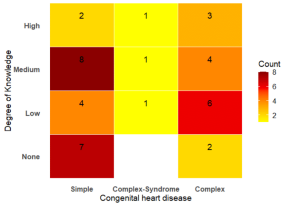

Figure 3. Joint distribution of disease complexity and degree of knowledge

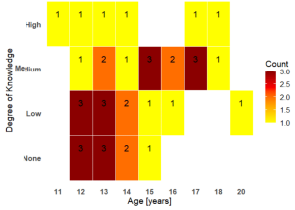

Figure 4. Joint distribution of age and degree of knowledge

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PP-153

Educational tools for enhancing rheumatic heart disease awareness in underserved populations

Jessica Abrams[1], Luz Marina Tacuri Chavez[2], Maria Do Carmo Pereira Nunes[2], Henrique Leão Fava[2], Matheus Henrique P Nunes[3], Mauro Henrique Agapito Da Silva[2], Beatriz Costa[2], Alexandre De Melo Ferreira[2], Fernanda Alves Gelape[4], Clareci Cardoso[5], Wanessa Compos Vinhal[5], Isabely Caroline Silva Ribeiro[5], Gabriela Aparecida Silva[5], Craig A Sable[6], Liesl J Zühlke[7] and Bruno Ramos Nascimento[2]

[1]Reach, Cape Town, South Africa; Paediatrics and Child Health, Red Cross War Memorial Children's Hospital, University of Cape Town, South Africa; [2]Universidade Federalde Minas Gerais, MG,Brazil; [3]Faculdade de Medicina de Barbacena, MG, Brazil; [4]Faculdade de Ciências Médicas, MG, Brazil; [5]Universidade Federal de São João Del Rei, MG, Brazil; [6]Children's National Hospital, Washington, USA; [7]Paediatrics and Child Health, Red Cross War Memorial Children's Hospital, University of Cape Town, South Africa; South African Medical Research Council, Cape Town, South Africa

Background and Aim: Rheumatic heart disease (RHD) continues to be a significant cause of premature deaths, emphasising the need for effective strategies to alleviate its burden. Education plays an important role in tackling this issue. The mission of Reach is to provide comprehensive technical support for local, regional, and global initiatives aimed at preventing and controlling rheumatic fever (RF) and RHD. This study aims to evaluate the effectiveness and usefulness of a set of educational flipcharts targeting healthcare providers, patients, and at-risk community in the state of Minas Gerais, Brazil.

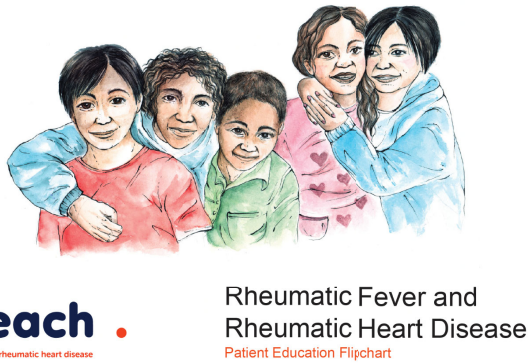
Method: Over 3-years, four flipcharts were developed, namely "Introduction to RF and RHD," "RHD and Pregnancy," "RHD and Surgery," and "RHD Community Awareness." These resources serve as instrumental tools for healthcare providers to engage with their patients and simultaneously raise awareness. Corresponding training and community education sessions were conducted, followed by evaluation surveys. These surveys were administered both electronically and in printed format in Belo Horizonte and Divinopolis throughout the year 2023.

Results: Between January and October 2023, a total of 46 training and education sessions were conducted. Flipchart training was delivered to 637 health and education professionals, while 458 patients, school students, and community members received education. Response to customised surveys were 84% (531/637) and 89% (408/458), respectively. Only 39% of professionals had prior training on RF/RHD, with 65% acquiring new information from the flipcharts, and 32% considering all the information to be new. Almost all professionals (97%) reported that the flipcharts could improve patients' lives. In the survey for patients, community members and schoolchildren, only 8 (2%) had prior education about RF/RHD, 211 (58%) reported they learned some new information and for 189 (46%) RHD information was all new. Notably, 78% (320 individuals) expressed their intention to discuss RHD with their families and communities.

Conclusions: The flipcharts can improve health and education professionals understanding of RHD and serve as a tool to guide their interaction with RHD patients. Simultaneously, they can be used to raise awareness among at-risk communities. We anticipate assessing the direct impact on RHD patients will bolster the rationale for continued investment in this program.

Keywords: rheumatic heart disease, rheumatic fever, health literacy, education, training

Flipchart Cover Page



The cover page for one of the educational flipcharts.

PP-154

Development of the parental e-learning resource to support discharge preparation using the congenital heart assessment tool

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Background and Aim: An early warning tool Congenital Heart Assessment Tool (CHAT) was identified in 2012 to address the gap in parents' understanding of signs of deterioration, appropriate contact and communication and earlier management by clinicians. A CHAT2 e-learning resource was developed in 2019 to enable health care professionals to appropriately prepare parents for discharge from hospital with their infant after complex cardiac surgery. There have been requests for a parental version, to further enhance consistency of parental preparation. AIM: To produce a digital resource that supports parents/carers to understand and use CHAT2 during preparation for taking their baby home.

Method: A quality improvement project, funded by the University of Worcester Learning and Teaching Fund, from August – September 2023. The Plan, Do, Study, Act method was chosen to design and manage the project, giving structure to meet NHS Patient Information guidance. Plan: Review the content of the healthcare professional's CHAT2 e-learning resource to ensure the content of the parental e-resource was aligned and consistent. Consultation with healthcare professionals to ensure accuracy of information regarding the clinical service delivery of parental CHAT2 education and support.

Results: Do: Create the parental CHAT2 e-resource using the web app Articulate Rise 360, a platform that enables the building of a fully responsive course through a variety of design applications. Use of the parental e-learning resource will be based on which parents are offered the Congenital Heart Assessment Tool by the medical and nursing teams in the cardiac centres. Parents will be encouraged to access the parental e-learning resource as part of the standard care at least 5 days prior to discharge from hospital and it will be the parents' decision whether to access the learning and they can access it whenever it suits them. In addition, an information leaflet has been created for community teams to enable them to support these families at home

Conclusions: Study: We are currently applying for ethical approval to engage parents in evaluation after a pilot use of the e-resource. Act: Following evaluation of parental feedback, the e-resource will be refined and launched.

Keywords: Congenital Heart Disease, Discharge, Home monitoring, Parental support, Education

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Fetal Cardiology

PP-155
Diagnostic yield of fetal echocardiogram referrals in a community based teaching hospital

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Background and Aim: The majority of congenital heart disease that is diagnosed in-utero is based primarily on suspected congenital heart disease from routine second trimester screening in low risk populations (no predisposing risk factors). However this data is predominantly based on academic centers with little focus on large community based teaching hospitals where the majority of children in the United States are born. Our hospital is a community based, academic affiliated, high volume birth center, averaging 3500 births per year. We sought to investigate our experience with regards to the diagnostic yield of congenital heart disease based strictly on referrals for suspected fetal heart disease.
Method: Retrospective review of initial referrals for suspected heart disease to a pediatric cardiologist for fetal echocardiogram over a 3 year period, 2020-2022. Referrals were divided between those from a general obstetrician and from a maternal fetal medicine specialists. The fetal echocardiograms were separated into three categories; no congenital heart disease noted, possible/mild heart disease or moderate/severe heart disease.
Results: The average total number of initial fetal echocardiograms during the study period was 325 per year (+/- 22). Of those an average of 19.2% were for suspected cardiac disease. The average yield, or summation of possible/mild disease and moderate/major disease, was 79% leaving 21% without any identifiable cardiac disease on fetal echocardiogram. The majority of those with suspected cardiac disease, 86%, were referred by maternal fetal medicine specialists, with only 14% referred by general obstetricians.
Conclusions: Our findings both agree and diverge from prior studies. The percentage of referrals for suspected cardiac disease, 19.2% is very similar to a recent large study (17.7%). However, our yield of 79% was significantly higher as compared to that study (41.1%). Importantly, in our center, a pediatric cardiologist is embedded within a maternal fetal medicine clinic. This allows for simultaneous collaboration, feedback and review prior to any referral for a formal fetal echocardiogram. In our view, regardless of the type of center (academic or community hospital) this setup is superior to the more common practice of referral for a fetal echocardiogram in a pediatric cardiology office.

Keywords: Fetal echocardiograms, fetal cardiac disease, fetal echocardiogram yield, maternal fetal medicine

PP-156
Prenatal diagnosis of coronary artery fistula connecting the left main coronary artery with the right atrium: A case report

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Background and Aim: We report a rare case of a CAF originating from the left main coronary artery and draining into the right atrium (RA).
Method: A pregnant woman was referred at the 21 weeks of gestation with a suspected congenital heart disease (CHD) on the ultrasound examination. Fetal echocardiography performed a large distal CAF arising from Left coronary artery (LCA) and draining into the right atrium (RA). The left CAF (diameter 2.6 mm) draining into the RA was visualized by color Doppler ultrasound. Pulsed Doppler recordings revealed characteristic bidirectional flow in the fistula. Turbulent diastolic reversed flow was detected from the blood steal phenomenon via CAF into the RA. The function of the left ventricle was within normal range. At 28 weeks of pregnancy, an increase in the size of the fistula to 3.6 mm was observed. However, the LV function was normal. Unfortunately, the woman did not follow up for a scheduled heart ultrasound because she had gone to another city. At 38 weeks of gestation, the woman gave birth. After birth, on ECHO, CAF. A fistula with a diameter of 0.55 cm from the lCA system was noted communicating with the right atrium. Moderate LV dilatation, FS 26%. In the first week of life, a percutaneous total closure of the fistula was performed at the cardiac catheterization laboratory. Follow-up echocardiogram the day after the procedure showed continued transformation into the LCA and RCA with a decrease in LV size and improvement in ventricular function.
Results: Our case report suggests that antenatal diagnosis of a CAF may aid in early intervention. Unfortunately, not all cases are identifiable before birth, and many of them are diagnosed after birth or later in life when symptoms become apparent. If there is a suspicion of a coronary artery fistula during pregnancy, it is important for a medical team with expertise in fetal cardiology to diagnose, evaluate, and plan appropriate care.
Conclusions: Early prenatal of CAF diagnosis is important so that transcatheter or surgical intervention can be made as early as possible. Total closure of the CAF in the cardiac catheterization laboratory is a safe and effective treatment.

Keywords: Congenital heart disease, coronary artery fistula, Fetal echo, cardiac catheterization.

Figure 1

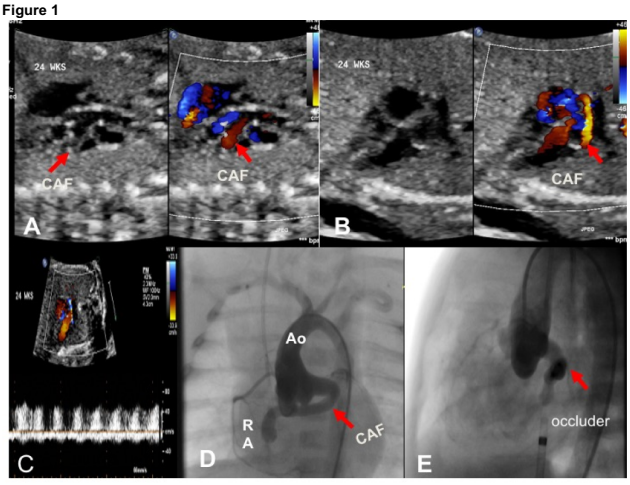

Figure 1. A,B, C - 2D and Color Doppler shows the presence of a fistulous tract arising from the left coronary artery and into the right atrium (RA). The filling of the fistulous tract is antegrade during ventricular diastole and reversed during systole. D- Selective left coronary artery angiography showing dilation of the fistulous tract and draining into the RA.E- After total closure of CAF. Ao- aorta; CAF - coronary artery fistula; RA - right atrium.

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PP-158

Umbilico-porto-systemic venous shunts

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Background and Aim: The poster aims to present the types of umbilico-porto-systemic venous shunts (UPSVS) and discuss their clinical relevance by using fetal cases and echocardiography images from our institute. We distinguish 3 main types according to the Achiron classification. The first is the umbilico-systemic shunt (USS), in which the ductus venosus (DV) is absent and the umbilical vein (UV) is connected to the right (rarely left) atrium, the subdiaphragmatic inferior vena cava (IVC) or the iliac vein. The other two types of UPSVS is the ductus venosus systemic shunt (DVSS) and the portosystemic shunts (PSS).
Method: The poster will present two USS cases by reviewing the fetal and postnatal periods. In both cases, UV drained into the right atrium. In the first case, maternal insulin-dependent diabetes mellitus and single umbilical artery, in the second case, persistent right umbilical artery and aberrant right subclavian artery were found along with significant cardiomegaly and USS.
Results: According to the literature USS is a severe abnormality and has a poor prognosis: circulatory failure, hydrops, high risk of intrauterine death and pulmonary hypoplasia may develop depending on the degree of cardiomegaly. In addition, liver lesions requiring transplantation may occur postnatally due to the underdevelopment of the v. portae system. Despite the poor literature data, our both cases showed excellent outcome during follow-up to date. The anatomy, clinical significance and outcome of the other two types of UPSVS (DVSS and PSS) will also be presented. In all types of UPSVS, cardiac developmental and chromosomal abnormalities (e.g. Down-syn., Noonan-syn., Turner-syn.) should be sought. The intrahepatic form of PSS has a better prognosis, but IUGR is common in these fetuses. In all types of UPSVS, cardiac developmental and chromosomal abnormalities should be sought.
Conclusions: The message of this poster is that systematic examination of the UV, portal vein and DV systems is of paramount importance in fetal echocardiography, as these complex abnormalities require both intrauterine and postnatal follow-up and may be associated with serious, even life-threatening complications.

Keywords: fetal echocardiography, umbilical vein, ductus venosus, umbilico-porto-systemic shunt

PP-159

Reference intervals for biventricular myocardial deformation in healthy fetuses using 2D speckle tracking echocardiography

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Background and Aim: In recent years fetal echocardiography has paid a special attention to the evaluation of ventricular function. The cardiopulmonary transition from fetal to neonatal circulation is a complex and multifactorial adaptation process. In this period, the ventricular physiology suffers important and progressive change. In this context, in addition to the management of structural heart diseases, quantification of ventricular function has been proposed for the evaluation and follow-up of the fetuses. Speckle-tracking ecocardiography (STE) has good fesability and reproductibility in assesing miocardial function in newborns and children, furthermore it overcomes many of the limitations of classic echocardiography. Nevertheless, one of the most important limitation of STE is that it is dependent of the echocardiographic devices and analytical programs used, thus pleading for individualized reference values. Our objective was to establish the reference interval in assessing fetal myocardial function, using two-dimensional (2D) STE for left ventricular (LV) and right ventricular (RV) longitudinal strain.
Method: We conducted a prospective study from May 2020 to September 2023, during which 150 healty fetuses were enrolled and underwent echocardiographic evaluation. 2D, apical 4-chamber view images of both ventricles were acquired using an Epiq echocardiograph and analyzed offline using a PhilipsQlab 13 autostrain function. The strain parameters analyzed were LV and RV peak global longitudinal strain (pGLS) and biventricular ejection fraction (EF).
Results: The gestational weeks median was 28.4 [20,42], all fetuses were with good intrauterine growth. The frame rate used was between 80-160 Hz. For the LV systolic function, the mean LVEF was 57.3% and the mean LV pGLS obtained was -15.2. For the RV systolic function, the mean RVEF was 42.7% and the mean RV pGLS obtained was 42.7.
Conclusions: Fetal myocardial function can be analized using STE. Larger scale studies may be of great interest for completion and validation of the depicted reference values.

Keywords: fetal cardiology, speckle-tracking echocardiography, myocardial function, peak global longitudinal strain

PP-160

How can an intrauterine wide ductus arteriosus mask a left hemitruncus associated with a rare complex cardiac malformation?

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Background and Aim: Hemitrunchus is an extremely rare cardiac malformation, with an incidence of 0.1%. The origin of the left pulmonary artery (LPA) from the ascending aorta is less common than its right counterpart. This malformation can be isolated or may coexist with other cardiac malformations, such as ventricular septal defect or tetralogy of Fallot (TOF). The absence of the pulmonary valve has been reported in 3-6% of patients with TOF.
Method: We present a case of a newborn with intrauterine growth retardation. The patient was prenatal diagnosed, at 34 weeks gestation with a complex cardiac malformation: Fallot type double outlet right ventricle with critical pulmonary trunkal and valvular stenosis, respectively what seemed to be two dilated pulmonary branches.
Results: In our case, the diagnosis of left hemitruncus was initially missed by prenatal echocardiography. Postnatal echocardiography revealed a complex cardiac malformation: Fallot type absent pulmonary valve syndrome with left hemitruncus. The pulmonary trunk was found in continuation to the right ventricular outflow tract, but at the valvular level the leaflets were rudimentary, producing both stenosis and insufficiency (z-score of -4 at the pulmonary annulus). Subsequently only the right pulmonary artery was

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found in the continuation of the dilated trunk, with the absence of the LPA. However a persistent ductus arteriosus was seen arising from the medial portion of the aortic arch and with continuity towards the LPA. To ensure blood flow through the LPA, continuous treatment with prostaglandin-E1 was initiated. Considering the low birth weight of 1900g and the hemodynamic stability, we decided a staged approach: continuation of prostaglandin-E1 treatment and temporization of the surgical correction until reaching a weight of 2500g. Thus at the age of 7 weeks, the patient underwent pulmonary valve commissurotomy and implantation of the LPA to the pulmonary trunk. Furthermore, as the pulmonary stenosis, given by the hypoplasia of the pulmonary annulus, has shown to be protective of the pulmonary vascular bed we decided to delay the intracardiac correction.
Conclusions: Fallot type absent pulmonary valve syndrome with left hemitruncus is a very rare cardiac malformation. A thorough and systematic prenatal echocardiogram is recommended to avoid missing this rare lesion.

Keywords: fetal cardiology, hemitruncus, absent pulmonary valve syndrome, echocardiography

PP-161
Restriction of the foramen ovale in a fetus with coarctation of the aorta- what next?

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Background and Aim: Restriction of the foramen ovale (RFO) or its premature closure represents a rare cardiac developmental defect. This anomaly may be found in otherwise structurally normal hearts or in association with congenital heart defects (CHD), such as hypoplastic left heart syndrome (HLHS). HLHS may be defined as a large spectrum of CHD characterized by the underdevelopment of the left sided heart structures, including the aorta, aortic valve, left ventricle, mitral valve and left atrium. Coarctation of the aorta (CoAo) is a narrowing of the isthmic region of the aorta.
Method: We present the case of a fetus in whom at the gestational age of 31 weeks the suspicion of CoAo arose, presenting a Z-score of -2 at the level of the isthmus. The mother underwent subsequent evaluations and was closely followed-up. At 36 weeks gestation fetal echocardiography revealed a RFO, alteration of the pulmonary venous flow pattern and ventricular disproportion with a borderline left ventricle. Given the diagnosis and its poor prognosis an emergency cesarean section was performed.
Results: Postnatal evaluation confirmed the intrauterine diagnosis: revealing a borderline left ventricle (mitral annulus Z-score= -3.75) with a moderate contractile dysfunction, bicuspid aortic valve, ductal dependent systemic circulation with a severe CoAo (Z-score -4.18 at isthmic level) and a large persistent ductus arteriosus. In this clinical setting prostaglandin-E1 infusion and inotrope support with Milrinone were initiated. At the age of 3 weeks the patient underwent surgical correction of the CoAo with termino-terminal anastomosis. The postoperative evolution was marked by the rebound of systemic hypertension in association with a residual gradient of 22 mmHg at isthmic level. The patient was discharged after 1 week and is currently under close follow-up.
Conclusions: RFO in association with CHD carries a poor prognosis, especially regarding the HLHS spectrum, as it may lead to a progressive underdevelopment of the left ventricle. These patients are in need of specialized, multidisciplinary care in order to establish optimal management protocols.

Keywords: restrictive foramen ovale, coarctation, fetal

Cardiac Dysrhythmias and Electrophysiology

PP-162
Flecainide friend or FOE?- The management puzzle of a case of persistent junctional reciprocating tachycardia

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Background and Aim: Persistent junctional reciprocating tachycardia (PJRT) is an accessory pathway mediated tachycardia of incessant nature, presenting a risk for tachycardia induced cardiomyopathy and often resistant to medical therapy, even with the use of multiple medications. Flecainide, a class IC antiarrhythmic drug, blocks voltage-gated sodium channels and has been found effective in treating supraventricular arrhythmias, particularly those of reentrant nature. Sustained fetal tachycardias are rare and the determination of the underlying arrhythmia mechanism is indispensable for choosing the appropriate medication.
Method: We present the case of a fetus with an ongoing regular tachycardia of supraventricular origin, showing a long ventriculo-atrial interval. Transplacental antiarrhythmic medication was initiated, but the treatment regimen (Digoxin monotherapy and in association with Amiodarone) failed to obtain a complete conversion of the dysrhythmia. The fetus was however in a good hemodynamic condition, showing no signs of cardiac decompensation until her birth at a gestational age of 41 weeks.
Results: The postnatal evolution was marked by the recurrence of the tachycardia showing the characteristics of a PJRT: long RP interval; regular, narrow QRS complexes and negative P waves in the inferior leads. Several antiarrhythmic treatment regimens (including administration of beta-blocker, Amiodarone and Flecainide) were attempted, but all failed to successfully control the arrhythmia. In evolution, during an attempt to increase the dosage of Flecainide, the patient presented broad complex tachycardia and hemodynamic compromise. As it was thought to be a modification given by Flecainide, the conversion of the dysrhythmia was obtained after the administration of sodium bicarbonate. The patient was later on referred to a pediatric electrophysiology center, where the ablation of the accessory pathway was successful and remained free from relapses at a follow-up of 2 years.
Conclusions: Fetal tachyarrhytmia that is refractory to transplacental treatment with multiple agents should raise concern for less common arrhythmia mechanisms, such as PJRT. In flecainide toxicity supraventricular tachycardias can present with wide QRS complexes, thus being easily mistaken for ventricular tachycardia.

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The management of these patients is based on the few cases published, sodium bicarbonate being considered a mainstay of treatment.

Keywords: fetal, tachycardia, flecainide

General Cardiology

PP-163
Echocardiographic early detection of cardiotoxicity induced by chemotherapy in children with leukemia

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Background and Aim: Cardiotoxicity induced by cytostatic therapy is a curent medical concern and early detection in order to institute supportive measures is essential. Cardiac dysfunction can occur during cytostatic therapy, immediately after or at a distance. Identification of robust non-invasive markers for risk stratification is very important. The current study aimed to assess cardiotoxic effects of chemotherapeutic regimens in a pediatric population, as revealed by assessment of baseline and end-of-treatment speckle-tracking echocardiographic indices.
Method: Study design - single center, prospective that included 28 children with acute leukemia.The inclusion criteria were acute onset of leukemia in pediatric population, who required chemotheraphy with normal structural and normal baseline functional heart. The exclusion criteria were patients with congenital heart disease, cardiomyopathy and patients who received prior chronic treatment with potential cardiotoxic adverse effects. All patients underwent complete transthoracic echocardiographic evaluation with Philips Epiq 7 machine. The examinations performed were M mode, two-dimensional, tissue doppler imaging and 2D speckle tracking prior to treatment initiation and at the following-up examination.
Results: The echocardiographic assessment after the first cycle of chemotherapy showed an impairment of the ejection fraction (EF) in 32% of cases, with a mean decrease of Teciholtz EF of 14% compared to the baseline assessment. Each was compared between the initial evaluation and the second evaluation, conducted after completion of chemotherapy/before maintenance treatment. A significant correlation between E' percentage variation and anthracycline cumulative dose, as well as antimetabolite agents was identified (p=0.03 in both cases) and cumulative dose of alkylating agents and global longitudinal strain (GLS). However, it has not been possible to identify an association between restrictive diastolic dysfunction or pseudonormalization after the first chemotherapy treatment and cumulative doses of chemotherapeutics
Conclusions: GLS is a useful echocardiographic parameter in the detection of cardiac dysfunction before clinical manifestations. In addition to the effect of chemotherapy on myocardial function, the effect related to the dose of chemotherapy must be evaluated. Further studies with larger population are needed.

Keywords: cardiotoxicity, echocardiography, speckle-tracking, pediatric cardiology, leukemia

Fetal Cardiology

PP-164
Influence of prenatal echocardiography and mri on postnatal outcome in fetuses with isolated double aortic ARCH

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Background and Aim: In the general population, double aortic arch (DAA) occurs in up to 0,01%. There is no consensus on perinatal management and prediction of early postnatal symptoms in isolated DAA. The possibilities of multimodal imaging in prenatal diagnosis of this pathology for predicting early postnatal outcome were analyzed.
Method: Between January 2012 and February 2023, 32 fetuses with isolated DAA were identified at our institution. For prenatal assessment of the degree of tracheal stenosis, since 2017 we have performed fetal MRI (n=20). According to the MRI results, all fetuses were divided into 2 groups: 1) low-risk group (n=9; 45,0%) without significant (<50%) tracheal compression; 2) high-risk group (n=11; 55,0%) with significant (>50%) compression. All symptomatic children underwent angio-CT after birth.
Results: The mean age of 32 fetuses at the initial diagnosis of DAA was 26,7±4,7 weeks. The mean age of 20 fetuses at MRI was 30,5 ±2,4 weeks. After birth, in all newborns (n=32) the diagnosis of DAA was confirmed by echocardiography at an average age of 10,3±11,7 days. In the postnatal period, 25 (78,1%) children became symptomatic and were operated on an average of 168,6 ±161,7 days, among them 8 (25,0%) were newborns. All patients survived, except 1 (4,0%), who had IUGR and was born prematurely with a weight of 1350 g. Of the 7 (21,9%) unoperated patients, 6 are asymptomatic, the average follow-up period is 796,5 days. One patient was lost to follow-up.
When comparing groups with fetal MRI, we found that all (n=11; 100%) children from the high-risk group had symptoms and underwent surgery (mean age: 146,8±135,6 days), including 3 (27,3%) neonates. Whereas, from the low-risk group, only 4 (44,4%) had surgery and were operated not significantly later (mean age: 294,7±198,7 days, p=0,44) with 1 (11,1%) neonatal case. Preoperative degree of tracheal stenosis according to angio-CT data was significantly higher in the high-risk group (75,8±11,1% vs. 44,8±6,1%, respectively, p<0,01), which correlated with prenatal MRI data.
Conclusions: Fetal MRI with assessment of tracheal stenosis allows to distinguish fetuses with isolated DAA who are at high risk for neonatal and early infant symptoms and interventions, which is an important for clear planning of perinatal management.

Keywords: isolated double aortic arch, prenatal diagnosis, fetal MRI, perinatal outcome

PP-165
Has the outcome of prenatally diagnosed absent pulmonary valve syndrome improved?

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Background and Aim: Absent pulmonary valve syndrome (APVS) has high morbidity and mortality reported. The objective was

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to report the outcomes in a contemporaneous cohort and compare the outcomes to a previous cohort from our unit.
Method: Retrospective observational study examining prenatal findings and postnatal course of fetuses prenatally diagnosed with APVS between August 2000-2022 and comparison to historical cohort from our unit (1988-2000). Inter-cohort outcomes were compared using Fisher's test.
Results: There were 35 fetuses diagnosed with APVS at median gestation 21weeks' (range: 12-39). All fetuses had a VSD and an arterial duct was present in 2/35 (6%). Genetic testing was undertaken in 25/35 cases of which 8/25(32%) had a genetic anomaly, most commonly 22q11 microdeletion (2/25, 8%). Continuity of pregnancy occurred in 22/35(63%), intrauterine demise occurred in 1/35(3%) and there were 3(14%) neonatal deaths (right ventricular failure, preterm neonate with necrotising enterocolitis, persistent pulmonary hypertension of newborn).
Eighteen infants underwent surgery: two in the neonatal period, one is on long-term ventilation (LTV) for 17 years and another is alive without respiratory morbidity. A further 16 had surgical repair in childhood at median 1.7years (range 0.2-6.3) of which one had a second surgery to alleviate ongoing airway compression (aortopexy and PA patch plasty) and subsequently left pulmonary artery stenting. 17/21(81%) are alive at median age 11.5years (range:1.4-23.1).
Four fetuses underwent fetal MRI and two showed features of fluid-trapping consistent with bronchial compression, one of which remains LTV dependent, the other requires no ongoing respiratory support. In total 7/21(33%) of live-born babies required intubation within 24-hours of birth, of those 3 died and 2 have ongoing LTV and two are alive without respiratory support. Of the 14 who did not require ventilation at birth none have long term respiratory morbidity.
Compared to the previous cohort from our institution (n=20) infant mortality has reduced and infant surgical outcomes have improved (see table).
Conclusions: The highest risk for mortality was in the neonatal period. Intubation at birth is a risk factor for respiratory morbidity and mortality. Outcomes from infant surgery and survival to 1 year have improved in those that survive the neonatal period.

Keywords: Absent pulmonary valve, fetal cardiology

Data table

	Current cohort (n=35)	Historic cohort (n=20)	*P value*
Termination of pregnancy	13/35 (37%)	6/20 (30%)	*0.77*
Intrauterine demise	1/22 (5%)	3/14 (21%)	*0.28*
Live births	21/35 (60%)	11/20 (55%)	*0.78*
Neonatal mortality	3/21 (14%)	5/11 (45%)	*0.09*
Infant mortality	0/21 (0%)	3/11 (27%)	*0.03**
Surgical survival (neonatal repair)	1/1 (100%)	0/3 (0%)	-
Surgical survival (childhood repair)	16/16 (100%)	2/4 (50%)	*0.03**
Alive at 1 year	18/21 (86%)	3/11 (27%)	*0.002**

Comparison in outcomes between previous and current cohort of APV patients.

PP-166
Management and outcomes of vein of galen malformation in a neonate: A case study

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Background and Aim: Vein of Galen malformation (VOGM) is a rare and complex congenital vascular anomaly involving the cerebral venous system with secondary consequences on the heart and the circulatory system due to abnormal blood flow patterns. This case study presents prenatal diagnosis, the clinical course, and the management of a neonate diagnosed with VOGM.
Method: CASE: We present a case of a newborn male child, delivered via cesarean section at 38 weeks gestational age following spontaneous conception. During the prenatal ultrasound examinations of the fetus, there was a suspicion of A-V malformation in the brain and evidence of hypertrophy on the right side of the heart.
Results: Fetal echocardiographic findings confirmed cardiomegaly (cardiothoracic index [CTI] 0.65), myocardial hypertrophy, dilatation of the right heart chambers, and relatively significant tricuspid valve regurgitation (+2/4) with a max. peak velocity of 2 m/s. No intraventricular septal defects were identified, but both atrial chambers displayed wall hypertrophy (4-5 mm), with observed changes that correspond to signs of fetal heart failure. Also, a fetal MRI and postnatal MRI brain imaging revealed an aneurysmal malformation measuring approximately 6.2 cm in length and 2.9 cm in width with a broad drainage in the rectus sinus and sinus confluence
Conclusions: Endovascular embolization treatment was undertaken to occlude the abnormal arteriovenous connections with the aim of normalizing blood flow and reducing potential heart complications associated with VOGM.
This case underscores the complexities inherent in managing VOGM in neonates. Timely diagnosis, a multidisciplinary approach, and minimally invasive endovascular procedures play a crucial role in the long-term prognosis.

Keywords: Vein of Galen malformation, Fetal heart failure, Endovascular embolization

PP-167
Prenatal diagnosis of the right aortic ARCH: The multicenter arcade study

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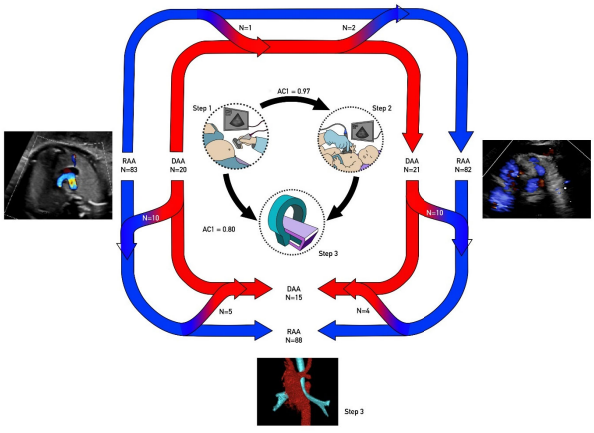
Background and Aim: To describe the various presentations of the prenatally diagnosed isolated right aortic arch (RAA) i.e., without associated congenital heart defect, and to evaluate the performance of prenatal diagnosis of isolated RAA in terms of postnatal outcome.
Method: In this multicentric retrospective study, from 2010 to 2019, all live births with a prenatal ultrasound diagnosis of isolated RAA were included, with a 1-year postnatal follow-up. The concordance between the different diagnostic stages (prenatal ultrasound, postnatal ultrasound, and post-natal CT-scan) was evaluated using Gwet's AC1 coefficient.
Results: A total of 309 cases of prenatally diagnosed RAA were analyzed, most of which had a left ductus arteriosus (83%). The concordance between prenatal and postnatal ultrasound diagnosis was excellent regarding the RAA type (AC1=0.97, 95% confidence interval (CI)= [0.94;0.99]). The rare discrepancies mainly involved non-diagnosed or misdiagnosed double aortic arch (2%). CT-scan was performed in 108 neonates (35%) and the concordance between prenatal ultrasound and postnatal CT-scan was good regarding the RAA diagnosis (AC1=0.80, 95%CI= [0.69;0.90]) but poor regarding the distribution of brachiocephalic vessels (AC1=0.21, 95%CI= [0.06;0.36]). An associated genetic anomaly was sought for in half of the cases and identified in 4% of the cohort. During the first year of life, 50 (18%) infants presented with vascular ring symptoms, and 24 (8%) underwent aortic arch surgery.
Conclusions: This multicentric nationwide cohort of 309 prenatally diagnosed isolated RAA demonstrated the reliability of prenatal screening, highlighted the rare cases of discrepancies between prenatal and postnatal diagnosis, and underlined the value of CT-scan to improve the postnatal follow-up.

Keywords: aortic arch, vascular ring, antenatal diagnosis, CT-scan, genetic counseling

Diagnostic concordance between Step 1 (prenatal ultrasound), Step 2 (postnatal ultrasound), and Step 3 (CT-scan).



The blue arrow represents the diagnostic stages in the RAA population between Step 1 (prenatal ultrasound), Step 2 (postnatal ultrasound) and Step 3 (CT-scan). The red arrow represents the diagnostic stages in the DAA population between Step 1 (prenatal ultrasound), Step 2 (postnatal ultrasound) and Step 3 (CT-scan). The color changes of the arrows represent the diagnostic changes at each step, between RAA and DAA (blue to red) and between DAA and RAA (red to blue). Abbreviations: S-RAA, solitary right aortic arch; DAA, double aortic arch; Step 1, prenatal ultrasound; Step 2, postnatal ultrasound; Step 3, CT-scan; AC1, concordance coefficient.

PP-168

Prenatal imaging of fetal cardiac valves using short axis view. Focus on isolated valvular diseases and its outcomes

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Background and Aim: To assess the feasibility of prenatal imaging of fetal cardiac valves anatomy using short axis view with particular emphasis on the detection of its morphology and its abnormalities
Method: This study was a retrospective study of fetuses with congenital isolated cardiac valvular disease which had undergone both 2D and 3D/4D STIC echocardiography using short axis view and its outcome. Study period from October 2019 to October 2023.
Results: A total of 3455 fetal echocardiography was performed during the study period. Congenital heart defects were identified in 156 fetuses. Isolated cardiac valvar disease were seen in 21 fetuses [Tricuspid valve (TV) dysplasia (10), absent inferior TV leaflet (1), TV prolapse (3); Mitral valve prolapse (3); Bicuspid aortic valve (7); Unicuspid aortic valve (1)]. Genetic evaluation was performed in 7 fetuses. Polyvalvar dysplasia noted in three fetuses was associated with a unique mutation [6q25.1 (TAB 2) deletion]. 12 [tricuspid valve dysplasia (8); polyvalvar dysplasia (2); bicuspid aortic valve (2)] couples opted for termination of pregnancy; 1 fetus had intrauterine death. 2D echocardiography of short axis view was superior to 3D/4D STIC echocardiography in predominant cases.
Conclusions: Prenatal imaging of fetal cardiac valves using short axis view gives additional information of the cardiac valve morphology when compared to standard fetal echocardiography views. This information will be useful in decision making and for providing effective prenatal counselling.

Keywords: Fetus, cardiac valves, short axis view

PP-169

Accuracy of the prenatal diagnosis of congenital heart disease

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Background and Aim: Prenatal diagnosis of congenital heart disease (CHD) leads to an improvement in postnatal survival. The main objective of the study was to determine the accuracy of prenatal diagnosis of CHDs in our center.
Method: A retrospective cohort study of CHDs diagnosed prenatally was carried out in a reference tertiary center in Spain during 2021, obtaining data from 111 pregnant women and their respective newborns. The severity of the CHD was classified using the Fetal Cardiovascular Disease Severity Scale (grading from 1 to 7); and depending on the scale of 1-3, 4-5 or 6-7, they were classified as low, medium or high severity. The accuracy of the diagnosis was determined based on whether it was complete/incomplete/incorrect; the correlation between the determination of pre- and postnatal severity was determined; and the possibility that different variables influenced the accuracy of the diagnosis.
Results: The most frequent diagnoses were suspected prenatal aortic coarctation (24.3%) and postnatal supra-aortic vessel anomaly (22%). The majority of CHDs were classified as mild severity (67.6% prenatal and 68.8% postnatal). The diagnosis was correct in 84.4% of CHDs, incomplete in 9.2% and incorrect in 6.4%. Prenatal severity grading using the Fetal Cardiovascular

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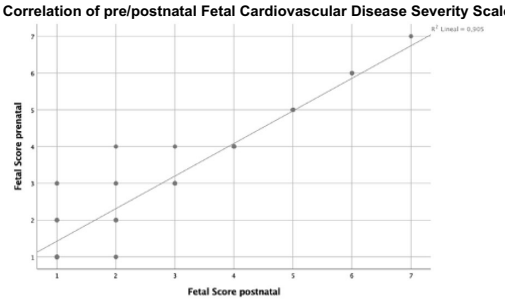
Disease Severity Scale was concordant with postnatal grading in 76.1% of cases; the prediction of severity as low, medium or high was correct in 98.2% of the CHDs. The correlation of the pre- and postnatal Fetal Cardiovascular Disease Severity Scale was high (Pearson's correlation coefficient 0.951 $p<0.005$); as well as the grading of severity as low, medium or high ($p<0.005$). There was a greater accuracy in grading severity at lower gestational age of the fetus at diagnosis ($p<0.005$).
Conclusions: In the prenatal diagnosis of CHD, it is important to correctly predict the severity of the disease. In our study, a correct diagnosis was made in 84.4% of the cases and, when determining the severity (low, medium or high), the correction was 98.2%.

Keywords: congenital heart disease, fetal diagnosis, accuracy



Representation of the high positive correlation between pre and postnatal Fetal Cardiovascular Disease Severity Scale (Pearson's correlation coefficient 0.951 p<0.005).

PP-170

Fetal arrhythmias: Twenty years of experience in a tertiary care center

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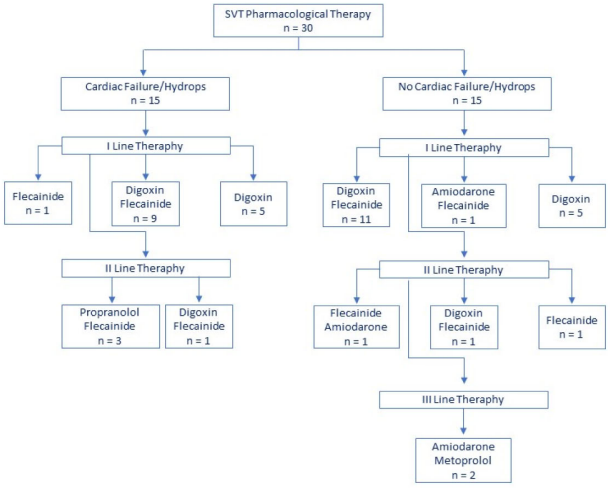
Background and Aim: Management of fetal arrhythmias has achieved great success in the last decades. Ultrasound evaluation and Doppler technology are indispensable in diagnosis. Treatment usually involves the second and third trimester of pregnancy. Even if the majority of fetal arrhythmias are benign, both bradi and tachi arrhythmias can bring to cardiac failure up to fetal hydrops requiring pharmacological intervention.
Method: Data of fetal arrhythmias were retrospectively collected from 2002 to 2023 in an Italian tertiary care centre, Policlinico Sant'Orsola Malpighi in Bologna. Detailed anatomical evaluation with fetal echocardiography were performed in all cases to determine whether a structural cardiac and extracardiac anomaly was also present. Fetal arrhythmias were classified under two main groups: tachyarrhythmia and bradyarrhythmia; a third group of unconfirmed diagnosis was reported including all referral from other centers. The following data were collected: prenatal diagnosis, gestational age at diagnosis, therapy administrated, outcome of pregnancy and outcome of neonates.
Results: Out of 87 women who received a diagnosis of fetal arrhythmia, 61 (70.9%) were classified as tachyarrhythmias, 22 (25.6%) as bradyarrhythmias and 3 (3.5%) were not confirmed. Most of tachyarrhythmias were supraventricular tachycardia (n = 30, 34.9%) with a mean gestational age of 27 ± 5 weeks, half of them had with signs of cardiac failure up to hydrops. All SVTs underwent pharmacological therapy. Digoxin was started primarily as first-line therapy in 28 patients, 20 in association with flecainide. As second and third line therapy, other antiarrhythmic agents (amiodarone, metoprolol). Twenty-seven (90%) of fetus converted on sinus rhythm in a mean time of 6,84 days, one (3.3%) died at birth; therapy was conducted for 57 ± 35 days. Bradyarrhythmias were mostly third grade AV-block (n = 14, 63,6%) with a mean fetal ventricular frequency of 63±3 bpm. Autoantibodies (antiRo-SSA and antiLa-SSB) were found in 64% of those patients (n=9). Desametasone was the drug of choice with a mean dosage of 6 mg.
Conclusions: In our centre, as well as in literature, the most common cause of fetal arrhythmias was tachyarrhythmias. The choice of treatment should depend on the condition of the mother and fetus as well as the center experience

Keywords: Fetal arrhythmia, fetal tachyarrhythmia, fetal bradyarrhythmia, hydrops, transplacental therapy

SVT Pharmacological Therapy



PP-171

Prenatal prediction of coarctation of the aorta: A systematic review and individual participant data meta-analysis

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Background and Aim: the prognostic value of prenatal ultrasound to diagnose postnatal coarctation of the aorta (CoA) remains unclear. The aim of the study is to report the strength of association and diagnostic accuracy of different US signs in detecting CoA prenatally.

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Method: systematic review and individual participant data meta-analysis. The reference standard was CoA diagnosed after birth defined as narrowing of the aortic arch and the index test included the most commonly evaluated parameters on US both in B-Mode and Doppler. Summary estimates of sensitivity, specificity, diagnostic odds ratio (DOR), and likelihood ratios were computed using hierarchical summary receiver–operating characteristics.
Results: Seventeen studies with 640 fetuses meeting inclusion criteria were analyzed. At random effect logistic regression analysis, most continuous variables were associated to CoA but had low diagnostic accuracy. Regarding categorical parameters, tricuspid/mitral valve ratio > 1.4 was significantly associated to CoA (p<0.001) and had a sensitivity of 72.6% (95%CI, 48.2-88.3), a specificity of 65.4% (95%CI, 46.9-80.2), and a DOR of 5.02 (95%CI, 1.82-13.9); the respective sensitivity/specificity (95% CI) figures for pulmonary artery/ascending aorta ratio >1.4 were 75.0% (61.1-86.0)/39.7% (27.0-53.4), aortic isthmus z-score <-2 at sagittal 47.8% (14.6-83.0)/87.6% (27.3-99.3) and aortic isthmus at 3-vessel views were, 74.1% (58.0-85.6)/62.0% (41.6-78.9). Hypoplastic arch had a sensitivity of 70.0% (42.0-88.6), a specificity of 91.3 (78.6-96.8), and a DOR of 24.9 (6.18-100). A multivariate scoring model was developed using the most commonly evaluated parameters (gestational age<28 weeks, tricuspid/mitral valve ratio >1.4, pulmonary/ascending aorta ratio >1.4 and aortic isthmus at the 3 vessel view z-score<-2) as well as one with those with the highest diagnostic yield in the meta-analysis (tricuspid / mitral valve ratio>1.6, hypoplastic arch; aortic isthmus 3-vessel view z-score<-2 and gestational age<28 weeks) and both showed poor performance with a highest sensitivity of 68.2% and specificity of 50.0% in the first one when 3 out of the 4 parameters were met.
Conclusions: Several prenatal ultrasound parameters are associated with an increased risk for postnatal CoA. However, diagnostic accuracy is only moderate, even when combined.

Keywords: Coarctation of the aorta, fetal, ultrasound, prenatal, prediction

PP-172

Pulmonary atresia/critical stenosis with intact ventricular septum: differences in perinatal management

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Background and Aim: Pulmonary atresia/critical stenosis with intact ventricular septum remains a complex entity with different proposed approaches both prenatal and postnatally. Our aim was to evaluate differences among different referral centers
Method: An online survey was developed by experts in prenatal and pediatric cardiology. The survey was distributed among centers in Europe and USA. One questionnaire was evaluated by center and results were evaluated in a descriptive manner.
Results: A total of 45 centers responded to the questionnaire. Of them, 25/45 evaluated at least 5 prenatal cases per year, and 13/45 evaluated at least 5 postnatal cases per year. All had neonatal cath lab and surgery units and 30/45 had a heart transplant programme. Gestational age at diagnosis usually occurred after the anomaly scan in 60.5% of cases (mostly between 22 and 28 weeks) and at 16-22 weeks in 39.5%. In 52.4% of cases, prenatal counseling was based on several models rather than one as a standalone and in 21.4% of cases solely on the z-score of the tricuspid valve. Pulmonary valvuloplasty was considered in 60.5% of cases and in 39.5% it was not considered an appropriate treatment for the condition. Termination of pregnancy rates varied among centers, mostly by geographical location but were <10% in 66.7% of centers. Regarding management and outcomes, one-and-a-half ventricle solutions were an alternative in all centers and non-univentricular outcomes were <25% in 26.2% of centers, between 25-50% in 33.3%, between 50-75% in 26.2% and >75% in 14.3%
Conclusions: There are wide differences in prenatal management and perinatal results of cases with pulmonary atresia with intact ventricular septum, reflecting a lack of solid evidence in a rare pathology. There is a need for large international studies that may pool efforts together in order to standardize management and offer better evidence-based recommendations.

Keywords: Pulmonary atresia, fetal medicine

PP-174

Fetal pericardial effusion and mca doppler survey among fetal cardiologist and fetal medicine specialists in the uk

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Background and Aim: Fetal pericardial effusion (PE) could be seen in a wide clinical spectrum, ranging from normal to severe fetal pathologies. There is no consensus on the normal appearance of PE in fetuses or the investigation of the causes. We report the information obtained from the fetal cardiologists and fetal medicine specialists in the UK centers on existing practices and individual preferences on the assessment and investigation of PE.
Method: Total 27 consultants were sampled, of whom 8 were fetal medicine doctors, 19 fetal cardiologists. Two questions were asked: (1) What is the normal limit for PE in fetus with no other pathology?" and (2) Do you think MCA Doppler is fetal cardiologist's remit to do in routine fetal cardiac examination?
Results: 14 participants chose the limit for normal acceptable size PE in fetus with no other pathology as under 2 mm. 7 participants added the opinion that the position and extent of the PE and cardiac cycle as being important when the measurement of the diameter was taken. Two fetal cardiologists would not measure it. One participant chose the normal limit for PE as under 2.5 mm. 10 participants accepted the normal limit of PE as under 3 mm. All responds are shown in the table 1. The latter thought that it would be more reliable to have a slightly higher limit to avoid false positivity. Only 2 participants were advocating MCA Doppler assessment by the fetal cardiologist. Overwhelming majority of the participants (25/27) indicated that MCA Doppler is the remit and responsibility of the feto-maternal medicine consultants.
Conclusions: Most of the UK fetal cardiologists and feto-maternal medicine specialists would consider under 2mm as the normal limit for physiological PE in the fetus. In addition, the position and extension of the fluid, the associated abnormalities were taken into

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account in grading PE severity. MCA doppler is almost universally agreed to be the business of feto-maternal specialists instead of paediatric cardiologists. Even in the presence of hydrops and other systemic abnormalities, PE should be primarily investigated by feto-maternal specialist rather than making an immediate referral to a paediatric cardiologist.

Keywords: fetal pericardial effusion, fetal cardiology, survey, MCA Doppler, fetal cardiologist, fetal medicine specialists

PP-175

Can the size of fetal pericardial effusion be predictive of aetiology?

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Background and Aim: Physiological fetal pericardial effusion (PE) could be detected during normal fetal anomaly scan without any pathology. Differentiation of pathological PE due to other severe diseases cannot be decided solely upon the size of separation between visceral and parietal pericardium. In this study, we evaluated the etiology and outcomes of fetuses with pericardial effusion in South Wales over the past 20 years.
Method: Fetuses diagnosed with PE between 2001 and 2022, were analyzed retrospectively at a tertiary pediatric cardiology center in Wales, UK
Results: 51 fetuses were estalished with PE. Mother's age was between 19 to 46. Gestational week at diagnosis was between 14 to 39 weeks. Pericardial effusion less than 3mm was defined as small-mild PE, 3 to 5mm as moderate, more than 5 mm as massive/large. Congenital heart diseases (CHD) were found in 16 cases: complete AVSD (5), large VSD (4), critic aortic stenosis (2), pulmonary hypoplasia/atresia (1), tricuspid atresia (1), Ebstein anomaly (1), and mitral atresia (1). Intrapericardial teratoma was diagnosed in one patient. 39 patients had mild/small PE. 5 patients had moderate PE. 7 patients had massive PE and hydrops.12 patients with mild PE and 2 patients with moderate and large PE had complex CHD. Massive PE and hydrops were noted in 2 patients with fetal tachycardia, whilst one patient with fetal AV block had also hydrops and large PE. Trisomy 21 were confirmed in 5 patients, of 1 had large massive PE with complete AVSD. 3 patient had fetal anemia, one patient had Vein of Galen aneurysm. A large number of mothers 17/51(33.3%) had history of smoking, alcohol drinking and one with heroin addiction. Mortality rate was 11/51-21.5% of which 6 had complex CHD (2: critic aortic stenosis, 1: Ebstein anomaly, 1: mitral atresia and hypoplastic left ventricle, 1: trisomy 21 and large VSD, 1: large VSD), 2 had fetal anemia, 1 had pericardial teratoma, 2 had fetal arrhyhtmia.
Conclusions: Fetal PE has usually a good prognosis when it is mild and not associated with genetic abnormalities, complex CHD, and/or prematurity. Other systemic diseases like fetal anemia, vascular malformations should be evaluated in the absence of cardiac pathology.

Keywords: fetal pericardial effusion, hydrops, congenital heart disease

Cardiac Dysrhythmias and Electrophysiology

PP-176

Epidemiology of supraventricular tachycardia in paediatric population: 30 years review in south wales

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Background and Aim: To identify the population characteristics of children presenting with supraventricular tachycardia (SVT) and their acute and long-term responsiveness to treatment.
Method: All patients with documented supraventricular tachycardia identified from the National Welsh Paediatric Cardiology database, Cardiobase were analysed retrospectively. For analysis, six age categories were defined and these were foetal, neonatal (<28 days), infant (< 1 year of age), childhood (<13 years of age) and in teenage years (<17 years of age). Those with unclear documentation of age at presentation were categorised under 'unknown'.
Results: A total of 796 patients were identified with documented SVT until July 2021. An additional 33 patients were identified from fetal cardiology database. 61 patients were excluded for not fulfilling inclusion criteria. 225 patients did not have mechanistic classification of their SVT diagnosis, hence they were excluded from this preliminary analysis, leaving a final population of 525 patients. 275 patients (52.38%) were male. The first episode of SVT most commonly occurred below 1 year of age followed by childhood (Table 1). Wolff Parkinson White syndrome was the leading cause of SVT in 227 patients (43.24%) followed by AVNRT (Table 2). 164/525 (31.24%) received adenosine with or without other adjunct treatment. 29/525 (5.52%) required emergency cardioversion for termination of acute SVT attack. 6/525 (1.14%) required CPR immediately. 264/525 (50.29%) underwent electrophysiology study and ablation procedures. In 41 (7.8%) infants (but not in older children) arrhythmia resolved spontaneously and medication could be ceased.
Conclusions: SVT mostly commonly manifest in children below 1 year of age and the leading mechanism is WPW syndrome. Although it is more prevalent and resistant to treatment in early periods, spontaneous resolution of SVT occur more often in this group compared to the older children.

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Keywords: cardiac arrhytmia, supraventricular tachycardia, cardioversion

Table SVT

Table 1. 525 patients at time of diagnosis by age category.

Age category	Frequency (n)	%
Fetal	71	13.71
Neonate	89	16.95
Infant	53	10.10
Child	197	37.52
Teenager	88	16.76
Unsure	26	4.95

Table 2. 525 patients stratified by arrhythmia subgroup.

Diagnosis	Frequency (n)	%
AVRT	358	68
WPW	227	43.24
AVRT due to CAP	95	18.1
PJRT	33	6.29
Mahaim	3	0.57
AVNRT	97	18.48
AET	29	5.52
Atrial flutter	22	4.19
Atrial fibrillation	7	1.33
Dual diagnosis	12	2.29
WPW + atrial flutter	3	
WPW + AVNRT	3	
WPW + AET	1	
WPW + atrial fibrillation	1	
AET+ atrial fibrillation	1	
AVRT + atrial flutter	1	

General Cardiology

PP-177
Cardiac biomarkers comparison between acute myocarditis/ myopericarditis and multisystem inflammatory syndrome in children

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Background and Aim: Acute myocarditis/myopericarditis is a heterogeneous disorder of unknown origin, the viral etiology leading the first row. There could be also myocardial involvement in multisystem inflammatory syndrome in children (MIS-C). In this study, we aimed to investigate cardiac biomarkers of acute myocarditis/myopericarditis and MIS-C and to compare these parameters between the two diseases.
Method: Patients who are diagnosed with MIS-C, isolated viral myocarditis/myopericarditis at a university hospital from October 2021 to March 2023 are included in this study.
Results: There were 38 MIS-C patients and 53 patients with myocarditis/myopericarditis. The mean age was 141.2 ± 38.2 months (4 to 18 years old) in MISC, and 145.8 ± 52.1 months (7 to 18 years old) in myocarditis/myopericarditis. Median troponin I level was 145 ng/L in MIS-C patients and it was 901 ng/L in myocarditis/myopericarditis patients. Creatinine kinase-myocardial band (CK-MB) median was 2.25 ng/mL (0.6-6.3) versus 6.7 ng/mL in MIS-C and myocarditis/myopericarditis, respectively. Pro Brain natriuretic peptide (Pro-BNP) median level was 2714.5 pg/mL (<300) in MIS-C, and it was 294 in patients with myocarditis/myopericarditis. Troponin I, CK-MB was significantly higher in myocarditis/myopericarditis, while Pro-BNP was significantly higher in MIS-C patients ($p < 0.05$). The separating power of CK-MB, troponin I, and Pro-BNP level was significantly higher in the differential diagnosis of these two group patients ($p < 0.001$). MIS-C patients with high pro-BNP levels had more prolonged hospitalization and left ventricular function impairment according to myocarditis/myopericarditis.
Conclusions: Cardiac biomarkers (CK-MB, troponin I, and Pro-BNP) could be good markers to estimate the course of the diseases.

Keywords: Cardiac biomarkers, myocarditis/myopericarditis, troponin, pro-BNP, multisystem inflammatory syndrome

Fetal Cardiology

PP-178
Congenital long QT syndrome and fetal bradycardia - a path to early detection and prevention

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Background and Aim: Fetal arrhythmias are uncommon diseases that occur in 2% of fetuses, with sinus bradycardia accounting for >10%. Congenital cardiac malformations, immune-mediated diseases and maternal infections can cause fetal bradycardia. In rare cases, fetal sinus bradycardia can be a marker of Congenital Long QT Syndrome. LQTS is an arrhythmogenic channelopathy characterized by delayed repolarization of the myocardium and increased risk for syncope and sudden cardiac death.
Method: We present the case of a newborn girl exhibiting sustained bradycardia, initially detected during the first trimester of gestation.
Results: At 29 weeks of gestation the mother was referred to the fetal cardiologist for sustained fetal bradycardia. The fetal echocardiogram showed normal function and structure of the heart. The AV interval was within normal limits, with 1:1 atrioventricular contractility, although the fetal HR ranged between 108-119 bpm. No familial history of cardiac disease or sudden death was reported. Maternal anti-Ro and anti-La antibodies were negative. Nevertheless, the maternal ECG showed a slightly prolonged QTc interval of 480 msec. Consequently, the suspicion of maternal LQTS led to genetic testing. The results identified a mutation in KCNQ1 gene, conclusively confirming the diagnosis of LQTS type 1. At term, a baby girl was delivered via cesarian

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section, receiving an Apgar score of 9/9. At NICU admission, clinical examination and laboratory investigations were unremarkable, except for bradycardia. Echocardiography revealed a small ASD and PDA. Standard electrocardiography revealed sinus bradycardia of 108 bpm and QTc interval of 537 msec. The Schwartz score was calculated at 4,5 points indicating high probability of LQTS. Consequently, treatment with Propranolol was started, gradually escalating the dosage up to 1,5 mg/kg/day. The beta-blocker therapy was well tolerated, maintaining HR >85 bpm and normal BP, while subsequent ECG assessments revealed the QTc interval between 510-516 msec. At discharge, the patient was in good condition and has not experienced arrhythmias to date.
Conclusions: Since LQTS account for 10% of Sudden-Infant-Death-Syndrome and effective therapy is available, early detection can prevent life-threatening events. When faced with fetal bradycardia, maintaining a high level of suspicion for LQTS and conducting familial screenings are crucial steps towards improving the survival rates of these patients.

Keywords: Fetal Sinus Bradycardia, Congenital Long QT Syndrome, KCNQ1 mutation

PP-179

Fetal diagnosis of an isolated aberrant right subclavian artery a normal variant?

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Background and Aim: An aberrant right subclavian artery (ARSA) is the most common aortic arch anomaly (1,5%). In prenatal scanning, the introduction of the three vessel and trachea view in routine fetal echocardiography has led to an increased detection of an ARSA in otherwise normal hearts. In some reports, the ARSA can already be seen as early as the first trimester examination. The presence of an ARSA has been reported to be an independent marker of trisomy 21. Clinically, the ARSA can become symptomatic due to the compression of adjacent structures. Dysphagia, respiratory distress and stridor have been reported. The presence of an ARSA leads to a referral to fetal cardiology for counseling postnatal symptoms. There is a lot of uncertainty if fetuses with ARSA should be born in tertiary cardiac centers. If a vaginal delivery is safe and if symptoms such as stridor due to tracheal compression or dysphagia occur early postnatally or later in childhood. Parents present with a lot of anxiety.
Method: The aim of this study was to describe the associations with chromosomal anomalies and the postnatal outcome of fetuses referred for cardiac assessment due to the suspicion of ARSA.
Results: A retrospective analysis was performed for fetuses referred for "isolated" ARSA from January 2020 until September 2023 in the fetal cardiology outpatient clinic of our hospital. 45 fetuses were included. All but one were confirmed at birth. One child was found to have only a bicarotid trunk. The ARSA was an isolated finding in 96%, while in 2 fetuses the ARSA was associated with a left superior caval vein and one fetus was found to have a VSD. One fetus was diagnosed with trisomy 21. There was no respiratory distress at birth or any signs of airway compression after birth. No child was referred for dysphagia.
Conclusions: The association with a chromosomal anomaly needs to be explained to the parents. This study helps to elude strategies to prevent unnecessary investigations for the fetus and anxiety for the parents, and identify children at risk who will benefit from intervention. A larger study is currently planned to get more data.

Keywords: ARSA,Fetal Echocardiography, Outcome

PP-180

Prenatal diagnosis and outcome of atrial isomerism: still a difficult counseling situation

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Background and Aim: Report our experiences in prenatal diagnosis of atrial isomerism (AI), describe the complex spectrum of cardiac and extracardiac associated anomalies and postnatal outcomes.
Method: Prospective and retrospective observational study was carried out on fetuses diagnosed of AI between March 2006- October 2023 in a single institution. Prenatal study was based on fetal echocardiographic evaluation supplemented by fetal MRI. An anatomopathological study was carried out in all interrupted pregnancies, except one case following family desire.
Results: 42 fetuses (50% females) were diagnosed; 11 (26%) had right atrial isomerism (RAI) and 31(74%) had left atrial isomerism (LAI). All fetuses with RAI had complex heart disease: atrioventriculo septal defect (AVSD) 82%, doble outlet of the right ventricle (DORV) 18%, pulmonary stenosis or atresia (PS) 54%, dextrocardia 45%, anomalous pulmonary venous drainage (APVD) 90%, persistent left superior vena cava (PLSVC) 73%, with no cases with heart rhythm disturbance or hydrops. Asplenia was found in all of them and other extracardiac anomalies were noted in 9 cases (81%), predominantly intestinal malrotation. All fetuses with LAI had interrupted inferior vena cava with azygous continuation. 7 (23%) have not heart disease or simple isolated one (VSD). AVSD was found in 48%, dextrocardia 26%, mesocardiac 9%, DORV 19 %, single atrium 19 %, APVD 32 %, PLSVC 42 %. 10 cases (32%) presented sustained bradyarrhythmia or complete block. Polysplenia was found in 42%, other extracardiac anomalies were diagnosed in 13 cases (42%), mostly intestinal malformations, highlighting brain malformation in a fetus with normal heart.
Termination of the pregnancy was carried out in 55% global (63% RAI- 52% LAI). There were 19 newborns, 57% of them required cardiac surgery (75% in RAI vs 53% in LAI) and two (5%) required pacemakers. After a median follow-up of 6,8 years the global survival was 73% (50% in RAI versus 80% LAI). 1 patient was lost to follow up.
Conclusions: Our study confirms the previous fetal findings, remarking that AI is one of the most complex forms of congenital heart disease except LAI with normal heart. Although our number of newborn babies is small, the postnatal outcomes for RAI is worse than those for LAI.

Keywords: Heterotaxia, Left isomerism, Right isomerism, Asplenia syndrome, Polysplenia syndrome.

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Table 1. Summary table of cardiac and extracardiac associated anomalies.

	RAI (n=11)	LAI (n=31)
Males	6/11 (54%)	15/31 (48%)
Females	5/11 (46%)	16/31 (52%)
Complex heart disease	11/11 (100%)	24/31 (77%)
Dextrocardia-mesocardiac	5/11(45%)	11/31 (35%)
AVSD	9/11 (82%)	15/31 (48%)
DORV	2/11 (18%)	6/31 (19%)
PA or PS	6/11 (55%)	5/31 (16%)
APVD	10/11 (90%)	10/31 (32%)
PLSVC	8/11 (73%)	13/31 (42%)
Single atrium	1/11 (9%)	6/31 (19%)
Tetralogy of Fallot	0%	5/31 (16%)
Interrupted inferior vena cave	3/11 (27%)	31/31 (100%)
Bradyarrhythmia or complete block	0%	10/31 (32%)
Asplenia	100%	0%
Polysplenia	0%	15/31 (48%)
Other extracardiac anomalies	9/11 (81%)	13/31 (42%)
Malrotation	4/11 (36%)	5/31(16%)
Brain anomalies	2/11 (18%)	2/31(6%)
Kidney anomalies	2/11 (18%)	1/31 (3%)
Other malformations	3/11 (27%)	7/31 (22%)

Table 1. Summary table of cardiac and extracardiac associated anomalies.

PP-182

A challenging case of refractory supraventricular tachycardia with hydrops fetalis in a twin pregnancy

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Background and Aim: The treatment regimen for fetal supraventricular tachycardia (SVT) lacks consensus, varying across institutions, and managing SVT in one fetus of a multiple gestation adds complexity, with few reported cases.
Method: Case Report
A 28-year-old secundigravida at 24 weeks' gestation presented with a dichorionic-diamniotic twin pregnancy conceived through in vitro fertilization. Ultrasound revealed persistent SVT (atrioventricular re-entry tachycardia) in twin 1, with a heart rate (HR) of 250 bpm, hydrops fetalis, and bilateral renal pelvic dilatation. Twin 2 was normal.
Results: After obtaining consent, the mother received oral Flecainide (300 mg/24 h) as first-line therapy. After 72 h, twin 1 showed persistent SVT with only short sinus rhythm periods, worsening mitral and tricuspid valves regurgitation, and cardiomegaly. Oral digoxin was initiated, switched to intravenous (IV) administration titrated up to 625 μg/day. After 23 days, twin 1 cardioverted stably to sinus rhythm, hydrops resolved, and HR stabilized at 115-120 bpm with prolonged PR interval (up to 180 msec). Twin 2 maintained normal sinus rhythm with prolonged PR interval (up to 150 msec). Flecainide continued, and digoxin switched to oral 0.125 μg/day thrice daily. Discharged after 30 days, the patient had bi-weekly appointments. No maternal side effects were reported, and both digoxin and flecainide levels were kept in the therapeutic range. At 36 weeks, spontaneous contractions led to a vaginal delivery. Twin 1 weighed 3030 g, twin 2 2120 g. One-month-old twin 1 displayed no arrhythmia but had microcephaly, dysmorphism, tremor, jitteriness, and bilateral pelvi-ureteric junction stenosis, with karyotyping revealing chromosome 4 deletion. Twin 2 had an uneventful neonatal period.
Conclusions: This case reinforces previous findings that transplacental antiarrhythmic treatment can successfully achieve cardioversion without harm to the healthy fetus or mother, even during prolonged treatment. Ongoing monitoring is crucial, as demonstrated by the successful delivery and postnatal care despite the observed complications in twin 1.

Keywords: Fetal Arrhythmia, Fetal Echocardiography, Transplacental antiarrhythmic treatment, Twin pregnancy

PP-183

Impact of arterial duct morphology on postnatal outcome in fetuses with isolated complete vascular rings

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Background and Aim: Objective: To share our experience in prenatal diagnosis and postnatal management of patients with isolated complete vascular ring (CVR) and ductus arteriosus (DA) anomalies. The association of isolated CVR with DA anomalies is rare and seldom reported.
Method: We examined all patients with prenatal diagnoses of isolated CVR from January 2010 to December 2022. Fetal heart examination included three-vessel and trachea (3VT) view and supraaortic branch view. We classified each detected CVR based on aortic arch (AA) type, DA position, and presence of an aberrant subclavian artery. Parents were offered karyotyping and prenatal testing for 22q11 microdeletion.
Results: Among 55 fetuses with CVR, 49 had a usual left DA, while 6 (10%) exhibited ductal anatomic variations. 4/6 had a right circumflex AA with mirror image branching and bilateral DAs of whom the right was dilated and the left was small and running from the proximal segment of the left subclavian artery to the left pulmonary artery (PA); confluent branch PAs were demonstrated in utero. After birth 2/4 underwent left ductal stenting and subsequent surgical connection to the main PA after absence of left PA branch was documented; 1/4 showed mild left PA branch stenosis, and CT scan revealed isolated left subclavian artery isolation, requiring longitudinal follow-up for risk of 'subclavian steal'; 1/4 didn't show any echocardiographic or clinical issue. 2/6 fetuses displayed left cervical and circumflex AA, aberrant right subclavian artery, and dilated right DA. In one of them main PA extended directly to right PA branch whereas left PA branch originates from an additional left DA and passes down into the ipsilateral lung parenchym. After birth surgical connection of left PA to main PA was performed at 6 months of age. The other one received

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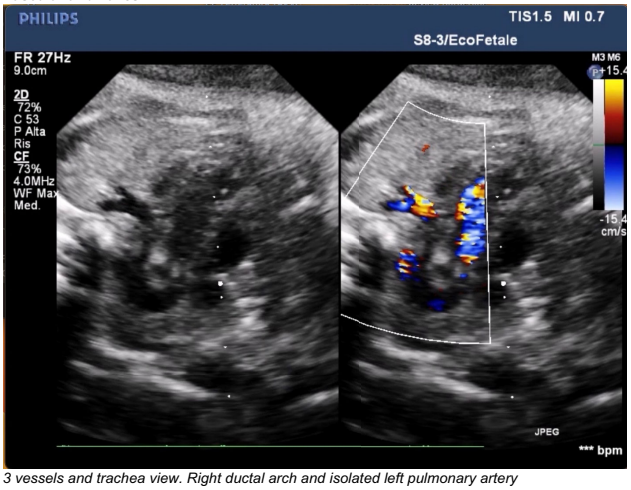
Vascular anomalies



3 vessels and trachea view. Right ductal arch and isolated left pulmonary artery

surgical right DA division and uncrossing arch procedure at 3 years of age. Both of them were positive for 22q11 microdeletion.
Conclusions: Defining DA characteristics during prenatal CVR diagnosis is crucial. Prenatal detection can prevent complications related to tracheal and esophageal compression and avoid late diagnoses of unilateral PA absence and other vascular complications, altering the natural history for affected children.

Keywords: Complete Vascular Ring, Fetal echocardiography, Arterial Duct, Absent pulmonary artery, Vascular Anomalies

PP-184

Outcomes of congenital heart diseases: Retrospective study since 2021 about 32 newborns

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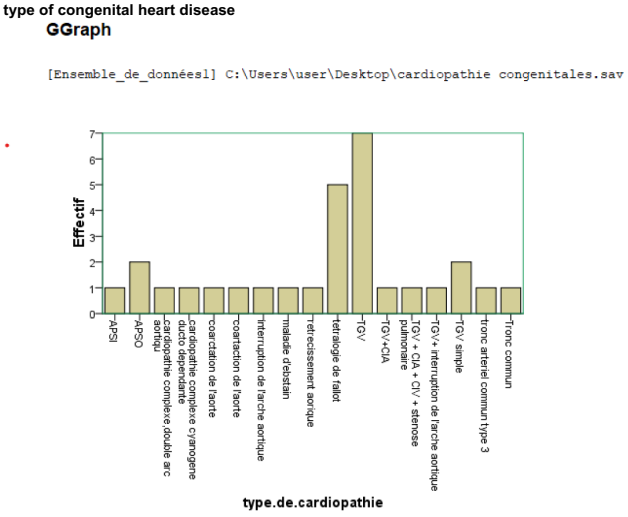
Background and Aim: Congenital Heart Disease (CHD) remains among the most common congenital malformation. Progress is made in terms of screening, antenatal diagnosis and treatment. This study aims to describe the clinical aspects and to determine the mortality rate of CHD in the Department of Intensive Care and Neonatal Medicine of Monastir.
Method: We underwent a retrospective study in the Department of Intensive Care and Neonatal Medicine of Monastir. During two years and ten months (January 2021- October 2023), we included all newborns diagnosed with CHD. We excluded non operable complicated CHD, persistent ductus arteriosus, atrial and interventricular septal defects.
Results: In total, 72 newborns with CHD were collected. After excluding 33 cases of atrial and interventricular septal defects, 7 cases of persistent patent ductus arteriosus and 3 cases of non-operable CHD, we enrolled 29 patients. The sex ratio was 1,5. Pregnancies were complicated by gestational diabetes in 7 cases and pre-eclampsia in 2 cases. Only one case had a family history of CHD. Antenatal diagnosis was made only in 24% of patients. In 75% of cases without antenatal diagnosis, CHD was revealed at the first 72 hours of life. The most frequent clinical symptoms were cyanosis (59%), heart murmurs (27%) and neonatal respiratory distress (14%). Transposition of the great vessels was the most common type of CHD (41%). Genetic study was carried out in 21% of cases. Ventilation was required in the majority of patients (62%). Vasoactive drugs were necessary for 8 cases. Half of these infants needed the use of prostaglandin E1. Atrial septostomy was performed for 9 cases. However, only 10/29 patients underwent surgery. The delay from diagnosis to intervention was long with an average time of 18 days. Mortality rate was 34%.
Conclusions: CHD have very wide spectrum of manifestations. Thus, more efforts are needed to improve antenatal diagnoses and organize neonatal management. This study underlines the difficulties with surgical management in developing countries.

Keywords: Heart defects, congenital, mortality,diagnosis

type of congenital heart disease



Cardiac Imaging

PP-185

Evaluation of right ventricle function using speckle-tracking echocardiography in beta thalassemia major patients

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Background and Aim: Cardiomyopathy mediated by iron disposition is a major cause of morbidity and mortality in patients with beta thalassemia major (BTM). Generally right ventricle (RV) injury is subtle. Therefore, cardiac imaging modalities based on myocardial deformation such as strain imaging are used for early detection of cardiac iron overload. To investigate whether longitudinal strain based on speckle tracking can detect subtle right ventricular iron overload in children with BTM.
Method: Sixty children (30 children with BTM and 30 healthy controls) were enrolled in this study. Conventional TTE study was performed in both patient and control groups. Conventional echocardiography parameters and RV strain were determined and compared between the two groups.
Results: Mean age was 10.4 ± 5 years in BTM group compared to 10.2 ± 5 years in control group (p=0.876). Compared to control

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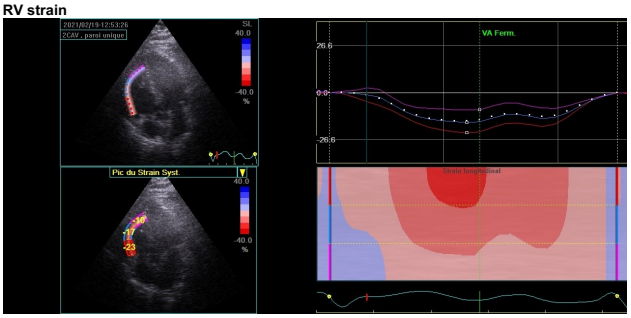
group, there was no significant difference in conventional TTE parameters except for indexed right atrium (RA) area and volume. RV strain was significantly lower in BTM children. RA area (cm^2) 10,38 ± 2,87 9,83 ± 2,61 0.44 Indexed RA area (cm^2/m^2) 10,18 ±2,93 9,08 ± 2,27 0,11 RA vlume (ml) 24,00 ± 9,07 20,53± 7,75 0,12 Indexed RV volume (ml/m^2) 23,16 ±6,69 18,39 ± 4,75 0,003 Tricuspid valve maximum velocity (m/s) 2,31± 0,40 2,19 ± 0,20 0,15 TAPSE (mm) 23,97 ± 5,22 24,33± 5,00 0,78 S'(cm/s) 14,97 ± 2,93 16,03± 3,01 0,17 RV GLS (mean ±SD) -24.6±4.5 -18.2±3.4 0.01.
Conclusions: In asymptomatic BMT children with normal RAconventional TTE RA parameters, longitudinal strain could be used for the detection of subclinical myocardial dysfunction.

Keywords: Beta thalassemia, strain, right ventricle



Paediatric Cardiovascular Intensive Care

PP-186
Prostaglandin E1 utilization for congenital heart disease in neonatal intensive care: A retrospective study

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Background and Aim: Prostaglandin E1 (PGE) is a vital treatment for neonates with ductal-dependent congenital heart disease (CHD) waiting for surgery. In addition to its cost, it can have many side effects. This study aims to determine the safe use of PGE and to evaluate the outcomes of those infants.
Method: Via a retrospective study during ten years (January 2013-October 2023), conducted in the department of Intensive Care and Neonatal Medicine, we collected all newborns diagnosed with curable congenital heart disease, who received PGE.
Results: A total of 63 cases were included. A male predominance was noted with sex ratio2.1. The median age of diagnosis was 2 days [1-17]. Thanks to antenatal diagnosis in only 33% of cases. The most common CHD included transposition of great vessels (24%), pulmonary stenosis or atresia (24%), and aortic coarctation (19%). The PGE use spanned a median of 4 days [1-29]. Adverse effects were: hyperthermia (34%), hypotension and rhythm disturbances (11%), apnea (10%), hypocalcemia (7%) and myalgia (20%). These effects appeared on average 5 hours after initiation [7-12h]. In 92% of cases, no severe respiratory deterioration needing mechanical ventilation has been observed. Duration of hospital stay was in the median of 10 days [1-49d]. Among these patients, 54 % were operated within an average time of 5 days. The overall mortality rate was 51%. One fifth of deaths occurred preoperatively.
Conclusions: This study underlines the difficulties of managing CHD in developing countries, revealing the high rate of mortality. Besides its potential side effects, PGE continues to be indispensable. This leads us to elaborate, in our department, protocols to improve efficacy and safety of PGE's use and with Cardio-vascular surgery team to reduce operating waiting time.

Keywords: Alprostadil, Heart Defects, Congenital, adverse effects, Mortality

Fetal Cardiology

PP-187
Prenatally diagnosed congenital heart block – a single center experience

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Background and Aim: Congenital heart block (CHB) is a life treatening condition for the fetus, especially when associated with a complex congenital heart defect (CCHD). We aimed to analyze the group of fetuses diagnosed with CHB for their initial characteristics, pregnancy course and perinatal outcome
Method: We reviewed data of fetuses with either isolated CHB (I-CHB) or CHB associated with CCHD (A-CHB) diagnosed in our Center between 2003-2023. In both groups we analyzed fetal heart rate (FHR), heart size and Cardiovascular Profile Score (CVPS) at the diagnosis and examined changes in peripheral flows (umbilical vessels, ductus venosus and middle cerebral artery) throughout pregnancy. Then we examined associations between forementioned parameters and pregnancy outcome.
Results: There were 125 fetuses diagnosed with CHB, 77 with I-CHB and, 48 with A-CHB. Among 59 mothers of I-CHB fetuses tested for Ro/La antibodies, 53 (90%) were positive. A-CHB occurred mostly with heterotaxy syndrome (39 fetuses) or congenitally corrected transposition of great arteries (3). The average gestational age and FHR at diagnosis were similar in both groups: 23 weeks in I-CHB vs. 21 weeks in A-CHB and FHR of 61/min and 59/min, respectively. Fetuses with A-CHB had larger hearts (Ha/Ca 0.42 vs. 0.39, $p<0.05$) and were in more severe condition (median CVPS 5 vs. 8, $p<0.05$). Heart size correlated inveresely with FHR ($r=0,32$, $p=0,005$) in I-CHB. There were 5 (6.5%) intrauterine deaths or terminations in I-CHB and 24 (50%) in A-CHB. 12/16 live-born patients with A-CHB died in the neonatal period. Peripheral flows in both groups showed typical changes: near-normal MCA-PI, increased UA-PI and often abnormal venous flow (90% negative a-wave in DV, 45% UV pulsations). In I-CHB, lower CVPS, lower FHR and UV pulsation correlated with fetal demise.
Conclusions: Prenatal and perinatal outcome of A-CHB is much worse than of I-CHB, in spite of similar heart rate and timing of diagnosis. Peripheral flows show typical changes which should not be interpreted as brain-sparing effect. Signs of heart failure

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expressed in CVPS scale may be useful to predict the outcome of I-CHB fetuses.

Keywords: Congenital heart block, fetal diagnosis, fetal condition, survival rates

PP-188

Long term survival of antenatally diagnosed functionally single ventricle congenital heart disease

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Background and Aim: Antenatal diagnosis of single ventricle congenital heart disease (SV CHD) allows prenatal counselling, better parental preparation and support as well as improved perinatal management. Expectant parents ask about likely outcomes and expected longer term survival. We reviewed the survival of this cohort presenting at our institutions between January 2000-October 2023.

Method: Retrospective data was collected from fetal cardiac and paediatric cardiology databases. Maternal data, fetal cardiac diagnosis, postnatal cardiac diagnosis and management were collected. The outcomes were separated into 2 groups – Cohort 1 (CO1) [2000 to 2007] and Cohort 2 (CO2) [2008 to 2023].

Results: 523 fetuses were diagnosed with SV CHD in the 23 year 10 months period (see Table 1, Figs 1, 2). For the small LV group with active management, survival at over 10 years was 46% for CO1 group and 64% for CO2 group, with no significant difference in the survival between the cohorts (log rank p=0.12). For the small RV group with active management, survival over 10 years was 71% for CO1 group and 91% for CO2 group. There was significant difference in the survival between the cohorts (log rank p=0.03). No analysis was made for the unspecified hypoplasia as the numbers were too small.

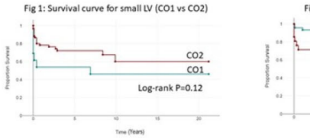
Conclusions: For both the small LV and small RV groups, the termination rate and compassionate care option remain unchanged between both cohorts. Overall, there has been improvement in the survival for the small LV and RV groups with active management, likely due to improved perinatal management with detailed management plan and designated delivery at tertiary neonatal hospital close to cardiac surgical centre, improved surgical expertise, improved PICU management as well as availability of ECMO for the sicker patients. In the small LV group, despite improvement in survival, this is not yet statistically significant due to more complex patients with more co-morbidity being accepted for surgery. For the small RV group, we see statistically significant improvement with survival reaching 90% at 10 years, due to increased use of non-surgical intervention like ductal stenting over conventional BT shunts which carry higher morbidity and mortality risk. Our survival outcomes are comparable with worldwide published data.

Keywords: Antenatal, Single ventricle, survival, outcome

Table and Survival Curve

Table 1: Demographics CO1 and CO2 details

	Cohort 1 (CO1) – 2000 to 2007	Cohort 2 (CO2) – 2008 to 2023	P value
Number of fetuses	124 84 (small LV), 38 (small RV), 4 (unspecified ventricular hypoplasia)	399 285 (small LV), 125 (small RV), 9 (unspecified ventricular hypoplasia)	
Small LV	84 fetuses	285 fetuses	
Termination of pregnancy/intrauterine demise	46/84	138/285	NS
Compassionate care (alive: dead)	11/84 (all died)	53/285 (3 alive,50 died)	NS
Active management (alive: dead)	13/84 (6 alive, 7 died)	69/285 (44 alive, 25 died)	
Loss to follow-up	2	5	
Survival for active management group	46%	64%	Log rank p=0.12
Small RV	38 fetuses	125 fetuses	
Termination of pregnancy/intrauterine demise	14/38	54/125	
Compassionate care (alive: dead)	3/38 (all died)	20/125 (all died)	
Active management (alive: dead)	21/38 (15 alive, 6 died)	49/125 (45 alive, 4 died)	
Loss to follow-up	0	2	
Survival for active management group	71%	91%	Log rank p=0.03
Unspecified ventricular hypoplasia	4 fetuses	9 fetuses	
Termination of pregnancy/intrauterine demise	2/4	7/9	
Compassionate care (alive: dead)	1,/4 (all died)	1/9 (all died)	
Active management (alive:dead)	1,/4 (all died)	1/9 (all died)	



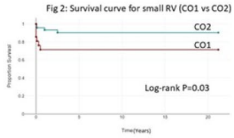


Table 1: Demographic data for CO1 and CO2 Fig 1: Survival curve for small LV Fig 2: Survival curve for small RV

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Pulmonary hypertension, heart failure and transplantation

PP-189

Novel, off-label use of the cardiomemstm remote pulmonary arterial pressure monitoring device in management of idiopathic pulmonary hypertension

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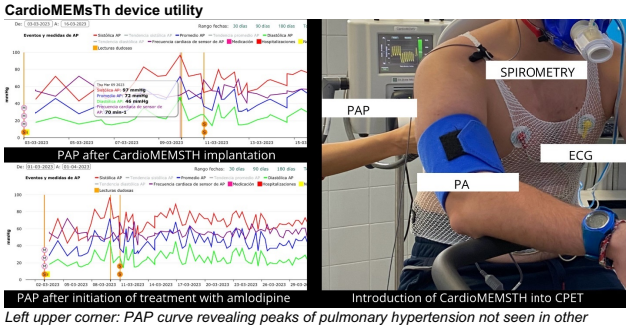
Background and Aim: Management of pulmonary arterial hypertension (PAH) precises close monitoring of pulmonary artery pressure (PAP). However, the risk associated with catheterization, precludes frequent monitoring. The CardioMEMSTM HF System is remote PAP monitoring device, approved by FDA for adult patients with heart failure.
Method: Single patient case study.
Results: We present a male patient who underwent annual echocardiographic controls since birth due to the family history of a brother who died at 15 months of age because of PAH, which resulted normal until the age of 10, when he started presenting presyncopes and disnea and echocardiography revealed signs of mild PAH, later confirmed by cardiac catheterization (PAP 43/7/22mmHg, PVR 3,39U/m2, negative vasoreactivity test). The guided interview revealed that 18 months prior the patient started treatment with therapeutic dosis of metilphenidate. Laboratory analyses for PAH workup including genetic tests resulted normal. The treatment included bosentan, sildenafil and suspension of metilphenidate, with complete remission of symptoms until the age of 16 years when he started presenting syncopes. Catheterization (PAP 49/9/29mmHg), CPET, cardiac MR and echocardiography did not reveal relevant changes with respect to previous studies. The symptoms persisted regardless of increase of bosentan and sildenafil doses and adding Selexipag. Due to high clinical suspicion of PHA as an etiology of his symptoms, CardioMEMSTM device was implanted and permitted continuous, real-time monitoring of PAP that demonstrated high daily variability with peaks up to 92/72/46mmHg, that had never been previously detected by other tests. Amlodipine was added to the therapy after which, lower variability and decreased PAP peaks (61/38/18 mmHg) were observed with remission of the symptoms referred by the patient. CardioMEMSTM was introduced into CPET revealing PAP of 46 mmHg during peak excercise.
Currently at 17 years of age, the patient remains asymptomatic with elevated PAP therefore the treatment will be further optimized.



Left upper corner: PAP curve revealing peaks of pulmonary hypertension not seen in other complementary tests previous to the date Left lower corner: Real-time observation of changes in PAP after change of the treatment Right part of the picture: Introduction of invasive PAP measurements into CPET

Conclusions: CardioMEMSTM is a safe, reliable and useful tool for the management of PAH, facilitating close monitoring of patients, enabling medication titration and anticipating the onset of symptoms. It´s introduction in to CPET allows measurement of excercise invasive hemodinamics, otherwise impossible to obtain due to necessity of sedation during catheterization in pediatric patients.

Keywords: CardioMemsTM, pulmonary hypertension, PAP

Fetal Cardiology

PP-190

Thrombus of isolated congenital ductus arteriosus aneurysm in an asymptomatic neonate

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Background and Aim: Ductus arteriosus aneurysm (DAA) is characterized by a saccular or fusiform dilatation of the ductus arteriosus. Although exact pathogenesis remains unclear; it seems to be related to turbulent flow or endothelial injury in the narrowed segment of the ductus. In most cases is an incidental finding during the third trimester, related to a good fetal and neonatal outcome. It's potential complication includes thrombosis that may lead to obstruction of fetal circulation and hemodynamic decompensation with hydrops in fetuses and mass effect with compression of adjacent structures and thromboembolism in infants. DAA risk stratification, management of fetuses and neonates with DAA thrombosis is controversial and there is no clear consensus regarding surgical treatment or indications for anticoagulation and antiplatelet therapy.
Method: Retrospective single case report.
Results: A 31-year-old woman, (gravida 1, para 0) was referred fetal cardiology department at 37 weeks' gestation due suspicion of coarctation of the aorta and echocardiography showed an enlarged dilated structure, 14 mm in diameter, that protruded leftward of the aortic arch with turbulent, swirling flow and no restrictive aspect of the Doppler ductal flow at that time (Figure 1). No other fetal structural anomalies were observed. The patient did not attend the planned follow up during the last weeks of pregnancy. A female infant was born at 40 weeks' gestation weighted 3170 gr, with an 10/10/10 Apgar score. Physical examination was normal. Echocardiography revealed PDA with left-to-right shunt, DAA, non-obstructive thrombus in PDA (arrow) with right ventricular hypertrophy (RVH) (Figure 2). A conservative approach was adopted with clinical and echocardiographic surveillance with good outcome and spontaneous ductus closure at 3 days and progressive remission of RVH.
Conclusions: Although the possibility of thrombi formation over the last weeks of pregnancy cannot be confirmed, it could explain the presence of RVH secondary to the blood flow obstruction across the arterial duct. Considering the possibility of serious complications related to thrombi of DAA, recognition and surveillance of DAA size and monitoring of thrombus development in pregnancy and neonatal period by serial echocardiographic examinations are critical in follow up.

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Keywords: Ductus arteriosus aneurysm, fetal echocardiography, trombosis

Echocardiography

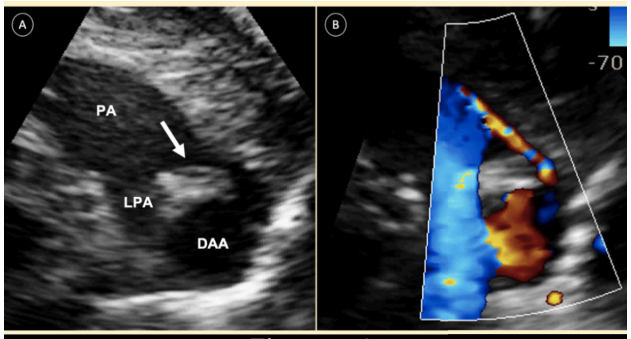

Figure 1.

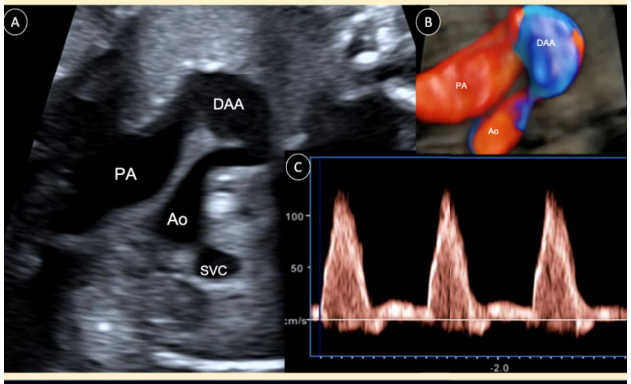

Figure 2.

Figure 1: Ductus arteriosus aneurysm during fetal life. (A) Two-dimensional echocardiogram demonstrates a ductus arteriosus aneurysm. (B) 4-D STIC ultrasound verifies the ductus shape and contour. (C) Pulsed Doppler in the ductus arteriosus showing a normal Doppler flow at 37 weeks. Ao, aorta; PA, pulmonary artery; SVC, superior vena cava; DAA, ductus arteriosus aneurysm Figure 2: Ductus arteriosus aneurysm in the neonatal period. Parasternal short-axis view. (A) Two-dimensional echocardiogram demonstrates a small thrombus (arrow) in the narrowed pulmonary ductus segment, on the left of the left pulmonary artery. (B) Color-flow Doppler. PA, pulmonary artery; LPA, left pulmonary artery; DAA, ductus arteriosus aneurysm.

PP-191

A rare case in fetal cardiology – congenitally corrected transposition of the great arteries with interrupted aortic ARCH

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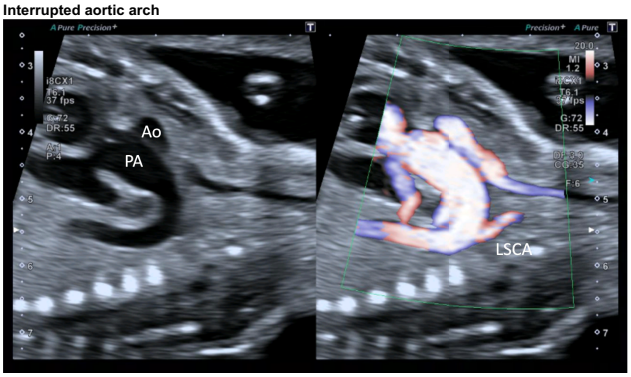
Bristol Royal Hospital for Children

Background and Aim: Congenitally corrected transposition of the great arteries (ccTGA) is a rare congenital cardiac abnormality, which is often associated with other cardiac lesions. We would like to share an unusual case, who was seen in our fetal cardiology clinic.
Method: We reviewed the outpatient data and fetal echocardiograms.
Results: A 27 year old woman was referred to the fetal cardiology clinic, because the fetal anomaly scan showed an abnormal 3 vessel view, and she was referred with a suspicion of transposition of the great arteries. She was first seen at 21 weeks + 6 days gestational age. She was fit and healthy and not on any medications. There was some family history of congenital heart disease, but not in any first degree relatives.
The fetal echocardiogram showed abdominal situs solitus, atrial situs solitus, levocardia, AV and VA discordance (ccTGA), large VSD. The pulmonary artery was overriding the VSD, the aorta was arising from the left sided anatomical RV. The aorta was anterior and leftwards compared to the pulmonary artery. The aorta was smaller than the pulmonary artery but within normal Z scores and there was no aortic valve stenosis. There was an interrupted aortic arch with interruption between left carotid artery and left subclavian artery. There were no signs of heart block, the fetal heart rate was 144bpm and there was 1:1 AV conduction.
There was no extracardiac abnormality and they declined invasive genetic testing.
Conclusions: There are only three case reports in the literature which describe an association of ccTGA and interrupted aortic arch and all of these patients were diagnosed postnatally. This is the first case report best to our knowledge which describes an antenatal diagnosis of ccTGA, and interrupted aortic arch.

Keywords: ccTGA, interrupted aortic arch, fetal cardiology

Interrupted aortic arch



Parallel great arteries, interrupted aortic arch type B

PP-192

Incremental value of 2d strain fetal echocardiography in the prediction of neonatal valvuloplasty in isolated MILD to critical pulmonary valve stenosi

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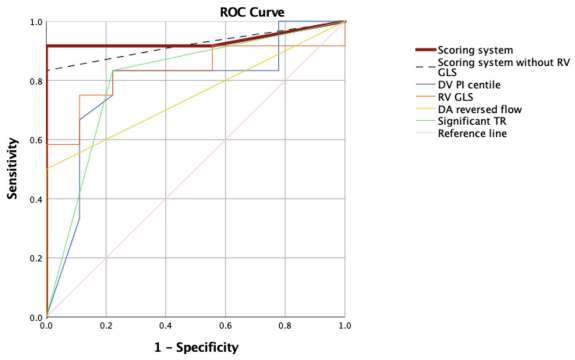
Background and Aim: To evaluate a prediction model of neonatal valvuloplasty by fetal echocardiography including speckle tracking echocardiography (STE) in a series of isolated pulmonary stenosis (PS).
Method: A prospective cohort study was conducted, which included 24 cases of isolated PS, comprising 7 critical and 17 mild-moderate cases. Cases were matched with 48 healthy controls adjusted for gestational age at echocardiography. The study was conducted at BCNatal, a referral center for congenital heart defects in Spain. Fetal ultrasound, comprehensive echocardiography and off-line STE were performed at the time of PS diagnosis. Echocardiographic parameters were analyzed according to the need for neonatal valvuloplasty to identify possible predictors of early intervention.
Results: Maternal and perinatal characteristics were similar in both groups. Mean gestational age at echocardiography was 31.4 weeks. Half of the PS fetuses required valvuloplasty within the first month of life.
Critical PS fetuses showed global RV functional impairment with significantly reduced GLS, FAC, TAPSE, a more pulsatile DV, and no changes in LV parameters. Worse RV function was observed in the valvuloplasty group. A combination of DV PI centile (cut-off value ≥79%), RV GLS (cut-off value of ≥-15.85%), reversed flow in the DA, and significant tricuspid regurgitation were predictors of neonatal valvuloplasty, and when combined in a scoring system the area under the curve was 0.931, p=0.001. Giving a score of 1 for each present variable, values > or equal to 2 predicted the need of neonatal valvuloplasty with a sensitivity of 91.7% and specificity of 100% (Figure 1). The prediction model based only on conventional echocardiographic parameters also had excellent performance with slightly lower sensitivity.

Figure 1



Receiver-operating characteristic (ROC) curves of Scoring System (red thick line) area under the curve (AUC) 0.931, scoring System without right ventricle global longitudinal strain (RV GLS) (dashed line) AUC 0.917, ductus venosus pulsatility index cenile (DV PI) (blue line) AUC: 0.782, RV GLS (Orange line) AUC 0.833, reversed flow at the ductus arteriosus (DA) (yellow line) AUC 0.750 and significant tricuspid regurgitation (TR) (green line) AUC 0.806; as predictors for neonatal valvuloplasty.

Conclusions: Decreased RV deformation is observed in critical PS fetuses, and correlates with mechanical dyssynchrony. A scoring system combining the DV PI centile and RV GLS with the presence of reversed ductus arteriosus flow and significant tricuspid regurgitation, is provided to identify those fetuses in need of neonatal valvuloplasty, who will thus benefit from an early neonatal assessment by pediatric cardiologists. Further studies are needed to evaluate the performance of the suggested scoring system, which seems promising.

Keywords: Fetal echocardiography, Pulmonary stenosis, Speckle tracking echocardiography.

PP-193
Early fetal ECHO – accuracy and incremental benefit of endovaginal imaging and early reassessment in cases with congenital heart disease

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Fetal and Neonatal Cardiology Program, Division of Pediatric Cardiology, Department of Pediatrics, University of Alberta, Edmonton, Canada

Background and Aim: The anatomical accuracy of early (EFE) compared with mid-trimester fetal echo and incremental value of additional endovaginal (EV) imaging and early reassessment in fetal cardiac diagnosis (FCD) is poorly quantified. We sought to assess these factors in our high volume EFE program.
Method: We identified all EFE performed from 10+0-15+6 weeks of gestation from 2009-21 with FCD. We developed a maximum 9-point scoring tool to quantify anatomical information obtained based on a segmental morphological approach, including at first EFE, at follow-up and on EV imaging for cases with FCD. We compared scores for initial EFE vs repeat EFE, initial EFE vs fetal echo at ≥ 18 weeks gestation, and score change when additional EV imaging was taken at EFE.
Results: 1660 EFE were performed in 1387 pregnancies; with FCD diagnosed in 130. In 14/130 with FCD EV imaging was required at EFE - gestational age was earlier (mean 12.1 vs 13.4w, p<0.0001) with mean score increasing from 1.6 for transabdominal studies to 3.5 with EV studies (p=0.049). 35/130 had repeat EFE (< 18 weeks gestation) of which we reviewed 17 patients with mid-trimester follow up data available. Repeat EFE studies had initial EFE at a mean of 13.2 weeks with mean score of 5.1, follow up 2 weeks (range 0.3-4.2) later showed a score increase to 7.4 (p < 0.0001). We reviewed a further 16 patients with mid-trimester follow up (≥ 18 weeks). In the total group, early studies were done at a mean of 14.4 weeks with a mean score of 5.6 vs 8.2 for the mid-trimester studies (p< 0.0001). Of the EFE FCD diagnoses 27 had no, 7 had minor and 1 had a major change at mid trimester follow up.
Conclusions: We have shown that when EFE is abnormal, results can be used for accurate counselling. When required, follow up EFE and EV imaging were highly effective in obtaining further diagnostic information. Fetal echo at ≥ 18 weeks gestation is particularly useful in demonstrating the pulmonary venous anatomy and branch pulmonary arteries, although it is uncommon for major diagnostic changes to occur.

Keywords: fetal, echocardiography, accuracy, transvaginal, first trimester

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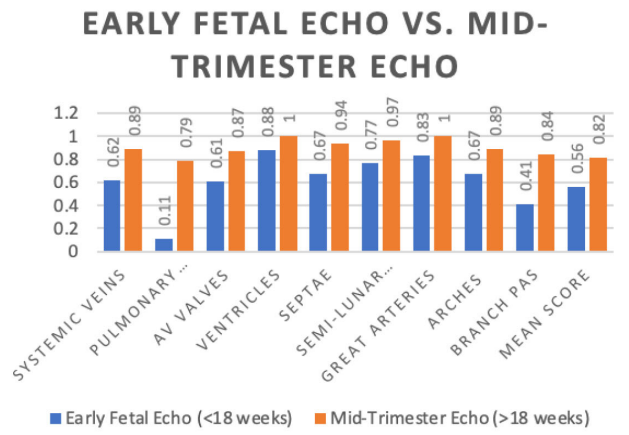
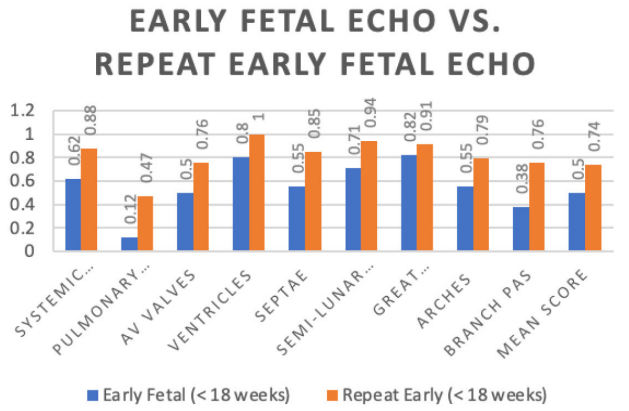
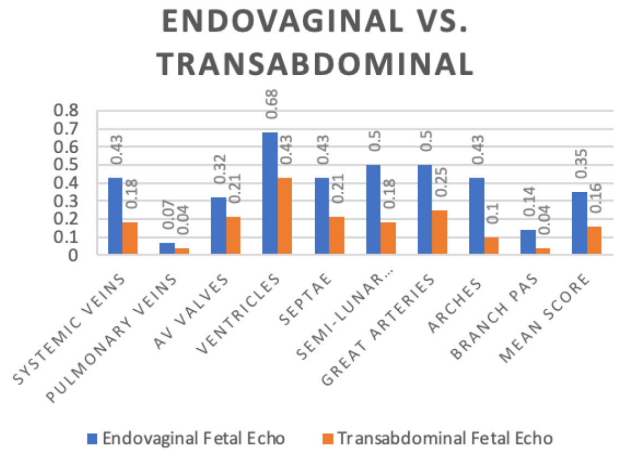
Additional information obtained with further imaging







General Cardiology

PP-194

Exploring risk factors for coronary complications in kawasaki disease in non-asian population

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Department of Paediatric Cardiology and Congenital Heart Defects, Medical University of Gdansk, Poland

Background and Aim: Kawasaki disease, known for its serious complication of coronary artery aneurysms, poses challenges in assessing the risk of coronary changes in non-Asian populations. This study, conducted in the Pomeranian region in Poland with 80 Kawasaki disease patients diagnosed between 1997 and 2021, aimed to develop a simplified risk assessment model for coronary artery aneurysms.
Method: The retrospective analysis of the acute phase included fever duration, symptoms, and abnormalities observed in transthoracic echocardiography (TTE) and electrocardiography (ECG). A comparative analysis was performed between patients with a normal coronary artery appearance during the acute phase of the disease or those with transient changes and patients showing coronary artery changes after the resolution of the acute phase of the disease.
Results: Among 80 patients, coronary changes were noted in 17 (21.3%), with a significant factor being diagnosis after the 10th day of fever. Multifactorial analysis identified independent risk factors, including late diagnosis, age below 6 months, repolarization disorders (ST-T abnormalities), and at least 4 additional symptoms (not constituting diagnostic criteria for the disease). The average age at onset was 2.3±2.2 years, and the short-term observation duration was 32.0±9.8 days. The area under the curve (ROC) was 0.934, indicating the model's reliability. The prognostic model categorized the risk of coronary changes into Grades I (1.9%), II (45%), and III (87.5%).
Conclusions: In conclusion, the developed risk assessment model proves valuable in identifying patients at a higher risk of coronary artery aneurysms. Emphasizing the significance of early Kawasaki disease diagnosis, particularly concerning aneurysm risk, the model serves as a practical clinical tool. Striking a balance between feasibility and accuracy, it helps identify patients requiring specialized attention and monitoring for potential coronary complications.

Keywords: Kawasaki disease, coronary aneurisms

PP-195

Everolimus as effective complementary treatment in refractory atrioventricular reentry tachycardia in a patient with tuberous sclerosis complex

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Heidelberg (KiTZ), Heidelberg, Germany. National Center for Tumor Diseases (NCT), NCT Heidelberg, partnership between German Cancer Research Center (DKFZ) and Heidelberg University Hospital, Germany. Department of Pediatric Oncology, Hematology and Immunology, Heidelberg University Hospital, Heidelberg, Germany. Division of Pediatric Glioma Research, German Cancer Research Center (DKFZ), Heidelberg, Germany; [7]Department of Pediatric and Congenital Cardiology, Heidelberg University Hospital, Heidelberg, Germany

Background and Aim: Tuberous sclerosis complex (TSC) is a rare autosomal-dominantly inherited multisystem disorder which is associated with the occurrence of cardiac rhabdomyomas - benign tumors consisting of altered and dysfunctional cardiomyocytes - that also can cause arrhythmias that are unresponsive to standard antiarrhythmic therapy. Pharmacological mTOR (mechanistic target of rapamycin) inhibition with everolimus may be considered as complementary therapy with antiarrhythmic effects, but data is still limited.
Method: We report the case of a preterm infant (33 weeks of gestation) with multiple cardiac rhabdomyomas who developed refractory atrioventricular reentry tachycardia (AVRT). Everolimus was initiated as add-on therapy at the age of 2 months. The follow-up time was 7 years.
Results: Pharmacological mTOR inhibition with everolimus rapidly improved arrhythmia within few weeks after treatment initiation as additional therapy to antiarrhythmics such as beta-blockers, flecainide, and amiodarone, which however were ineffective as sole treatment in controlling arrhythmia. Further, we observed a correlation between arrythmia control and reduction of rhabdomyoma size. Attempts to discontinue everolimus resulted in rhabdomyoma size rebound and recurrence of arrhythmic episodes (AVRT). No relevant side-effects of the long-term use of everolimus were noted. Finally, at the age of 6 years, electrophysiologic testing confirmed an accessory pathway that was successfully ablated.
Conclusions: In summary, we present data on long-term medication with everolimus in a child with TSC-associated AVRT. This case report provides important information on the safety and efficacy of an mTOR inhibitor for the treatment of a potentially life-threatening cardiac disease manifestation in TSC for which the optimal treatment strategy is still not well established.

Keywords: Tuberous Sclerosis, Rhabdomyoma, AVRT, Everolimus

PP-196

Early detection of accelerated atherosclerosis, in a cohort of survivors suffered from childhood malignancies

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Background and Aim, Introduction: Adverse effects the cardiovascular system secondary to oncology treatment is a significant fact. The increasing rates of pediatric survivors, and the invention of new drugs of which late effects as still unknown provoke medical interest. New indexes, easy to use in clinical work are vital for early detection of cardiotoxicity. Echocardiography has a key role in detecting cardiotoxicity induced cardiomyopathy, similar indexes have to be established for detecting accelerated atherosclerotic cardiovascular disease (ACVD).
AIM: To examine if combining calculated epicardial adipose tissue (EAT), diastolic disfunction (DD) of LV and measurements of intima media thickness of the internal carotid artery can serve as early markers of ACVD
Method: 105 survivors from childhood malignancies were examined. Males: 58, Females:47. Age range: 9-36 years, mean age 17.7 years. Divided in 3 age groups: Children: 24, Adolescents: 47, adults 34. They initially suffered from Leukaemia's (45), CNS tumours: (21), Lymphomas (16), Neuroblastomas (11), Miscellaneous (12). Time elapsed from completion of treatment: 5.5 to 25 years. Mean time of completion treatment: 8 years and 6 months. Firstly, biochemical risk factors (RF) for atherosclerosis, as well as clinical indexes as BP and waist to hip ration were measured. A second team, blinded to the first obtained: 1. EAT, 2. DD of LV by Echo-2D and TDI techniques, 3. calculated Intima Media Thickness (IMT) of the internal carotid artery by high frequency resolution ultrasonography, following AEPC guidelines. Correlated a cluster of seven or more RF's with pathological measurements of IMT, LV/DD and EAT, to calculate the incidence of ACVD in our cohort and determine conditions related to that.
Results: Pathological combination EAT+DD + IMT was found in 20% of our cohort. All of them had a cluster of seven or more RF's for ACVD. Age, length of completion of treatment, combined treatment (chemo and radiation), were contributing for ACVD.
Conclusions: Although a primary study, we show a trend of detecting early subclinical ACVD among survivors from childhood malignancies using clinical indexes deriving from first line imaging of the cardiovascular system.

Keywords: accelerated atherosclerosis, childhood malignancies

PP-197

The case of a teenager's broken heart- high voltage electric shock causing rupture of chordae tendinae, leading to acute mitral valve insufficiency

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Background and Aim: The mechanisms of cellular injury by electricity are: the conversion of electric energy into thermal energy causing burns, alteration of cell membrane resting potential and eliciting tetanic muscular contraction, mechanical injury through falls, and irreversible electroporation leading to cellular death. We

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aim to present the case of a high-voltage electrical injury in a pediatric patient and the cardiac complication that followed- acute mitral valve insufficiency caused by chordae tendinae rupture.
Method: A 16-year-old male patient climbed a transformer pole to take a photograph and touched a 25.000 V source of alternating current, leading to cardio-respiratory arrest with a favorable response to resuscitation measures. He was brought to the hospital intubated and ventilated, GCS 3. We ran complete lab tests and cardiac, neurologic, and plastic surgery evaluations.
Results: On admission, the patient presented with grade III burns on 5% of the body surface. The entry point was on the right hemithorax. Cardiac ultrasound showed moderate systolic dysfunction of the left ventricle (LV), prolapse of the anterior mitral valve due to chordae rupture (that could also be visualized moving freely in the LV cavity), and moderate mitral regurgitation, with normally-sized left atrium. Although the electrocardiogram was normal at admission, it revealed sinus tachycardia and ventricular premature beats 5 days later. Lab tests showed elevated markers of myocyte injury that normalized within 8 days. Upon detubation, the patient showed a minimum state of consciousness and spastic cerebral palsy.
Conclusions: Electrical injuries are an intriguing aspect of medical care, as the extent of the external burns does not predict the severity of the internal injuries. When the electricity penetrates the thorax, cardiac lesions are most probably present. The electric current causes patchy necrosis of the myocardium rather than following the distribution of a coronary artery, therefore correlations of the electrocardiogram aspect with cardiac ultrasound and biomarkers are useful. A particular aspect of our case is that mechanical lesions such as cardiac ruptures are hardly diagnosed because of their poor survival rates. The patient requires close monitoring in dynamic, as the intricate cardiac and neurologic pathology poses a challenge in terms of therapeutic management.

Keywords: high voltage, electric shock, chordae tendinae rupture, mitral regurgitation

Interventional Cardiology

PP-198
Sharing is learning- the experience of a tertiary care center in percutaneous atrial septal defect closure

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Background and Aim: The technological breakthrough in the last decade has changed the approach to atrial septal defects (ASD) closure, shifting the therapeutic option from the surgical gold standard to transcatheter procedures. This paper is intended to share the experience of percutaneous closure of atrial septal defects in children in a tertiary care institute.
Method: In this cross-sectional study we included the pediatric patients who underwent transcatheter closure of ASD at the Department of Pediatric Cardiology, "M.S.Curie", Emergency Clinical Hospital for Children, Bucharest, Romania, from January 2012 to December 2022. We included patients with hemodynamically significant ostium secundum ASD or a history of cryptogenic shock, with satisfactory defect rims and no or mild pulmonary hypertension. Patients with defects other than ostium secundum, too large or lacking rims, were excluded, as well as those with high pulmonary resistance and those with associated congenital heart defects requiring surgical correction.
Results: Of 103 patients, 74 (71,84%) were females and 29 (28,16%) were males. Mean age at the moment of the intervention was 8,6 years old and mean defect size was 14,6 mm. Personal history of congenital cardiac disease was noted in 30 (29,12%) patients, out of which pulmonary stenosis and mitral regurgitation were the most common. The devices used were Amplatzer Septal Occluder in 42 (40,77%) patients and Cocoon Septal Occluder in 61 (59,23%) patients. In 92 (89,32%) patients the procedure was successful, whereas in 11 (10,68%) cases, conversion to surgery was required because of difficulties in positioning the device or device migration. Postprocedural complications were noted in 12 (11,65%) patients and consisted of small intradevice shunt, mild anemia, and pericarditis.
Conclusions: Device closure of secundum ASD is a safe and efficient method preferred in many centers in Europe. The first Pediatric Interventional Cardiology center in Bucharest has been established at "M.S.Curie" Emergency Clinical Hospital for Children and the results obtained are accordingly. Therefore, it is noted that despite the increasing difficulty of the cases over the years, the rate of complications and conversion to open surgery has lowered. Continuous study and practice are the keys to skills improvement and successful management of complex cases.

Keywords: congenital heart disease, atrial septal defect, device closure, transcatheter closure

General Cardiology

PP-200
Spontaneous coronary and cerebral artery dissections in a 15 year old girl

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Background and Aim: Spontaneous coronary artery dissection (SCAD) is an uncommon and potentially life-threatening cause of myocardial infarction. An intramural hematoma, not associated to arteriosclerosis or trauma, leads to compression of the true lumen causing an obstruction of the blood flow. SCAD has been associated with extra- coronary arteriopathies, in particular fibromuscular dysplasia. It affects predominantly female adults with a higher occurrence during peripartum period. Pediatric cases are extremely rare and mostly associated with an underlying connective tissue disease.
Method:

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Results: We describe an exceptional case of a 15 year old girl presenting with a Wallenberg syndrome: headache, vomiting and rapidly progressive dysarthria and dysphagia. MRA (magnetic resonance angiography) revealed recent ischemic events with absence of flow in segments of the vertebral artery and left posterior inferior cerebellar artery (PICA).
Transesophageal echocardiography in search of a patent foramen ovale to rule out a paradoxical embolism as cause for this ischemia, surprisingly revealed severe left ventricular dysfunction. Further imaging with computed tomography (CT) and coronary angiography demonstrated a non-recent dissection of the mid- and distal LAD (left anterior descending artery) and left ventricular remodeling.
Late and specific imaging of the affected vertebral artery was suggestive of dissection as cause of the stroke she presented with. Genetic analysis is ongoing.
Based on the scarce literature about SCAD, the patient was started on beta-blocker and antiplatelet therapy, as well as angiotensin converting enzyme (ACE)-inhibition in the context of heart failure treatment.
Conclusions: SCAD is very rare in the pediatric population, though needs to be considered in primary cases of myocardial infarction. Extra-coronary abnormalities are well described in adult literature but exceptional in children. They need to be thoroughly investigated and close follow- up is warranted.

Keywords: SCAD coronary artery dissection pediatrics

Congenital Heart Surgery

PP-201
LV decompression via valved conduit from apex to aorta descendens in a patient with hutchinson-gilford progeria syndrome and severe aortic stenosis

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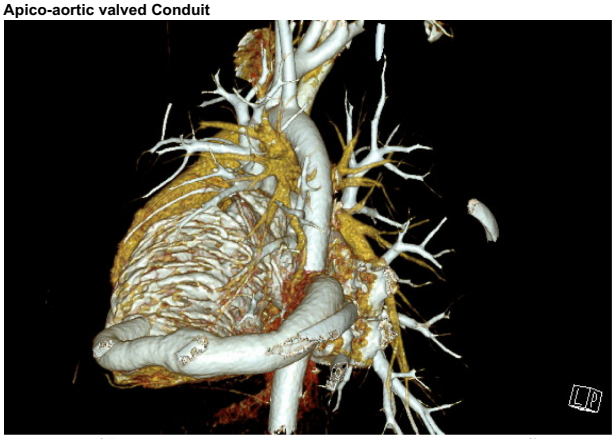
Background and Aim: Hutchinson-Gilford Progeria Syndrome (HGPS) is an extremely rare autosomal dominant syndrome in which progerin is produced, causing a characteristic nuclear blebbing. Patients infrequently live till adult age due to early coronary or carotid occlusion (dissection). Natural history has been changed by the Boston trial with Lonafarnib; as patients grow older, they develop in their 3rd decade premature thickening of the aortic and mitral valve leading to severe stenosis. Balloon dilation typically will provide only temporary result; surgery is esteemed extremely risky due to high friability; transcatheter aortic valve replacement is mostly no option because of small annular size, risk of coronary compression due to close proximity and size mismatch, and absence of adequate vascular access.
Method:
Results: A 22 year old male with HGPS presented with angina due to severe aortic stenosis and arterial hypertension; the coronary arteries appeared normal on coronarography. A percutaneous balloon dilatation was performed with good result. Unfortunately 9 months later the patient presented in lung edema due to rapidly evolving restenosis of the aortic valve, for which an emergency redo balloon dilation was performed. Again this brought only temporary resolution and a strategy was discussed with Pedro Del Nido, Boston USA. Informed consent was obtained. The patient, now age 24 years (L 129cm, W 20kg), was brought in for elective surgery 7 months after the second balloon dilation. An apico-aortic valved custom made off-label conduit was constructed: it consisted of a 10 mm apical Berlin Heart canula connected to a 12 mm Venpro (wrapped in 12 mm Dacron) leading onto a 10 mm Gore-Tex tube which was anastomosed end-to-side on the thoracic aorta. The surgery was well tolerated and post-surgery echocardiography demonstrated a decompressed left ventricle and well-functioning conduit with only trivial aortic regurgitation. The native aortic valve opens with antegrade flow and the patient reports clear improvement of complaints: no more angina pectoris and good exercise tolerance (NYHA I) with 6 months of follow-up.
Conclusions: HGPS is an extremely rare genetic condition associated with premature aortic stenosis requiring difficult management. We report the third valved LV apical to aorta conduit with successful decompression of the left ventricle.

Keywords: HGPS, Aortic stenosis, apico-aortic valved conduit

Apico-aortic valved Conduit



post-surgery CT scan demonstrating the apico-aortic valved custom made off-label conduit

General Cardiology

PP-202
The prevalence and natural history of persisting ductus arteriosus in full-term children

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Background and Aim: Ductus arteriosus (DA) is a part of fetal circulation. Typically, the ductus arteriosus (DA) closes during the neonatal cardiac transition period shortly after birth. However, in some neonates, the closure is delayed. When this closure does not occur, the condition is referred to as a persisting ductus arteriosus (PDA). PDA has previously been associated with preterm birth. However, cases of PDA with no known predisposing factors are a well-known phenomenon. Utilizing the Copenhagen Baby Heart study, the aim of this study is to assess the prevalence and natural history of persisting ductus arteriosus when not associated with preterm birth.
Method: The Copenhagen Baby Heart Study prospectively included 25,000 neonates with an echocardiography performed in the neonatal period. In this study, we included individuals with an open DA at the time of echocardiography as cases. All cases were offered a follow-up echocardiography. A retrograde jet in the pulmonary projection were diagnostic of PDA and referred to a pediatric cardiologist.
Results: Out of 565 participants, 409 underwent a follow-up examination (73%). The mean age at examination was 3.5 years (IQR 1.9). Among the 409 who were successfully examined, 36 cases of PDA were found (9%). Data analysis is ongoing.
Conclusions: Our preliminary results provide an understanding of the prevalence of PDA in a Danish child population. After data extraction and data management, we will test maternal, prenatal, postnatal, and intrapartum variables as predictors of having a PDA. Furthermore, we will compare echocardiographic parameters between children with PDA and otherwise healthy children in the Copenhagen Baby Heart cohort. We hope to enhance the understanding of factors associated with the presence of PDA and investigate whether an otherwise asymptomatic PDA has echocardiographic measurable consequences for the heart.

Keywords: Congenital heart defect, Patent ductus arteriosus, Ductus arteriosus, Echocardiography, Neonatology

PP-203
Maternal and neonatal characteristics associated with delayed closure of ductus arteriosus in the copenhagen baby heart study

Anton Friis Mariager[1], Alberte Hammeken[2], Mikkel Malham[1], Raheel Altaf Raja[3], Johan Navne[8], Anna Sellmer[4], Signe Levring Jessen[5], Kasper Iversen[6], Henning Bundgaard[7] and Dorthe Lisbeth Jeppesen[1]
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Background and Aim: The ductus arteriosus (DA) is a part of fetal circulation. Normally, the DA closes during the neonatal cardiac transition period immediately after birth, but for some neonates, closure is delayed. Although delayed closure is thought to be a common phenomenon, little is known about the etiology of such cases in otherwise heart-healthy and term-born children. Utilizing the Copenhagen Baby Heart Cohort, the aim of this study is to assess maternal, prenatal, and perinatal factors associated with an open DA in term-born neonates within the first 28 days after birth.
Method: The Copenhagen Baby Heart Study database consists of more than 25,000 examinations. We created a cohort consisting of neonates found with an open DA at baseline. All neonates born at full-term who underwent an echocardiography within the first 28 days of birth were included (N=21,610). Neonates with other congenital heart defects than atrial septal defects were excluded. The primary outcome was an open DA based on echocardiography. The cohort was stratified by age at examination: under seven days and after day seven. We employed log-binomial regression to test the association between the binary outcome of having an open DA and predictors.
Results: Under day seven (n=6,010), 6% had an open DA (n=382). In this group, hypoglycemia (RR=5.5, 95%CI=2.4-12.8), a persisting foramen ovale (RR=1.8, 95%CI=1.3-2.5), and African (RR=4.1, 95%CI=2.5-6.7) or Asian (RR=2.1, 95%CI=1.4-3.1) ethnicity were significant predictors. After day seven (n=15,600), 0.7% had an open DA. In this group, poor Apgar score after one minute was a significant predisposing factor (RR=3.5, 95%CI=1.3-9.3). Including all participants, female sex (RR=1.2, 95%CI=1.0-1.4), hypoglycemia (RR=5.6, 95% CI=2.3-13.6), African (RR=2.7, 95% CI=1.2-5.9) or Asian (RR=1.9, 95% CI=1.3-2.8) ethnicity, obesity (RR=1.7, 95% CI=1.1-2.6), larger neonates (RR=1.64, 95%CI=1.0-2.6), and type one diabetes (RR=2.8, 95%CI=1.2-5.9) were significant predictors.
Conclusions: Our study identified several factors that predicted the presence of an open DA in an early echocardiography. We found indications of a difference between cases of open DA before and after day seven, perhaps indicating differences in the mechanism of persistence. The results provided can give us valuable insight into the etiology of one of the most common congenital heart defects.

Keywords: Congenital heart defect, Patent ductus arteriosus, Ductus arteriosus, Echocardiography, Neonatology

PP-208
Particular cardiac changes in a group of children with spinal amyotrophy

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Background and Aim: Spinal Amyotrophy (SMA) is an autosomal recessive genetic condition which consists in the loss of function of the SMN1 gene, resulting in the degeneration of alpha motoneurons from the anterior horns of the spinal cord and leading to motor deficits. Cardiac complications in these patients are poorly described in the literature, particularly regarding ECG

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abnormalities. In patients with Type I SMA, ECG baseline tremors have been reported, while those with Type II SMA may exhibit sinus tachycardia, right bundle branch block, Q waves, ST segment changes.
Method: We conducted a study on 22 patients with Type I or II SMA who were undergoing treatment with Onasemnogene, Nusinersen or Risdiplam. The study was conducted between May and October 2023. Cardiac evaluations included clinical cardiological examination, ECG, echocardiography, cardiac enzymes (for those treated with Onasemnogene), and ECG holter(in one case). Monitored ECG changes included Q waves >3 mm and/or duration >0.04 seconds, present in ≥2 leads (excluding DIII and AVR leads), PR interval, QRS duration, QT, ST segment changes, and presence of extrasystoles.
Results: Out of 13 patients with Type I SMA, ranging in age from 2 months - 5 years, 9 patients (69%) presented ECG changes: 4 patients had Q waves>3 mm in ≥2 leads, and 5 patients had sinus tachycardia. Echocardiographic findings revealed 3 patients with atrial septal defect/patent foramen ovale (PFO) and 3 with recurrent/persistent pericardial effusion. Laboratory results showed elevated troponin I (175 pg/ml) in 1case. Out of 9 patients with Type II SMA, aged between 1-13 years, 8 patients (88%) presented ECG changes: 3 patients had Q waves>3 mm in ≥2 leads, 4 patients had sinus tachycardia, and 1 patient had supraventricular extrasystoles. Echocardiographic findings revealed 1 patient with PFO. Notably, 3 patients had Q wave amplitude>10 mm in DIII or AVR.
Conclusions: Treatment options for SMA Type I and II have significantly advanced in recent years with new drugs improving and increasing the life expectancy of these patients. As a result, previously undescribed cardiac changes can now be highlighted.
We consider that additional comprehensive studies on the cardiological aspects of these patients are necessary to establish a global monitoring protocol.

Keywords: Spinal amyotrophy, ECG, Q waves, cardiac complications

PP-209
Real-life use of chatgpt-4 to write letters in the paediatric cardiology outpatient clinic

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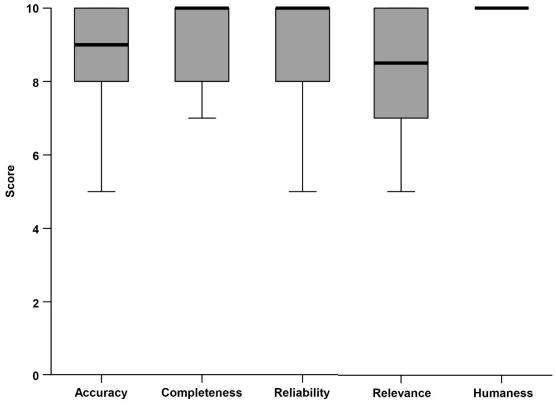
Background and Aim: Chat Generative Pre-Trained Transformer (ChatGPT) based on large language models may help generate clinic letters. The real-life use of ChatGPT to write patient referral letter has not been determined. This study aims to determine the quality, consistency, and limitations of ChatGPT-4, released in March 2023, for the generation of letters in real-life paediatric cardiology outpatient clinic setting.
Method: We created 10 clinical scenarios based on real-life encounters with children and adolescents in the paediatric cardiology outpatient clinic. The ChatGPT-4 was instructed to write 4 clinic letters to the dentist regarding antibiotic prophylaxis against bacterial endocarditis, 3 letters to schoolteachers regarding exercise prescription, and 3 letters to family doctors cautioning on drug use and vaccination. The 10 prompts were entered twice 24 hours apart. The first letter was scored in terms of "factual accuracy", "completeness", "reliability", "relevance", and "humanness" by 3 paediatric cardiologists using a Likert scale of 0-10. Inaccurate and missing information was noted. Consistency of information provided in the first and second letters was determined.
Results: The intraclass correlation coefficient of scores of the 3 raters was 0.79. The first ChatGPT-4 generated letter provided correct major recommendation in 8 (80%) of the 10 case scenarios. ChatGPT-4-generated letters achieved the highest scores in 'humanness' (median 10.0, IQR 10.0 to 10.0), 'completeness' (median 10.0, IQR 8.0 to 10.0) and 'reliability' (median 10.0, IQR 8.0 to 10.0). The scores in "factual accuracy" (median 9.0, IQR 7.5 to 10.0) and "relevance" (median 8.5, IQR 7.5 to 10.0) were lower. Inaccurate information, related to incorrect citation of guidelines, exercise recommendations, and general medical advice, was found in 7 (70%), while missing information related to drug prescription and medical complications was found in 4 (40%) of the 10 letters. Comparisons between the first and second letters revealed that ChatGPT-4 provided consistently correct major recommendations in 8 (80%) out of 10 scenarios, consistently incorrect recommendation in 1 scenario, and discordant major recommendation in 1 scenario. Neural hallucination was noted in 1 letter.
Conclusions: ChatGPT-4 can generate letters of reasonably high quality and consistency on commonly encountered clinical scenarios in paediatric cardiology.

Keywords: ChatGPT-4, clinic letters

Box plots showing the distribution of scores for the 5 aspects of the quality of ChatGPT-4-generated clinical letter



The line within the box represents the median, the box represents the interquartile range, and the whiskers represent the range of the scores.

PP-210
Appropriateness of kawasaki disease recommendations to parents and paediatric trainees from CHATGPT-4

Bosco Wing-ki Chik[1], Kwok Lap Chan[2], Sabrina Siu Ling Tsao[2] and Yiu Fai Cheung[2]
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Background and Aim: Appropriate Kawasaki Disease (KD) recommendations are necessary for parents of KD patients and paediatric trainees to optimise patient care. This study aims to determine the quality and appropriateness of advice regarding KD generated by

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Chat Generative Pre- Trained Transformer -4 (ChatGPT-4) for parents of KD patients and paediatric trainees.

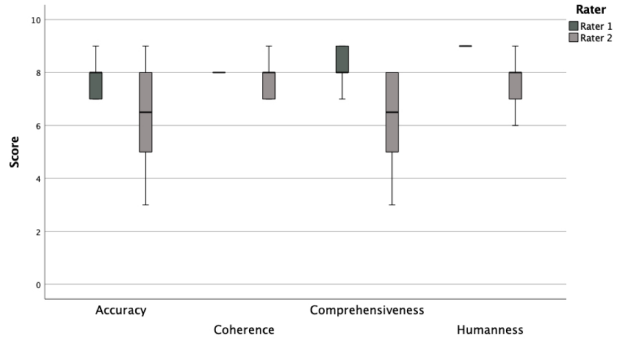
Method: We created 2 sets of clinical scenarios. In the first set, ChatGPT-4 was instructed to respond to 10 questions based on enquiries from parents of KD patients. Responses were scored in terms of "factual accuracy", "coherence", "comprehensiveness", and "humaneness" by 2 paediatric cardiologists using Likert scale of 0-10. Readability were calculated using Flesch reading-ease test. In the second set, ChatGPT-4 was instructed to respond to 8 KD-related questions based on enquiries from paediatric trainees. Responses are graded based on "relevance", "reliability" and "comprehensiveness" using Likert scale of 0-10 by 2 paediatric cardiologists independently. Reviewers would determine whether major advice from chatGPT-4 would be adopted in clinical judgement. Ratings were presented as median and interquartile range (IQR). Readability score were presented as mean and standard deviation (SD).

Results: For parent-targeted responses, ChatGPT-4 achieved the highest scores in 'humaneness' (median 9.00, IQR 8.00 to 9.00), 'coherence' (median 8.00, IQR 7.00 to 8.00) and 'comprehensiveness' (median 8.00, IQR 6.25 to 8.00). Inaccurate information regarding drug prescription, surgical indications, and disease prognosis is found in 80% of scenarios. Missing information regarding coronary complications and long-term cardiac assessment is found in all 10 scenarios. Mean readability of parent-targeted responses is 71.70 ± 6.26, a readability level easily understood by 12-year-olds. For paediatrician-targeted responses, ChatGPT-4 achieved the highest scores in 'relevance' (median 9.50, IQR 8.25 to 10.0). Scores in 'reliability' (median 8.50, IQR 6.00 to 9.75) and 'comprehensiveness' (median 7.00, IQR 6.25 to 7.00) are lower. Inaccurate information regarding thrombophylaxis, coronary imaging or interventions, and immunization is spotted in 37.5% of the scenarios. Missing information regarding patient education and stress imaging is found in 25% of the scenarios. Both reviewers would adopt ChatGPT-4's advice in 87.5% of the scenarios.

Conclusions: ChatGPT-4 has significant limitations in accuracy and lacks salient information when providing KD recommendations for parents and paediatric trainees.

Keywords: ChatGPT-4, Kawasaki Disease, Disease Recommendations

Boxplot showing distribution of scores for the 4 aspects of the quality of parent-oriented recommendations generated by ChatGPT-4.



The dark grey box represents ratings from rater 1, and the light grey box represents ratings from rater 2. The line within the box represents the median, the box represents the interquartile range, and the whiskers represent the range of the scores.

PP-211

NT-probnp a useful marker for diagnosis in the emergency room of cardiological disease?

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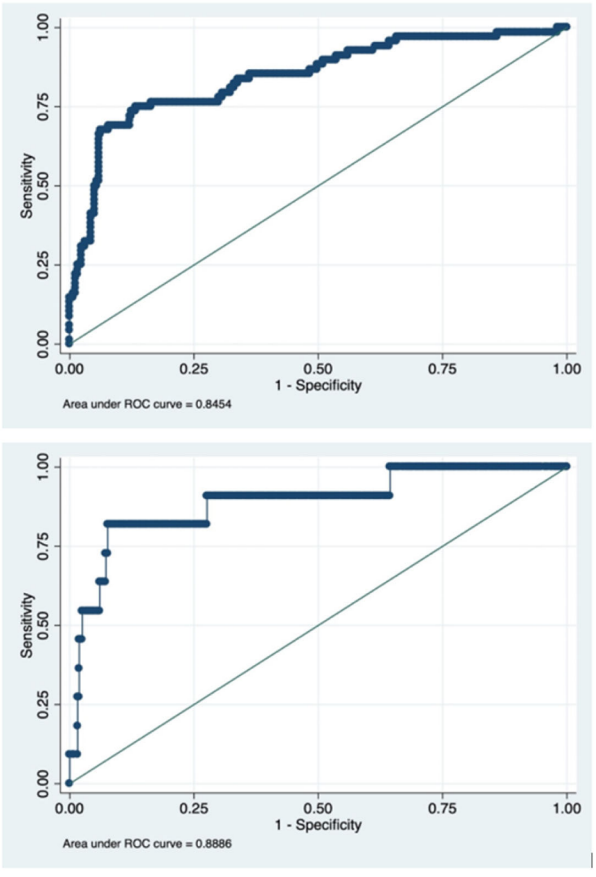
Background and Aim: In pediatrics, there is growing evidence of the role of the NT-proBNP as a severity biomarker in several clinical contexts.

Our main goal was to find out if the presence of myocardial stress detected by the elevation of this biomarker in previously healthy patients evaluated in the pediatric emergency rooms might be usefull for the detection of cardiological alterations which require treatment.

Method: An observational retrospective study of transversal cohort which include patients evaluated in the pediatric emergency room of a third level Spanish hospital with NT-proBNP analysis from January of 2018 to December 2021.

The NT-proBNP Zlog was calculated adjusted to age in days of life, considering high a Zlog>1,96. Populations was divided in 2 groups in base to this value. Prognosis variables were exitus and need for cardiological treatment during the episode. Descriptive statistics was performed. Though analysis of logistic regression

ROC curves



Area under ROC curves, sensibility, and specificity for need of cardiological treatment (First image) and for exitus (Second image).

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multivariant the independent association of high NT-proBNP with the prognosis variables adjusting by comorbidities as a confounding factor. Analysis by ROC curves were performed to explore the validity and prognosis value of high NT-proBNP. The statistical analysis was performed with STATA v.16. p value of <0.05 was considered significant.
Results: 316 patients were included (medium age 3 years (0.5-8 years); 62% male; 28% with comorbidities, with a medium level of 295 (95-1378) pg/ml (Zlog of 1.6). Cardiological studies was performed in 48%, 21% required cardiological treatment and mortality was of 3.5%.
A 35% presented high NT-proBNP, presenting theses patients more exitus risk (OR 20 CI 95%) in the multivariate analysis.
The area under the RCO curve, sensibility and specificity of the Zlog value of NT-proBNP 1.96 for the need for cardiac treatment were: 0.84 (IC95% 0.78-0.90); 0.76 and 0.80 respectively and for mortality they were: 0.88 (IC95% 0.77-0.95): 0.91 and 0.70.
Conclusions: The presence of high values of NT-proBNP in patients in which this biomarker is requested from the emergency room seems to be associated with a higher risk of exitus and need for cardiological treatment, for this, NT-proBNP might be a useful biomarker for screening of potential patients with cardiological risk or underlying cardiovascular conditions in the emergency room.

Keywords: NT-proBNP, biomarkers, emergency

PP-215

A case of aortic atresia, ventricular septal defect & normally developed left ventricle, with disconnected right common carotid & subclavian arteries

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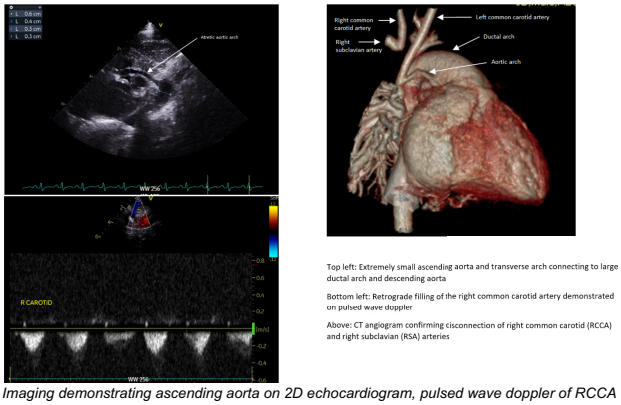
Background and Aim: Aortic atresia is often associated with a hypoplastic left ventricle, but approximately 4-7% of patients with aortic atresia develop a normal size left ventricle due to an associated ventricular septal defect (VSD). A Yasui operation, performed in stages, can be used to maintain a biventricular circulation in these patients. Good outcomes can often be achieved but cardiopulmonary bypass (CPB) typically involves cannulation of the ascending aorta or innominate artery to preserve cerebral blood flow during cardiac surgery.
Method: We report a rare case of aortic atresia and hypoplastic aortic arch (HAA) associated with a large VSD, normally developed left ventricle and disconnected right common carotid (RCCA) and subclavian (RSA) arteries. The patient was antenatally diagnosed and delivered at full term following induction of labour. A prostaglandin infusion was commenced and the patient was transferred to the tertiary paediatric cardiac surgical centre. Postnatal echocardiogram confirmed aortic atresia with a tiny ascending aorta measuring 2-3mm and HAA, with retrograde aortic flow perfusing the coronary arteries. The left common carotid arose from the distal aortic arch; the ductal arch gave rise to the left subclavian artery posteriorly. The RCCA and RSA were demonstrated on echocardiogram, with retrograde filling of the RCCA from head/neck vessels, but with no visible connection to the aortic arch. Cross sectional imaging confirmed these findings.
Results: The patient was discussed in the joint congenital cardiac meeting, considering possible surgical strategies and level of risk associated with a tiny ascending aorta, but also the complex difficulties that CPB would present with a disconnected RCCA and RSA. The decision was made to offer compassionate medical care only as any surgical strategies were felt to carry an unacceptably high risk and low chance of a good result.
Conclusions: Surgical decision making in congenital heart disease is complex and requires detailed, high quality echocardiographic and often cross-sectional imaging to ensure decisions are made based on the correct information. All aspects of the cardiovascular anatomy need to be considered both in terms of the possibility of surgical palliation or repair but also the practicalities of CPB and the associated risk profile.

Keywords: Aortic atresia, hypoplastic aortic arch, disconnected common carotid artery, disconnected subclavian artery,

Echocardiographic and cross-sectional imaging of ascending aorta and disconnected RCCA & SCA



Imaging demonstrating ascending aorta on 2D echocardiogram, pulsed wave doppler of RCCA and CT angiogram of aortic and ductal arches with disconnected RCCA & SCA

PP-218

Comparative metabolomics in single ventricle patients after fontan palliation: A strong case for a targeted metabolic therapy

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Background and Aim: Most studies on Single-ventricle circulation take a physiological or anatomical approach. Although there is a coupling between cardiac contractility and metabolism, the metabolic outlook in this patient population is very recent. Early findings point to major metabolic changes, with both impaired glucose and fatty acid oxidations in the cardiomyocytes. Also, Fontan patients have systemic metabolic disorders such as abnormal glucose metabolism and hypocholesterolemia.
Method: Our review compares the metabolism of the different types of heart failures, as HfrEF, HFpEF, Right-sided HF (RV-HF), as well as Non Failing Single-Ventricle (SV-NF) and Single-Ventricle Heart Failure (SV-HF), through a Pubmed search of the literature on cardiac metabolism, published in English from 1939 to 2023.

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Results: Early evidence demonstrates that Single-ventricle is not only a hemodynamic burden requiring staged palliation, but also a metabolic issue. Deep alterations of fatty acid and glucose oxidation, cellular respiration were found, resulting in metabolic instability, impaired energy production, which may be a cause of circulatory failure. Stimulating Ketone Oxidation may be an effective treatment strategy for heart failure in patients with BV. Few, but promising, clinical trials have been conducted thus far to evaluate therapeutic ketosis with HF using a variety of instruments, including Ketogenic Diet, Ketone Esters, and SGLT2 inhibitors. Initial trial from a small cohort demonstrated favorable outcomes for Fontan patients treated with SGLT2 inhibitors.
Conclusions: The rationale for a therapy targeting the metabolic disorders seen in Single-Ventricle patients was analyzed. Inducing cardiac ketolysis may provide an additional substrate for the single-ventricle heart and pleiotropic effects of ketone bodies may also be beneficial. To our knowledge, our review is the first to highlight the therapeutic potential of ketosis in the therapy of patients with single ventricle circulation.

Keywords: heart failure, Fontan, ketones, metabolism, single ventricle

PP-219

Drug-nutrient interactions of 45 vitamins, minerals, trace elements and aging-related cofactors with aspirin and warfarin

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Background and Aim: Aspirin and warfarin are among the most widely used prophylactic therapies in cardiology against thromboembolic events. Drug-drug interactions are generally well known. Less known are the drug-nutrient interactions, impede drug absorption and altered micronutritional status.
Method: Our review is comparing the impact of aspirin and warfarin on drug-nutrient interactions, based on a PubMed review of the literature on micronutrient interactions with acetylsalicylic acid (aspirin), or warfarin from 1936 to 2023.
Results: Both therapies might influence the micronutritional status of patients through different mechanisms such as absorption, transport, cellular use or concentration, excretion. Some of these mechanisms could be investigated to potentiate the drug effects. Scientific evidence on several essential micronutrients is scarce. Study designs vary greatly, rendering it difficult to assess their clinical relevance. Aspirin is generally recognized for altering micronutritional status of C vitamin, E vitamin, iron, folate. It may also: increase absorption of chromium; increase serum levels of magnesium; decrease serum levels of folate and ascorbic acid; increase urinary excretion of ascorbic acid, thiamine, folate, calcium and phosphorous; decrease urinary excretion of magnesium and sodium; increase urinary excretion of ascorbic acid, thiamine and folate, alters homeostasis of B12 vitamin, ascorbic acid, iron; and be potentiated by folate, tocopherols, tocotrienols, or taurine. Warfarin is generally recognized for altering micronutritional status of K vitamin, and altered C vitamin, E vitamin, iron, magnesium and zinc status are controversial. It may not interact with iron, nor magnesium, but may interact with polymorphism of folate homeostasis, reduce folate levels through K-vitamin food avoidance, have INR controlled in part by serum magnesium, and be potentialized by niacin, tocopherols and tocotrienols
Conclusions: Drug-nutrient interactions are seen in both aspirin and warfarin and could be managed through simple strategies such as a risk stratification of drug-nutrient interactions specific to the patient; a micronutritional status assessment as part of the medical history; extensive use of the drug-interaction probability scale to reference little known interactions, and apply a personal, predictive and preventive medical model using the omics.

Keywords: micronutrient, micronutrition, micronutritional deficiency, aspirin, warfarin

PP-220

Melas syndrome associated with congenital aortic stenosis: A case report

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Background and Aim: Mitochondrial encephalomyopathy, lactic acidosis and stroke-like episodes (MELAS) is a rare mitochondrial disease. It mainly affects nervous system and muscles, and it can be associated with a congenital heart disease.
Method: we present the case report of a newborn with MELAS syndrome and congenital aortic stenosis
Results: A preterm neonate was admitted to the neonatal Intensive care unit. She was born by cesarean section at 34 weeks gestation, to a 35-year-old-mother with hypothyroidism,sonsorineural hearing loss,recurrent headaches and history of 2 sons affected by MELAS syndrome. At birth, the newborn presented respiratory distress requiring intubation. Physical examination revealed an intense heart murmur with weak peripheral pulses. Laboratory finding showed a severe metabolic acidosis (pH: 6.9, HCO3-: 10.6 mmol/l) with hyperlactacidemia (11,7 mmol/l). Transthoracic echocardiography revealed congenial aortic stenosis with severe biventricular hypertrophy, and post-capillary pulmonary hypertension. Balloon aortic valvuloplasty was performed on the 3 th day of life. But the newborn succumbed due to severe hypoxemia. Considering family history, lactic acidosis and severe hypoxemia, diagnosis of MELAS syndrome with cardiac involvement in newborn was made.
Conclusions: Management of MELAS syndrome is largely symptomatic and should involve a multidisciplinary team. Patients with associated cardiac injury have overall higher morbidity and mortality.

Keywords: MELAS, aortic stenosis, mitochondiral diseace

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PP-221

The circular shunt - a challenge in the management of neonatal ebstein's anomaly

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Background and Aim: Neonatal presentation of Ebstein`s anomaly (EA) represents the most severe form of this condition, especially when it is associated with circular shunt (CS), a particularly lethal situation when the right ventricle (RV) is unable to generate forward pulmonary flow (RVFF) and there is severe pulmonary regurgitation (PR). This creates a large volume overload on the heart with low cardiac output. We review our experience with management of neonates with EA associated with CS.
Method: Between January 2021 and January 2023 four neonates with EA and CS were treated in our tertiary centre. The echocardiographic evaluation focused on: Carpentier type classification, GOSE index, pulmonary valve (PV) annulus size, grade and peak velocity of tricuspid regurgitation (TR), RVFF, presence and degree of PR.
Results: The initial echocardiography revealed Carpentier B type, GOSE II index (0.82/0.81/0.53), and normal PV size in 3 neonates. Two of them were hemodynamically unstable immediately after birth requiring mechanical ventilation (FiO2100%). The RV pressure was over 30mmHg (37/42/61mmHg) in all cases. RVFF was absent after birth, it commenced approximately 10 hours after birth (1-24hours), with the association of Sildenafil and Milrinone. In all cases serial echocardiographic evaluation revealed the appearance of moderate PR. In the presence of large ductus arteriosus (DA), all neonates developed CS, with elevated lactic acid levels (3.6/5.3/6.2mmol/l). The CS was interrupted by the spontaneous/pharmacological closure of the DA, with the subsequent reduction of PR, lactate normalization, and improvement of hemodynamics. The fourth case, a full-term newborn, severe cyanotic and hemodynamically unstable at birth, presented a more severe EA anatomy: Carpentier C type, GOSE III (1.4), severe TR, and low RV pressure (20mmHg). Severe PR was present at birth. In the absence of RVFF detection, Prostaglandin infusion was started. He developed a devastating CS with low cardiac output (lactic acid:10.5mmol/l), with death within 24-hours after birth, despite aggressive medical treatment with FiO2 100%, iNO, Milrinone.
Conclusions: Severe anatomy causes severe physiology with unfavourable results. Interruption of the CS is the immediate step to secure systemic oxygen delivery, by eliminating the DA flow (spontaneous closure or surgically). Aggressive treatment of pulmonary hypertension promotes RVFF.

Keywords: Ebstein`s anomaly, circular shunt, neonate

PP-222

A rare encounter – group B streptococcus infective endocarditis in a neonate with structurally normal heart

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Background and Aim: Infectious endocarditis (IE) is defined by the colonization of cardiac valve endocardium by infections agents. Since most cases occur in the presence of risk factors such as acquired or congenital heart disease, only 4% of IE arise in infants <1 year old with normal hearts. Although group B Streptococcus is a leading cause of neonatal sepsis, IE caused by this pathogen in infants is exceptionally rare, entailing a high morbidity and mortality rate.
Method: We present the case of a 20-day-old baby girl, with an uneventful birth and no risk factors, admitted due to one-day-old fever, lethargy, and lack of appetite.
Results: Upon admission, the clinical exam identified mild dehydration. Laboratory investigations revealed mild leukocytosis with neutrophilia, thrombocytopenia, and significantly elevated CRP (21.30 mg/dl) and procalcitonin (97,83 ng/ml). Despite extensive investigations, no infectious focus was identified. The case was interpreted as neonatal sepsis and empiric antibiotic treatment with Ampicillin, Gentamicin and Cefotaxime was initiated, with apparent clinical and biological improvement over the next days. Blood cultures identified the causative agent as Streptococcus agalactiae. On the fourth day of hospitalization, the physical examination identified a new diastolic murmur. This prompted the decision to conduct an echocardiogram that revealed two masses on the aortic valve, on the non-coronary cusp and on the right coronary cusp, causing moderate aortic insufficiency, with otherwise normal cardiac architecture. Subsequent echocardiographic assessments indicated a lack of reduction of the aortic mases, alongside with progressive increase in aortic regurgitation, left ventricle dimensions and a decline in LVEF to 44%, consistent with clinical deterioration. The patient was transferred to ICU, where inotropic therapy with Dobutamine and Milrinone was started, resulting in an improved clinical picture. Due to the patient's young age, the group B streptococcus etiology, lack of reduction of the vegetations under antibiotic treatment, and progressive heart failure, surgical therapy was proposed. The Ross procedure was performed, yielding no postoperative complications and a favorable outcome.
Conclusions: Even in front of septic neonates without any known risk factors, clinicians must remain vigilant for IE, as its management is complex, and its prognosis is burdened by high morbidity and mortality rates.

Keywords: Infectious Endocarditis, Neonatal Sepsis, Group B Streptococcus

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PP-223

Anthracycline effects on heart rate variability in children with acute lymphoblastic leukemia: Early toxicity signs

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Background and Aim: Anthracycline treatments are known to cause cardiotoxic long term side-effects in cancer survivors. Recently, a decrease in heart rate variability (HRV) has been identified in these patients, signaling the autonomic dysfunction, and altered cardiac fitness.

This study aimed at evaluating changes in HRV in children treated with anthracyclines.

Method: A total of 35 consecutive pediatric patients with acute lymphoblastic leukemia were evaluated by means of 24-hour Holter ECG, at baseline and after reaching half the total cumulative dose of doxorubicin equivalent (120mg/m2). Both time-domain and frequency-domain parameters of HRV were assessed, as well as any arrhythmic episodes, bradycardia, and tachycardia percentages.

Holter ECG parameters

Table 2. Holter ECG parameters

Parameter	Before treatment	After 120 mg/m² of doxorubicin equivalent	p value
HR	87.28 ± 13.8	103.2 ± 21.8	**<0.001**
Tachycardia (%)	7.79 ± 11.96	15.44 ± 22.26	0.19
Bradycardia (%)	30.07 ± 26.57	18.77 ± 22.56	**0.011**
PR interval (ms)	121.28 ± 19.3	135.58 ± 17.14	**0.012**
QTc (ms)	458.62 ± 73.6	442 ± 27.56	0.61
Time domain parameters			
SDNN (ms)	157.66 ± 49.37	114.95 ± 54.63	**0.002**
SDANN (ms)	125.48 ± 43.12	97.7 ± 47.61	**0.007**
rMSSD (ms)	109.76 ± 51.8	63.85 ± 42.35	**0.003**
pNN50 (%)	36.65 ± 18.87	17.84 ± 16.96	**<0.001**
Frequency domain parameters			
LF (ms)	0.47 ± 0.15	0.34 ± 0.15	**0.019**
HF (ms)	0.8 ± 0.35	0.51 ± 0.31	**0.018**
LF/HF ratio	0.65 ± 0.2	0.82 ± 0.32	**0.036**

*Data are shown as mean ± standard deviation.

Statistically significant decrease of HRV parameters following half the total cumulative dose of Doxorubicin equivalent.

Results: Results have shown a significant decrease in both time domain and frequency domain HRV parameters, following anthracycline treatment. Low-frequency (LF) to high-frequency (HF) parameters ratio, also displayed a significant difference (p= 0.035), suggestive of an early cardiac autonomic dysfunction. Of note, none of the patients presented symptoms of heart disease, elevated troponins, and only two patient presented echocardiographic signs of diastolic disfunction. In conclusion, the present study has shown that cardiac autonomic nervous system regulation is compromised in children treated with anthracyclines even before reaching the total cumulative dose.

Conclusions: Our findings emphasise the importance of cardiac monitoring during the oncological treatment, as HRV parameters could be used to determine early cardiotoxicity signs and need for treatment. Further studies are needed to establish the exact implications of these results for the long-term cardiac function and cardiotoxicity prophylaxis.

Keywords: cardiotoxicity, heart rate variability, childhood cancer, early cardiac dysfunction

PP-224

Integrated school-based screening for rheumatic heart disease and dental caries in MOZAMBIQUE

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Background and Aim: Rheumatic heart disease (RHD) and dental caries (DC) disproportionately affect children and young adults in sub-Saharan countries, with major impact on schoolchildren's health and education. DC in children with RHD constitutes an important risk for fatal complications. Our study aimed at assessing the feasibility of simultaneous RHD and DC screening in school environment.

Method: March 20–24, 2022, we performed an observational descriptive study of schoolchildren in a public school in Maputo City, Mozambique. RHD screening involved two stages: first, a physical examination (including cardiac auscultation and direct observation of the oral cavity), and second, an abbreviated echocardiography performed by a cardiologist. Rapid testing for group A Streptococcus (GAS) was done to every eighth child in the classroom and for those with signs suggesting recent infection, in accordance with the study protocol developed for screening. A multidisciplinary team collected the data. Data were analyzed using descriptive statistics.

Results: A total of 954 students (median age 9; range 6–15) were screened. One hundred and twenty-five participants were eligible for a rapid antigen test, of which 6 (4.8%) tested positive. On clinical evaluation 52 children (5.3%) presented a heart murmur. Echocardiography on 362 children showed borderline RHD in 35 children and definite RHD in 2 (0.6%); 1 child had a ventricular septal defect. Dental cavities were present in 444 (48.4%), despite 904 out of 917 students reporting brushing of their teeth once to three times daily (98.6%).

Conclusions: School-based integrated oral and cardiovascular screenings and use of rapid tests for GAS carriage provide crucial

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information to create customized preventive strategies for rheumatic fever (RF) and RHD in low- and middle-income countries (LMICs), in addition to detecting children at very high risk of bacterial endocarditis. The sustainability of such interventions and acceptability by health providers needs to be assessed.

Keywords: Integration, rheumatic heart disease, dental caries, schoolchildren

PP-227

Evolution of cardiac involvement in patients with multisystem inflammatory syndrome in children (MIS-C)

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Background and Aim: The aim of the study was to determinate the course of the cardiovascular involvement in patients with Multisystem Inflammatory Syndrome in Children (MIS-C) secondary to coronavirus 2 (SARS-CoV-2) infection.
Method: A prospective and retrospective observational study was carried out on patients under 18 years of age who met clinical criteria for MIS-C (With or without a confirmed diagnosis of COVID- 19) in a tertiary hospital. The study was based on ECG, echocardiographic evaluation, laboratory test supplemented with MRI or other studies when necessary.
Results: A total of 22 patients (median age 7.4 years, 64% boys) were hospitalized an average of 20,4 days, (longest 182 days). Half of them had relevant personal history (1 Graves' disease, 2 operated esophageal atresia and 4 had an underlying congenital heart disease). All of them had elevated inflammatory biomarkers: Serum D-dimer, C-reactive protein, NT-ProBNP and dehydrogenase lactate in the 100%, interleukin - 6 (95%) procalcitonin (90%), leukocytosis (63%). Among then, 64% had cardiovascular system involvement: 31,8% had decreased left ventricular ejection fraction, 27% pericardial effusion, 23% valvular insufficiency (atrioventricular valves), 8% alterations in baseline ECG and there was only one case of arrhythmia (supraventricular tachycardia). Coronary involvement (moderate aneurysm) was found in only one patient. MRI was performed in 6 patients with myocarditis data in 3 of the 6 studies. 68% of the cases were admitted to the Paediatric Intensive Care Unit (predominantly females and most extreme age groups) requiring vasoactive drugs up to 50%. Only one patient needed to be supported with Extracorporeal Oxygenation Membrane and unloading atrial septostomy with stent. Most of these abnormalities improved within a few days. No patient died, 100% were discharged home. Subsequent follow-up was completed by 95% of the sample, with resolution of the recorded heart failure and coronary involvement. There were no readmissions.
Conclusions: Data suggest that cardiac involvement is common in children affected by MIS-C. However, the prognosis appears to be favorable with very low mortality despite frequent multisystem involvement. In our serie, the results are similar to others published in multicenter studies, except that we found a lower rate of coronary aneurysm.

Keywords: COVID-19; Kawasaki disease; acute heart failure in children; coronary artery aneurysm; myocarditis, paediatric inflammatory multisystem syndrome temporally associated with SARS-CoV-2

PP-228

Appearances can be deceiving – a rare case of total anomalous pulmonary venous return associated with ALCAPA

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Background and Aim: We describe the case of an infant diagnosed with anomalous left coronary artery emerging from the pulmonary artery (ALCAPA) after successful surgical repair of infracardiac total anomalous pulmonary venous return (TAPVR). This association of heart lesions is a very rare entity and to our knowledge there is only one other similar case described in the literature.
Method: Multiple imagistic and angiographic exams were performed in order to establish the final diagnostic and the adequate course of treatment.
Results: The baby was transferred in our unit with a diagnosis of TAPVR for further investigations and treatment. Upon admission the echocardiography revealed non obstructive infracardiac TAPVR. The right coronary artery had a normal origin and course by both two-dimensional imaging and color Doppler. The left coronary artery appeared to have a normal origin by two-dimensional imaging but was not clearly seen by color Doppler, while the ECG showed no signs of ischemia. In post operative setting the patient continued to present respiratory distress syndrome and hemodynamic instability while his echocardiography demonstrated a dilated left ventricle with severe dysfunction, severe mitral regurgitation and endocardial fibroelastosis. The left coronary artery originated from the pulmonary artery and had retrograde flow while the ECG showed ischemic changes. The findings prompted cardiac catheterization which subsequently confirmed the diagnosis. Afterwards the patient underwent successful reimplantation of the coronary artery and went on ECMO support for 10 days. The patient continued to present severe hemodynamic instability despite adequate inotrope support and eventually died because of multiple system organ failure.
Conclusions: Although ALCAPA is a rare heart defect and it can be difficult to diagnose, the presence of a left-to-right shunt elevating pulmonary artery pressures or resistance can make it even more challenging to diagnose. Hence it is of upmost importance to have adequate coronary imaging both in two-dimensional echocardiography and color Doppler with serial evaluations.

Keywords: anomalous left coronary artery emerging from the pulmonary artery, ALCAPA, total anomalous pulmonary venous return, TAPVR

PP-229

Cardiac manifestations in a cystic fibrosis adolescent: Unveiling a diagnostic challenge

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Background and Aim: Cystic fibrosis (CF) is an autosomal recessive genetic condition characterised by the production of thick and sticky mucus, mainly affecting the respiratory, digestive, and reproductive systems. Although primarily recognized as a pulmonary disorder, emerging evidence suggests involvement of the CFTR protein (the underlying affected protein in CF) in the cardiovascular system. Postmortem studies in CF patients revealed cardiac structural changes, including ventricular fibrosis, lymphoedema, necrosis, and myocarditis. Other noteworthy literature findings include right ventricular hypertrophy and dilatation, prevalent in both adult and paediatric populations with CF.
Method: We present the case of a 16-year-old male with CF (homozygous F508del mutation) with chronic Burkholderia cepacia infection and moderate lung function reduction, who had recently migrated from Brazil to Portugal. Upon admission to the Paediatric ward for treatment of a pulmonary exacerbation, the patient exhibited frequent ventricular and supraventricular extrasystoles, prompting a comprehensive cardiology evaluation. Despite lung function deterioration and cardiac changes, no exercise intolerance was reported. Echocardiogram showed mild systolic dysfunction (LVEF 52%). Holter monitoring revealed 9% isolated ventricular extrasystoles and 15% isolated supraventricular extrasystoles. Cardiac MRI demonstrated normal biventricular dimensions and function with small (point-like) non-ischemic late gadolinium enhancement foci located in the basal mid-septal (RV insertion zone), basal anterior, basal mid-inferior and mid-lateral segments.
Results: Given the patient's origin in Brazil and epidemiological considerations, Chagas disease was suspected but ruled out through negative T. cruzi testing. Other diagnostic hypotheses included previous, unnoticed myocarditis and/or concurrent CF-related cardiac involvement. He was recently started on elexacaftor/tezacaftor/ivacaftor (which specifically targets the underlying CF protein defect) and heart rate has been controlled under atenolol.
Conclusions: This case explores the intricate relationship between CF and cardiac manifestations, emphasising diagnostic challenges. This presentation aims to underscore a potential impact of CFTR modulator therapy on CF in this patient. Given the pre-existing cardiac changes, we advocate for a particularly cautious approach during treatment initiation, while remaining open to the prospect of revealing potential beneficial effects. The observed complexity of this case emphasises the necessity of a holistic approach to CF care, integrating cardiac considerations into diagnostic and therapeutic strategies.

Keywords: Cystic fibrosis, Diagnostic Challenges, Holistic Health

PP-231

Multiple giant aneurysms in infants with kawasaki disease

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Background and Aim: Kawasaki disease (KD) is the most common vasculitis syndrome with coronary artery involvement in young childhood. However, forms with multiple giant aneurysms are rare. The authors would highlight the problems caused by this condition.
Method: We present the cases of three patients treated in our institution. The initial care and the studies performed during the follow-up are detailed.
Results:

1. The 11 years old boy developed KD at the age of 3 months. On echocardiography both the right and left coronary arteries were involved. The giant aneurysm on the right coronary artery regressed, while the scarification of the string of pearl-like dilatations on the LAD (left anterior descending) resulted in subtotal occlusion. Follow-up is performed by ultrasound, CT and dobutamine stress echo. He did not require any intervention. Ten years after he is on aspirin therapy.
2. The 1-year-old boy had KD at the age of 4 months. Echocardiography 1 week after the onset showed right coronary dilatation and giant aneurysms on the LAD and CX (circumflex) coronaries. The abnormalities confirmed by the initial CT one month after the first symptoms showed moderate regression on coronary angiography performed during the follow-up at the age of 1 year. Currently, the child is receiving thrombocyte aggregation inhibitor and anticoagulant therapy. No surgical or catheter intervention was necessary.
3. The 9 months old boy developed symptoms of KD at the age of 5 months. All coronaries were affected. 1 month after the onset CT angiography showed dilation of all the left main, LAD and CX coronaries. A string of pearl-like aneurysm was depicted on the right coronary. During follow-up giant aneurysms were observed in all the three vessels. There are no symptoms of myocardial ischemia. He is receiving thrombocyte aggregation inhibitor and anticoagulant treatment. Surgical or interventional therapy was not an option for him either.

Conclusions: All three patiets are alive. In the cases of the two younger children the course is still a question. Based on our experience and literature data, mainly drug therapy is available for Kawasaki disease in infancy. Surgical or interventional therapeutic options are limited in this age.

Keywords: Kawasaki disease, giant aneurism, infancy, multiple coronary involvement

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PP-233

What does meningoechephalitis look like at a conference like this?

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Background and Aim: Bacterial endocarditis is a low incidence entity in pediatric age, especially on native valve, and specially without associated congenital heart disease. It involves the formation of a thrombus or vegetation that can injure the endocardial or valve tissue, as well as embolize to distant extracardiac territories. The presentation is usually non-specific, following a subacute course in which fever and constitutional syndrome are the most frequent clinical expression and, although they may be accompanied by complications, these are a very rare mechanism of onset of the disease. We present the case of a 9-year-old male who came to the emergency room with high fever and acute neurological symptoms (decreased level of consciousness, meningeal irritation and decreased focal strength of 48 hours of evolution) in which a right fronto-temporal ischemic lesion was observed prior to echocardiographic visualization of vegetation in the anterior leaflet of the mitral valve secondary to bacterial endocarditis due to "Staphylococcus aureus". Despite the prompt broad-spectrum antibiotic coverage and the sensitivity of the causative germ (sensitive to cloxacillin, clindamycin and vancomin), the patient suffered an early valve perforation (first 12 days of admission) and, consequently, severe mitral valve insufficiency that has required prosthetic valve replacement in the first 2 months of evolution.
Method: Unique tertiary hospital centre case report
Results: Staphylococcus aureus complicated endocarditis whose onset corresponded to an acute cerebral isquémic embolic complication.
Conclusions: Although it is not frequent, bacterial endocarditis can debut with one of the extracardiac complications

Keywords: Native valve, embolism, endocarditis, perforation, Staphylococcus

PP-234

Cardiovascular phenotyping of children and adolescents with post-covid syndrome (PCS) at the time of diagnosis

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Background and Aim: Post-COVID Syndrome (PCS) is an emerging, highly relevant topic in public health and a condition that negatively affects quality of life and educational/work performance at all ages. To date, there is hardly any robust data on cardiac function in PCS (Long-COVID) available, particularly not in children and adolescents. The aim of our study is to pursue deep cardiovascular phenotyping in pediatric patients with PCS at baseline, using cardiac MRI and echocardiography, including strain/tissue tracking analysis.
Method: Prospective, single center cohort study at Hannover Medical School, Germany (10207_BO_K_2022). PCS was defined as follows: persistent symptoms such as reduced physical performance, poor concentration, mood symptoms, headaches, sleep disorders and dysosmia, for at least 12 weeks after PCR-confirmed SARS-CoV-2 infection. A total of 42 patients (age 9-18 years, 26 female) and 20 age/gender matched healthy controls (age 8-18 years, 12 female) were enrolled. All patients had persistent PCS-typical symptoms as defined above. Baseline data collection consisted of 12-lead EKG, protocol-driven echocardiography (echo), including tissue Doppler echo and biventricular strain analysis (TOMTEC, Philips; data pending), and cardiac MRI, including cine mass/volumes quantification in SAX and tissue tracking (strain) analysis of the RV and LV (cvi 42). Baseline laboratory studies included serum NTproBNP and troponin c.
Results: MRI-derived RV global radial strain (RVGRS) was significantly decreased (22.6±6.4 vs 26.8±6.9%; p=0.02), and RV mass index (19.1±3. vs 16.8±3.1 g/m2; p=0.008) increased in LCS patients vs. controls. Children with PCS also tended to have mildly reduced RVEF (50.9±5.1 vs. 53.1±3.6%; p>0.05) and RV circumferential strain (-13.5±3.5 vs. -15.0±2.8%), and slighty increased RVESV (45.1± 9.6 vs. 42.2±7.9%; p>0.05). In contrast, MRI-derived LV variables (LVEV, LVEDV, LVESV, LV mass), including tissue tracking (strain) analysis (LVGLS, LVGCS, LVGRS), revealed similar values in PCS and control subjects. EKG and echo data analysis is underway.
Conclusions: Post-COVID syndrome (Long-COVID) was associated with mildly decreased radial RV contractility (RVGRS) and moderate RV hypertrophy in our pediatric PCS cohort. In contrast, LV contractility (strain) and mass were not affected. Whether the aforementioned RV alterations are causal for the reported cardiopulmonary exercise limitations in pediatric PCS is unknown, and should be investigated further.

Keywords: Post-COVID, Long-COVID, COVID-19, cardiac function, MRI, children

PP-235

The use of automated analysis of the audio signals of the heart in the diagnosis of structural heart disease in children

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Background and Aim: Our aim was to investigate the ability of the artificial intelligence (AI) -based algorithm to identify a benign murmur from a pathological one.
Method: An AI -based algorithm was developed by fine-tuning a state of the art speech model using heart sound recordings collected at five university hospitals in Finland. Data collected from 1413 patients were used in training the model. The corresponding heart condition was verified using ultrasound. In the second phase of the study, patients referred to Helsinki University Hospital (HUS) due to a heart murmur were prospectively analyzed first with an

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algorithm and compared with clinical evaluation and echocardiography.
Results: Heart sounds of 98 pediatric patients were included. The algorithm identified 72 (73.5%) of the heart sounds as normal and 26 (26,5%) as abnormal. The echocardiography was normal in 63 (64,3 %) and abnormal in 35 (35.7 %) children (Table 1). The algorithm recognized abnormal heart sounds in 24/35 and normal heart sounds with normal echocardiography in 61/63 children. The details of algorithm analysis are presented in table 2. The confidence level of the algorithm analysis was confident in 92 (93.9%) and uncertain in 6 (6.1%) cases. When the murmur was audible, the sensitivity and specificity of the algorithm were 85.7% (24/28) and 96.7% (59/61) ($p<0.001$), respectively (Table 3). The average probabilities of defect (set by the algorithm) were 0.147 (0.028-0.265) and 0.810 (0.788-0.991) in children with normal and abnormal echocardiography and an audible murmur, respectively.
Conclusions: The algorithm was able to distinguish a murmur caused by a heart defect from a benign murmur with good sensitivity and specificity. Some of the structural heart defects do not cause a murmur and are thus not recognizable by the algorithm. Further research is needed on the use of the algorithm in screening for heart murmurs in primary health care among children. Innocent murmurs burden healthcare and this algorithm could be used in the future to screen those pediatric patients who need an ultrasound examination of the heart.

Keywords: AI, heart murmur, congenital heart defect, validation, heart sounds

Attachment

Image 1

Heart sounds analysis equipment

Table 1 Basic patient information

	N (% from patients)
Gender	
Male	48 (49 %)
Female	50 (51 %)
Age	
0-1 months	3 (3 %)
1-6 months	14 (14 %)
6-12 months	7 (7 %)
1-4 years	20 (20 %)
4-12 years	31 (32 %)
>12 years	23 (24 %)
Heart murmur	
Yes	89 (91 %)
No	9 (9 %)
Diagnosis	
Normal	63 (64 %)
Abnormal*	35 (36 %)
Shunt lesions	22 (22 %)
Valvular lesions	17 (17 %)
Other lesions	3 (3 %)

*One patient can have more than one diagnosis

Table 2 The diagnoses of patients with abnormal echocardiography

Algorithm analysis: Abnormal

Diagnosis (n)	Probability of defect
VSD 7	0.54-0.99
ASD 4	0.58-0.99
AS 4	0.87-0.97
PS 3	0.99
CoA 2	0.78-0.87
PDA 1	0.92
AS+PS 1	0.98
Others 2	0.94-0,95

Algorithm analysis: Normal

Diagnosis (n)	Probability of defect
BAV (no audible murmur) 3	0.03
ASD (no audible murmur) 2	0.18-0.33
VSD (no audible murmur) 2	0.09-0.25
ASD (audible murmur) 1	0.49
ASD (benign murmur) 1	0.11
VSD (audible murmur) 1	0.41
AI (audible murmur) 1	0.04
PDA (audible murmur) 1	0.07

Table 3 Patients with audible murmur evaluated by pediatric cardiologist (N=89)

		Echo		
		Abnormal	Normal/ Benign	
Algorithm	Abnormal	True positive 24	False positive 2	Pos. predictive value 0,92
	Normal/ Benign	False negative 4	True negative 59	Neg. predictive value 0,94
		Sensitivity 0.86	Specificity 0.97	Accuracy 0,93

Images and tables

PP-236

Marfan's syndrome: an overview

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Background and Aim: Marfan syndrome (MFS) is a rare genetic disorder with autosomal dominant inheritance, caused by pathogenic variants in the fibrillin-1 (FBN1) gene, causing defective microfibrillar organization and function in multiple tissues and organs. Around 75% of individuals with MFS have an affected parent and in 25% of cases FBN1 pathogenic variants occur de novo. The most prominent manifestations are aortic root (AO) dilation, mitral valve prolapse (MVP), ectopia lentis and skeletal abnormalities. Diagnosis is based on the Ghent II nosology. MFS is the most common cause of aortic aneurysm in children. Whereas surgical therapy remains the primary treatment to prevent aortic complications, medications that reduce hemodynamic stress on the aortic wall, such as β blockers or angiotensin receptor blockers may slow or prevent aortic growth. Our aim is to describe the cardiac features and molecular data of a pediatric population of MFS followed in a tertiary center.
Method: Descriptive retrospective review of patients' records.
Results: The study includes 25 patients with MFS, 56% male, with a median age at presentation of 9.8 years (minimum: 2.3 years, maximum 16.4 years) and a median follow up of 5 years. Eight patients (31%) had a family history of MFS and 6 were first referred due to an affected parent. All patients underwent FBN1 testing, and 18 patients (72%) exhibited a variant in FBN1. Throughout the follow-up period, 16 cases (61%) presented aortic root dilatation (zscore≥2.5), 19 cases (73%) exhibited ascending aorta dilatation and 14 cases (54%) demonstrated mitral valve prolapse and mitral regurgitation. Major manifestations in other organs comprised ectopia lentis (44%) and musculoskeletal abnormalities (16% underwent orthopedic surgery). Medical therapy was instituted in 16 patients (61%), and one underwent multiple open-heart surgery. No deaths were reported during the follow-up.
Conclusions: Early diagnosis and timely intervention are imperative to mitigate subsequent morbidity and mortality in cases of MFS among paediatric patients. Also, these patients should be managed using a multidisciplinary approach. This cohort underscores the importance of referring paediatric-aged first-degree relatives of individuals with MFS. This proactive approach is essential as these individuals are at heightened risk for aortic dissection and sudden death.

Keywords: Marfan, genetic, cardiac features

PP-237

Heart challenges in an adolescent patient with bulbar and spinal muscular atrophy

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Background and Aim: According to contemporary literature, rare individuals with bulbar and spinal muscular atrophy (BSMA) develop heart disease, characterized by ST-segment abnormalities,

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Brugada syndrome, dilative cardiomyopathy, or sudden cardiac death.
Method: we present case study, describing neurological and cardiac data of a patient diagnosed with BSMA.
Results: A 14-year-old adolescent was admitted to our Pediatric department, presenting with a history of proximal muscle atrophy, ubiquitous muscle cramps, general myalgias, and postural tremors since the age of 7. He also reported a progressive decline in mobility and fine motor control, as well as dysarthria and high CK level in medical history (>800U/L). Neurological assessments revealed progressive muscle weakness, predominantly affecting the bulbar and spinal regions, consistent with the BSMA diagnosis, which was subsequently validated through genetic investigations. Two months later, the patient presented with newly emerged symptoms, including fatigue, dyspnea, and palpitations, indicative of heart failure functional class III. Clinical and instrumental cardiologic investigations were conducted to comprehensively assess the cardiac status of the patient. Electrocardiogram findings demonstrated sinus rhythm with tachycardia (115 bpm, >98P according to age), small Q-waves in II, III, and aVF, QRS width of 90 ms, coved ST-segment elevation in right precordial leads (V1–V3), early repolarization pattern in II, aVR and aVF, normal QTc of 380 ms. The echocardiography revealed characteristics in line with dilated cardiomyopathy, encompassing ventricular dilation (LVEDD 55 mm), compromised systolic function (EF 20%), and the presence of pericardial effusion distributed around the heart (0.8–1 cm). The neurologists and cardiologists in our multidisciplinary team worked together to enhance the quality of care for the patient, starting a therapeutic regimen with steroids, beta-blockers, digoxin, ACE inhibitors, and diuretics. Over time, the patient's respiratory function deteriorated, necessitating the installation of a tracheostomy. Subsequently, the patient was transferred to a palliative care unit.
Conclusions: This clinical case underscores the significance of vigilance for cardiac involvement in adolescents diagnosed with BSMA, which shows abnormal electrocardiograms and structural anomalies, potentially contributing to an elevated susceptibility to sudden death. These observations underscore the crucial role of routine ECG screening and consideration of transthoracic echocardiography to evaluate cardiac risk factors in their health management.

Keywords: Bulbar and spinal muscular atrophy, cardiac involvement, adolescent health, dilated cardiomyopathy

PP-238

Heart rate and blood pressure patterns in duchenne muscular dystrophy

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Background and Aim: Duchenne muscular dystrophy (DMD) results in disability, respiratory issues, and heart failure. Numerous aspects of the pathophysiology related to cardiovascular system involvement remain unclear. The study's objective was to evaluate alterations in heart rate and blood pressure among boys with dystrophinopathy.
Method: We examined clinical data from the initial cardiology visits of 50 genetically confirmed DMD patients, aged 3.0 to 17.9 years. Notably, none were on cardiac medication, irrespective of their ages and disease stages. Throughout their ambulatory follow-up appointments, heart rate (HR) was assessed via pulsoximetry, and blood pressure (BP) was recorded using the oscillometric method. Normalization of measurements (z-scores) was performed using the Boston Children's Hospital Z-score. Results are presented as mean ± standard deviation. Statistical analyses involved the application of Pearson correlation to explore relationships within the dataset.
Results: The average HR was 107.92±14.89 (69.0 – 132.0) and the calculated z-score 2.07 ±1.03 (0.16 – 3.87). In 44 (88%) cases sinus tachycardia was diagnosed. The tachycardia prevalence was dependent on age ($r = 0.752$, $p < 0.01$). Only benign arrhythmias were found. SBP was 110±15 mmHg (73.7±23.88 percentile; z-score -0.69±1.0) and DBP 60±5 mmHg (68.8±13.2 percentile; z-score 0.49±0.42), anormal in 10% and 4% of measurements respectively. High SBP was found in patients on steroid therapy, and SBP values did not indicated low values at all. The correlation analysis reveals a modest positive relationship between the z-score of SBP and age ($r = 0.277$, $p = 0.051$), and, in contrast, the z-score of DBP demonstrates a negligible and statistically non-significant negative correlation with age ($r = -0.082$, $p = 0.573$).
Conclusions: These findings offer insights into cardiovascular dynamics in DMD, highlighting sinus tachycardia prevalence, and raising questions about its etiology - whether it is a compensatory mechanism to heart failure or progressive autonomous dysfunction. Additionally, the study reveals age- and treatment-related blood pressure variations. Further research is needed to enhance our understanding of these relationships and their clinical implications.

Keywords: Duchenne muscular dystrophy, arrhythmia, blood pressure variations

PP-239

Improving outcomes in neonatal enterovirus myocarditis

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Background and Aim: Neonatal enterovirus myocarditis (NEM) is uncommon and has a high mortality rate. Published literature states less than two thirds survive and approximately half requiring extra corporal membrane oxygenation (ECMO). Between July 2022 and March 2023, a spike of cases were managed in intensive care units (ICUs) in the South West of England and South Wales with an aggressive antiviral and anti-inflammatory strategy. We describe presentation and outcomes of this cohort and all historical cases from the previous two years.
Method: All children aged less than 30 days with a clinical diagnosis of myocarditis and positive polymerase chain reaction for enterovirus admitted to ICUs in Wales and the Southwest of England between March 2021 and March 2023 were identified and included.
Results: Fourteen neonates met inclusion criteria: 12 from the contemporaneous cohort; two historical cases retrospectively

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identified. All cases presented at nine to 24 days of life with short histories of dyspnoea, poor feeding and cardiogenic shock and required inotropes before transfer to ICU. At presentation, lactate, troponin-T, NT-pro brain-type natriuretic peptide and ferritin levels were universally significantly elevated. Electrocardiograms met criteria for myocarditis and echocardiograms demonstrated structurally normal hearts with severe left ventricular dysfunction and hypokinetic or akinetic left ventricular free walls. 11 of 12 contemporaneous cases were treated with combinations of antiviral and immunomodulatory medications (intravenous immunoglobulin, methylprednisolone, anakinra, pocapavir); all survived. Of the three cases not treated (one contemporaneous, two historical), one (33%) survived (p-value for difference=0.0048, 95% CI 14-94%). None were placed on ECMO. In survivors (n=12), median durations of inotropes, ICU and hospital stay were 35, 36 and 75 days respectively. One required a mitral valve repair and transplant referral due to myocardial dysfunction. Two required tracheostomies for protracted ventilation. All survivors were alive at 6-months.
Conclusions: This represents the largest cluster of NEM cases published to date. Despite extremely poor cardiovascular status at presentation and prolonged ICU support, all those treated with immunomodulatory and antiviral medications survived. This is in contrast to the published literature and our own historical cases prior to the implementation of immunomodulatory and antiviral medications in NEM, which should be explored further.

Keywords: Myocarditis, Cardiac Intensive Care, Heart failure

PP-241
Echocardiographic strain evaluation at one year post-MIS-C diagnosis: Persistent cardiac changes noted

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Background and Aim: The coronavirus infection disease 2019 (COVID-19) rapidly escalated into a global pandemic, leading to the emergence of Multisystem Inflammatory Syndrome in Children (MIS-C). It is characterized by a severe inflammatory response affecting multiple organ systems, with cardiac involvement in up to 80% of cases. This study aims to evaluate the long-term cardiac impacts of MIS-C, focusing on echocardiographic outcomes such as left ventricular global longitudinal strain (LV GLS), left atrial (LA) strain, and right ventricular (RV) strain one year post-diagnosis, comparing these results with a control group.
Method: In this retrospective study, we analyzed the cases of 22 children diagnosed with MIS-C who were admitted to Jeonbuk National University Hospital from January 1, 2021, to October 1, 2022. To facilitate a comparative analysis, an equal number of patients were selected to form a control group. Echocardiographic data were collected and compared at two distinct time points: at the time of diagnosis and at a one-year follow-up. The average interval between these two assessments was approximately 348.3 days.
Results: At the initial diagnosis, the MIS-C cohort exhibited a significant reduction in left ventricular ejection fraction (LVEF), as measured by the Simpson method, and in longitudinal strain (LS) across the apical 4 and 2 chamber views, as well as in GLS. Upon re-evaluation at the one-year follow-up, these patients continued to show decreased LVEF in the apical 4 chamber and overall, along with reduced LS in the apical 4 chamber and GLS. Additionally, the right ventricular (RV) free wall and global strain remained diminished when compared to the control group. Notably, there was a significant change in both LVEF and LS at the apical 4 chamber from the time of diagnosis to the one-year follow-up. Furthermore, the z-scores of all coronary arteries demonstrated notable changes over this period.
Conclusions: This study implies the persistent cardiac alterations in children with MISC even one-year post-diagnosis, particularly in both ventricular functions as assessed by strain calculation. These findings underscore the necessity for ongoing cardiac monitoring in MIS-C patients to better understand and manage the long-term cardiac implications of this post-COVID-19 syndrome.

Keywords: MIS-C, COVID-19, Echocardiography, strain

PP-242
Evolving trends in congenital heart disease: Insights from portugal (2000-2019)

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Background and Aim: We present data related to prenatal diagnosis (PND), demographic characteristics, prevalence, mortality and associated malformations of congenital heart disease (CHD) in Portugal from 2000 to 2019.
Method: The cohort comprises 6,521 CHD cases from the Portuguese National Registry of Congenital Malformations.
Results: The overall incidence of CHD increased from 326.6 per 100,000 live births in 2000 to 437.7 in 2019. The most common CHD type was left-to-right shunts, peaking at 314.62 per 100,000 in 2015. Critical CHDs showed a significant increase in PND, particularly for Tetralogy of Fallot and Hypoplastic Left Heart Syndrome, indicating advancements in diagnostic technology, improvement in prenatal diagnosis and increased awareness.
The progressive improvement in medical care is shown through a decreased in non-survivors beyond the first week. Notably, there was also an increase in medical terminations of pregnancy after 2009, possibly due to enhanced detection of severe anomalies.
In a sub-analysis of our cohort, 3,734 had other associated malformations, primarily from the Q80-Q89 group (other congenital malformations), followed by blood diseases (D50-D89 group). The average birth weight was approximately 2,731.6 grams. Prematurity and additional malformations significantly impacted survival, with non-survivors having a lower average birth weight (836.7 grams).
Conclusions: This comprehensive study provides insights into the evolving landscape of CHD in Portugal, emphasizing the importance of PND and the impact of associated malformations on patient outcomes.

Keywords: congenital, heart defects, trends

PP-243
Incidence and risk factors for pericardial effusions in children with anorexia nervosa

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Background and Aim: Anorexia nervosa (AN) is a growing medical problem worldwide associated with cardiac complications such as pericardial effusions, the incidence and etiology of which remains unclear. The aim of this cross-sectional study was to assess the

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incidence of pericardial effusions and it's possible risk factors in children with AN.
Method: The presence of pericardial effusions, defined as >3mm in any echocardiographic projection, was assessed at admission in 47 children (3 boys, 44 girls) aged 11.5-18 years (median: 14.75) with AN and severe malnutrition (BMI: 8.8-15.6; median: 13.6) hospitalized in years 2020-2023 in the Nutritional Clinic for nutritional treatment. The correlation of the amount of pericardial effusions with the patients' age, BMI, percentage of weight loss, time of weight loss and serum protein concentration was assessed and the difference between the subgroups with and without effusions was analyzed.
Results: Thirty of 47 patients (64%) were found to have pericardial effusions ranging from 4 to 27 mm. There was no correlation between the fluid amount and any of measured parameters: correlation coefficient (CC) for age (-0.01), time of weight loss (+0.04), protein concentration (-0.07)), except for BMI (CC=-0.5) and percentage of weight loss (CC=+0.5), where the correlation was close to statistical significance.
Differences between two subgroups of the study group: 17 patients without and 30 patients with pericardial effusions, were also analyzed. A statistically significant difference was stated only in the case of percentage of weight loss (median 18.5% vs 31%) ($p<0.01$). The difference between median BMI of the subgroups (14.1 vs 12.75) was on the verge of statistical significance ($p=0.05$). Other analyzed parameters: age (14.7 vs 14.75); time to weight loss (5.5 vs 6.5 months) and protein concentration (72 vs 68g/l) did not differ between the subgroups.
Conclusions: The presence of pericardial effusions is a common cardiac complication in children with AN, more common than previously thought
[1, 2]. There were no obvious risk factors for its occurrence other than percentage of weight loss above BMI alone, suggesting, according to recently postulated hypothesis
[2], that the pericardial space becoming too large for the wasted heart and filling with fluid instead, may be the cause.

Keywords: pleural effusions, anorexia nervosa, children

PP-244
Clinical symptoms in children with genetic aortopathies - a monocentric analysis with over 25 years of experience

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Background and Aim: Genetic aortopathy (GA) is a rare form of cardiovascular disease in childhood. The rarity of each disease makes it difficult to systematically describe the characteristics of these diseases. Here we present clinical data we have collected over the last 25 years from children with all forms of GA. From classic Marfan-syndrome (MFS) to Ehlers-Danlos-Syndrome (EDS) to Loeys-Dietz-Syndrome (LDS). Through our years of experience, we have one of the largest patient populations for these rare diseases.
Method: Since 1998 we investigated 846 patients in 3020 visits with suspected GA of which 304 patients were diagnosed clinically or genetically. We retrospectively analyzed the type of mutation, prevalence and age of onset of symptoms from birth to transition into adult medical care.
Results: Out of 304 patients, a mutation could be genetically detected in 231 patients. We have a patient population with 189 MFS, 4 EDS, 22 LDS patients and 16 with other mutations. The mutations occur between 0.3% (BGN) and 62.2% (FBN1). In 17.1% (n=52) despite clinical diagnosis no genetic variant was found. Especially in pediatric patients, diagnosis based on the Revised Ghent Criteria (RGC) alone is difficult. In our cohort only 23% of patients had a systemic manifestation at first clinical presentation at an average age of 7,9 yrs. At the time of transition, 43% had a systemic manifestation. Each symptoms present itself age dependent. E.g. Sinus Valsalva (SV) dilatation is present in 51% of cases at the age of 11,3 years. At the time of transition 72% show an aortic root dilatation. In contrast the symptom pneumothorax, with a prevalence of 3.7% (n=7) in our cohort, does not come close to these marks over the entire follow-up period.
Conclusions: In our large, monocentric pediatric patient group with GA, MFS remains the most frequent form of GA. Each of the typical symptoms presents itself age dependent and must be known in order to correctly diagnose and treat patients. Because of these data, we call for the development of a separate clinical score for children with GA to take more account of strict age dependency and to allow earlier clinical diagnosis.

Keywords: Genetic aortopathy, MFS, LDS

PP-245
Presence of bicuspid aortic valve in children with a genetic aortopathy changes the phenotype of aortic dilatation. implications for risk prediction

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Background and Aim: Bicuspid aortic valve (BAV) is a common congenital abnormality. Precise correlation with genetic aortopathies is missing. It is not known whether the presence of BAV in children with genetic aortopathies leads to a different phenotype of aortic dilatation. We hereby evaluated the correlation of appearance of BAV with other cardiovascular pathologies and the degree of dilatation of the ascending aorta in genetic aortopathies in childhood.
Method: Since 1998 we investigated 847 patients with suspected genetic aortopathies of which 305 patients were clinically or genetically diagnosed. 23 patients had a bicuspid aortic valve, of whom 15 children (65%) had a mutation in the FBN1 gene. We retrospectively analyzed the correlation of prevalence of aortic pathologies dilatation of sinus of valsalvae (SV) and ascending aorta, systemic manifestation of Ghent Criteria and mutations of genetic aortopathies with the appearance of BAV.
Results: Prevalence of BAV in pediatric patients with genetic aortopathies was 7,5%. Patients with a dilatation of the ascending aorta, had significantly more often a bicuspid aortic valve ($p<0.01$). Dilatation of sinus of valsalvae did not occur more often in patients with BAV. Analysis of correlation of BAV with other cardiovascular pathologies and systemic manifestation did not show a correlation.
Conclusions: In our large pediatric patient group prevalence of BAV in genetic aortopathies was 7,5%. The presence of BAV has no impact on the degree of dilatation of the SV, but the

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ascending aorta is significantly more often dilatated. Thus the phenotype of aortic dilatation is dependent of the presence of BAV even in patients with genetic aortopathies. This might have implications for the risk prediction of aortic dissection.

Keywords: Bicuspid aortic valve, genetic aortopathy, MFS

PP-246
Syncope in an asmathic patient somefact is missing?

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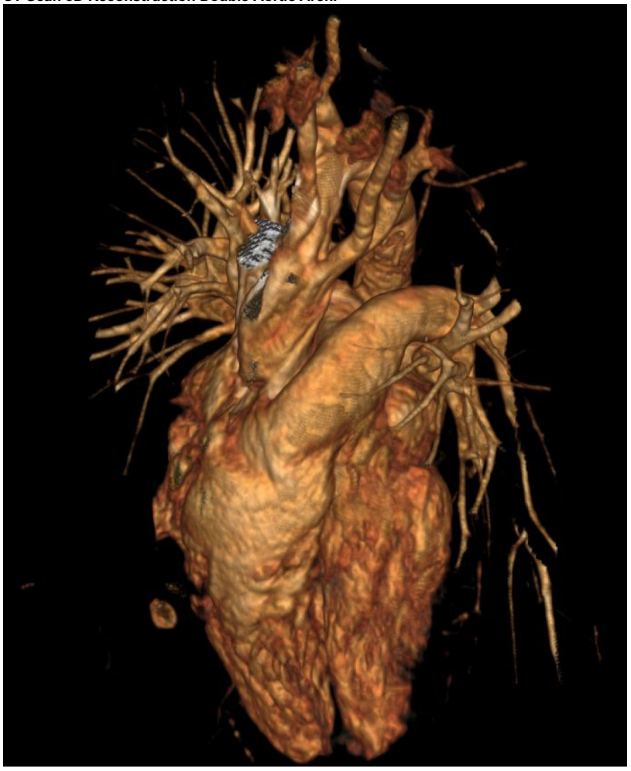
Background and Aim: Double aortic arch is the most common type of vascular ring. It involves the complete encirclement and compression of the trachea and/or esophagus by the aortic arch, its branches, or atretic ligamentous segments. Left untreated, it may lead to significant morbidity for the patient and can result in sudden collapse from airway compromise.
All of these symptoms are non-specific; hence, these patients can remain undiagnosed for many years. The incidence of double aortic arch is generally unknown.
A 14 year old patient with 4 year history of Asthma and Allergic Rhinitis diagnosis was remitted to a Pediatric Cardiologist evaluation due a syncope during coughing episode.

CT Scan 3D Reconstruction Double Aortic Arch.



Method: Case presentation 14 Year old female with 5 year diagnosis of intermittent exertional asthma and allergic rhinitis The symptons were intermittent episodes of persistent cough, nasal congestion and shortness of breath during maximal efforts mostly in running no symptoms during any other sports like dancing or swimming.The patient's medical history rebuilt a long standing history of intermittent respiratory symptoms which have been attributed to exercise induced asthma. The patient presented Cough mostly during night 1-2 episodes/ week despite appropriate treatment. A syncope during a coughing attack after exercise was presented.
Results: A Pediatric cardiology consultation and Echocardiogram were done, aortic arc abnormality was suspected. Then a CT scan showed a double aortic arch. A surgical intervention was performed. The patient was successful repaired with improving her symptoms after the surgery.
Conclusions: This case highlights the importance of considering alternative diagnosis when patients do not respond well to Asthma standardized treatments. Double aortic arch is a rare congenital heart defect and this clinical scenario remember us the importance of thinking about the possibility when you have no improvement like in this case. The early recognition of this condition, the appropriate diagnosis and management can lead to improved outcomes for patients with complex and rare congenital heart defects like a double aortic arch.

Keywords: Asthma, syncope, cough, double aortic arch

PP-247
Four cases of Q fever endocarditis in congenital heart disease

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Background and Aim: Q fever is a worldwide zoonosis caused by Coxiella burnetii. The clinical course of Q fever endocarditis is generally subacute and chronic; the disease may be present for years with only subtle symptoms and no vegetation visible on echocardiography while the bacteria gradually degenerates the heart valves. The aim of this study is to report four cases of Q fever endocarditis in congenital heart disease (CHD) patients at our Adult CHD Unit.
Method: Description of four cases of Q fever endocarditis diagnosed between 2015 and 2023 at our center. In this work we describe the clinical features, microbiologic results and imaging techniques; as wells as the treatments and outcomes of the 4 patients.
Results: None of the patients presented with fever except one. Time from initial symptoms to diagnosis was close to a year. In patient 2 it was suspected earlier because a family member was diagnosed with Q fever. PET-TC was helpful for diagnosis only in patient 1. All patients improved their symptoms after the specific Q fever treatment was initiated.

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Conclusions: Q fever endocarditis is easily misdiagnosed, not only because it is a rare condition, but also due to its chronic behavior with insidious symptoms and negative blood culture. We should be aware of Q fever in CHD patients with unexplained deterioration, splenomegaly and/or lupus nephritis.

Keywords: Coxiella burnetii, endocarditis, Q fever, treatment.

Table 1: main data of the 4 patients.

	PATIENT 1	PATIENT 2	PATIENT 3	PATIENT 4
AGE	44	41	24	17
CHD	PA+VSD	Tetralogy of Fallot	DTGA + VSD	PA+VSD
CHD REPAIR/ RESIDUAL LESIONS	Residual severe pulmonary regurgitation	Prosthetic pulmonary valve (2012)	Rastelli repair and Melody valve (2020)	Prosthetic pulmonary valve (2022)
INITIAL SYMPTOMS AND DATE	May 2016: diagnosis of lupus nephritis	May 2015: intermittent fever, profuse sweating, asthenia and dyspnea	April 2022: severe deterioration of functional cass (NYHA IV), pulmonary embolism and splenomegaly	December 2022: Pulmonary embolism. February 2023: diagnosis of lupus nephritis
BLOOD CULTURE	Negative	Negative	Negative	*S.lugdunensis*
TIME TO DIAGNOSIS	13 months	4 months	9 months.	11 months
TREATMENT	Doxycycline+HCQ	Doxycycline+HCQ	Doxycycline+HCQ	Doxycycline+HCQ

PP-248

Serum adipocytokines profile in children born small and appropriate for gestational age

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Background and Aim: Adipose tissue is not only a place where fat is stored, but also an endocrine organ that secretes bioactive molecules that influence the body's metabolism. Such molecules are known as adipocytokines.
In recent years, adipocytokines have been found to be associated with fetal growth restriction disorders.
The aim of this study was to determine the concentration of adiponectin, leptin and resistin in the serum of children born small for gestational age (SGA) compared to children born appropriate for gestational age (AGA).
Method: The study included 35 children aged 7 to 9 years (18 girls, 17 boys), born on term as small for gestational age (SGA), and 25 healthy peers (14 girls, 11 boys), born on term with normal birth weight (appropriate for gestational age - AGA). All children underwent physical examination, manual oscillometric measurements of arterial pressure. Adiponectin, leptin, and resistin concentration in serum were measured by an enzyme-linked immunosorbent assay.
Results: All patients had mean manual blood pressure values below the 95th percentile for sex and height. Adiponectin and leptin levels were significantly higher in the SGA group compared to the AGA group ($p = 0.023$, $p = 0.018$, respectively).Resistin values were comparable in both patient groups. In the SGA group, a positive correlation was found between serum leptin concentration and current body weight ($r = 0.28$; $p = 0.108$).In turn, adiponectin concentration in this group of patients correlated negatively with actual body weight ($r = -0.51$; $p = 0.002$). A negative correlation between body mass index and plasma adiponectin levels was found only in children born with SGA.In children with SGA, a negative correlation was found between serum adiponectin concentration and systolic blood pressure. In the SGA group, the phenomenon of catching-up growth was observed in 32 children.
Conclusions: Children born SGA have abnormal adipose tissue biomarkers profiles.

Keywords: adipocytokines, adiponectin, leptin, resistin, small for gestational age, children

PP-249

Enhancing neonatal screening for left heart obstructive defects: The added value of using perfusion index

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Background and Aim: Early detection of critical congenital heart defects has undergone significant improvement during the last decades due to advances in prenatal screening and the implementation of pulse oximetry screening (POS). Despite this, newborns with left heart obstructive defects, especially coarctation of the aorta (CoA) continues to be sent home without an in-hospital diagnosis. This underscores the need for improved neonatal screening methods.
Method: Retrospective, population-based study including all 38 full term live-born infants with aortic arch obstruction (AAO) born in Sweden 2014-2019, screened postnatally with perfusion index (PI) in addition to pulse oximetry (POS). As controls, 512 consecutive full-term newborns who passed the screening and remained healthy, were included. Optimal PI-values for cut-offs were assessed by Youden´s rule and evaluated by ROC-curve area under the curve (AUC).
Results: With conventional screening strategies 21/38 (55%) with AAO were discharged undiagnosed, including one in-hospital death. Median screening age in the AAO group was 10 (3-36) hours. Four of 38 (11%) had a positive POS, 3/38 (8%) had PI-value <0.7% ($p=0.04$ versus controls), 22/38 (58%) had a PI-value >3% in right hand ($p<0.0001$ versus controls), and 15/38 (39%) had a difference of >2% between right hand and foot PI values ($p=0.003$). Fourteen of 38 (37%) cases had a positive neonatal physical examination (NPE). Maximal screening sensitivity was obtained by using PI value in right hand >3% in combination with POS and NPE: sensitivity was 76% and specificity 85% and AUC 0.81 (0.73-0.89) $p<0.0001$. POS and NPE had a sensitivity of 45% and identified a significantly lower proportion of AAO-patients ($p=0.009$).
Conclusions: Although current contribution of PI in neonatal screening in addition to POS is limited, the sensitivity can be substantially increased by using a combination of criteria; a PI-value >3% in the right hand in combination to POS and NPE. Future prospective studies should investigate to what extent false-positive rate could be brought down when repeated PI-tests are conducted. Utilizing additional PI-criteria could significantly reduce the proportion of newborns with AAO being discharged undiagnosed, and thereby reducing long-term negative health outcomes for affected neonates.

Keywords: aortic coarctation, perfusion index, neonatal screening, early detection

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Table 1

	AUC (95% CI)	P-value	Sensitivity	Specificity
POS + NPE	0.72 (0.61-0.82)	<0.0001	17/38 (45%)	505/512 (99%)
PI >3% right hand	0.72 (0.62-0.82)	<0.0001	22/38 (58%)	441/512 (86%)
PI hand and foot diff >2% or >-2%	0.63 (0.53-0.74)	0.006	14/38 (37%)	460/512 (90%)
PI <0.7 %	0.53 (0.43-0.63)	0.57	3/38 (8%)	500/512 (98%)
POS + NPE + PI >3% + PI hand and foot diff >2% or >-2%	0.79 (0.72-0.87)	<0.0001	30/38 (79%)	399/512 (78%)
POS + NPE + PI >3% right hand	0.81 (0.73-0.89)	<0.0001	29/38 (76%)	435/512 (85%)

Table 1. Comparing different cut-off criteria to discriminate between the AAO-group and the control group. Results given as AUC (95% Confidence Interval), sensitivity and specificity (Unpublished data)
AUC: area under the curve, POS: pulse oximetry screening, NPE: neonatal physical examination, PI: perfusion index

Table showing different combination criteria in screening

PP-250

Exercise testing in clinical context: reference ranges for interpreting anaerobic threshold as an outcome for congenital heart disease patients

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Background and Aim: Change in the oxygen consumption at the ventilatory anaerobic threshold (VO2 at VAT) is an important outcome in response to treatment or interventions for patients with congenital and pediatric acquired heart disease (CHD). For context when assessing a measured change in VAT in clinical studies, the baseline range of values for any given VAT reported by different raters (similar to error of the measurement) is needed. The distribution of reported values in VAT for reference had not been reported.
Method: Sixty maximal cardiopulmonary exercise tests (CPET) for patients with CHD aged 8-21 years old were independently reviewed by six exercise physiologists and four pediatric cardiologists. For each of the 60 unique pairs of raters, the absolute difference in VO2 at VAT was calculated. Since the true physiologic VAT is not known, error of the measurement cannot be determined but differences between pairs can be displayed to demonstrate the distribution of inter-rater variability. Distribution percentiles of the differences were calculated. This method was repeated for subgroups of test modality (cycle/treadmill), patient factors (diagnoses, exercise capacity), and rater factors (cardiologist/physiologist, years of experience).
Results: Rater agreement was good with an intraclass correlation coefficient of 0.77-0.89 but the distribution of differences was broad. The median difference, or 50th percentile, was 2.7% of predicted peak VO2 (60 mL/min, 1.0 mL/kg/min), the 75th percentile was 6.4% predicted (140 mL/min, 2.5 mL/kg/min), and the 95th percentile was 16.3% (421 mL/min, 6.5 mL/kg/min) (Table 1, Figure 1). Distributions were similar for CPET modality and years of experience of rater, but differed for other factors.
Conclusions: The baseline distribution of reported VAT was relatively broad and varied by reported units (mL/min, mL/kg/min, %VO2). This variability was not explained by rater experience or differences in exercise test modality, but did vary by patient factors. When evaluating clinical relevance, a change in the VO2 at VAT in response to treatment of <6.5% (140 mL/min, 2.5 ml/kg/min) would fall within the majority (75th percentile) of expected variability and should be interpreted with caution.

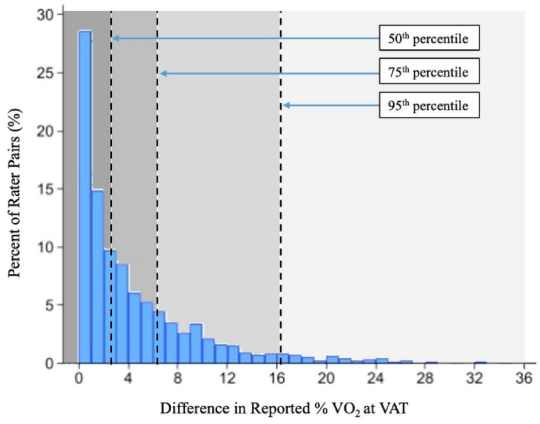
Keywords: Anaerobic threshold, inter-rater variability, reference range, cardiopulmonary exercise test, congenital heart disease

Table 1 & Figure 1

Table 1: Percentiles and values for the differences in reported VAT

Percentile	VO_2 at VAT (mL/min)	VO_2 at VAT by weight (mL/kg/min)	%VO_2 at VAT (%)
50th	60	1.0	2.7
55th	74	1.2	3.1
60th	88	1.5	3.7
65th	102	1.7	4.4
70th	118	2.1	5.4
75th	140	2.5	6.4
80th	185	2.9	7.7
85th	238	3.6	9.5
90th	301	4.5	11.2
95th	421	6.5	16.3
99th	744	10.6	24.7
Intra-class correlation coefficient	0.91	0.87	0.79

Figure 1: Distribution of the range of reported differences in percent of predicted VO_2 at VAT



PP-251

Does high intrathoracic pressure impact lymphatic flow? – a study of lymphatic contractility during positive pressure ventilation (LYMPH-UP)

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Background and Aim: The lymphatic system plays an important role in immune response, uptake of dietary fats, and maintaining fluid homeostasis. Most of the lymphatic fluid returning from the body runs through the thoracic cavity via the thoracic duct, before draining into the subclavian vein. This transport is unidirectional and dependent on contraction forces of peripheral and central lymphatic vessels, venous pressure at the thoracic duct outlet, and pressure in the thoracic cavity. Positive pressure ventilation (PPV) increases central venous pressure, reduces venous return, and affects the pressure in the thoracic duct. However, it is not known if positive pressure ventilation also affects the peripheral lymphatic vessels and thus the drainage of lymphatic fluid. We

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aim to investigate the impact of positive pressure ventilation on peripheral lymphatic contractility.
Method: Inclusion of 20 cardiopulmonary healthy patients 18 ≤ 35 years old undergoing surgery for malformations in the jaw, requiring general anesthesia and mechanical ventilation. Lymphatic contractility is investigated using Near-Infrared Fluorescence Imaging of the lower limb to determine lymphatic contraction frequency and velocity. Investigations are performed for 1) 30 minutes at standard ventilation pressure (PEEP 4-7 cmH2O), 2) 30 minutes at elevated ventilation pressures (PEEP 10 cmH2O > standard), 3) during breath-hold, 4) immediate post-op, and 5) during spontaneous respiration (baseline).
Results: Data collection and analysis is ongoing. Inclusion is expected completed by January 2024. Preliminary results from the first 11 patients show a median lymphatic contraction frequency at baseline of 1.8 (IQR 1. 3 - 2.5) contractions min-1. Standard pressure PPV decreased contraction frequency with 61% (median 0.7 (IQR 0.3 - 1.0) remaining here at elevated pressure ventilation (median 0.5 (IQR 0.3 - 1.1) and breath-hold (median 0.7 (IQR 0.0 - 0.8)). Immediate post-op median frequency was 0.8 (IQR 0.5 - 1.4) contractions min-1.
Conclusions: These preliminary results suggest that peripheral lymphatic contractility is affected by PPV. If so, it could instigate new initiatives to reduce fluid retention after prolonged exposures to PPV in e.g. intensive care patients, as well as help improve our understanding of especially patients with a Fontan circulation who either have or are at risk of developing lymphatic complications.

Keywords: lymphatic function, lymphatic system, positive pressure ventilation, mechanical ventilation, lymphedema

PP-252

The effects of physical activity levels on body composition and exercise performance in children and adolescents with a fontan circulation

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Background and Aim: Physical active lifestyles are important for children and adolescents living with congenital heart disease. This study aims to determine the effect of physical activity levels on the body composition and exercise capacity in children and adolescents living with a Fontan circulation.
Method: Thirty children and adolescents with a Fontan circulation, were prospectively recruited in this case control study. Mean age was 14.8±2 years, at mean 11.8±2 years after Fontan operation. The participants underwent cardiopulmonary exercise testing and bioimpedance analysis. Physical activity level was assessed using accelerometer (Fitbit Inspire 3 wristwatch), patients with daily step counts over 10000 considered active Fontan and under 10000 daily steps are considered less active Fontan.
Results: See Table 1 for summary of results.
43% (13 of 30) of cohort were considered active Fontan, predominantly male at a younger age (mean age of 13.9±1.7 vs 15.4±2.2 years). The overall body mass index (BMI) was within normal limit, however 17% (5 of 30) of the Fontan cohort were overweight or obese (BMI >85th centile). Both groups had normal height for age, with lower BMI in the active Fontan group.
The less active Fontan group was associated with higher percentage fat mass (22.6±8.7 vs 16.3±4.9%), lower percentage appendicular skeletal muscle mass, SMMa (30.9±4.2 vs 33.6±2.6%; z score -0.21±0.6 vs 0.15±0.7), and muscle to fat ratio (1.7±0.9 vs 2.3±0.9). The less active group had lower peak oxygen consumption, VO2peak (26.5±7.2mL/kg/min at 66.6±16.1% predicted vs 35.5±3.4 mL/kg/min at 80.6±9.5% predicted), lower peak oxygen pulse (76.2±13.7 vs 78.3±9.9%). They achieved lower work rate (100±43.8 watts at 57±18.3%predicted vs 120 ±39.4 watts at 71±5.14.3% predicted) and peak heart rate (92±8.2 vs 97±5.2% predicted) when compared with the active Fontan group.
Conclusions: Higher physical activity levels (>10000 steps daily) in children and adolescents with Fontan circulation are associated with normal skeletal muscle mass and higher exercise capacity, conversely lower physical activity levels are associated with skeletal muscle mass deficit, increased adiposity and reduced exercise capacity.

Keywords: Fontan; CHD, Physical activity; VO2peak; Bio-impedance-analysis.

Table 1 Summary of Results

Table 1: Summary of results on the Effects of Physical Activity levels to Body Composition and Exercise performance in children and adolescents with a Fontan circulation, values in mean±SD.

Variables	All patients	Active Fontan >10000 steps/ day	Less Active Fontan <10000 steps/ day
Number of patients	30 (19 male)	13 (11 male)	17 (8 male)
Age, years	14.8±2.1	13.9±1.7	15.4±2.2
Height, cm (z score)	163±11.7 (0.03±0.9)	162±11.6 (0.03±1.2)	164±12.2 (0.03±0.7)
Weight, kg (z score)	54±14.7 (-0.07±1.1)	48.6±11.6 (-0.4±1.1)	58.1±15.8 (0.2±1)
BMI (z score)	20±3.7 (-0.18±1)	18.3±2.6 (-0.56±1)	21.3±3.9 (0.12±1)
Number of patients with normal BMI >5th & < 85th centile	25	11	14
Number of patients with BMI >85th centile (Overweight)	4	2	2
Number of patients with BMI >95th centile (Obese)	1	0	1
Body Composition			
Appendicular Skeletal Muscle Mass (SMMa), kg	17.2±4.8	16.4±4.2	17.9±5.2
Percentage SMMa, % (z score)	32.1±3.8 (-0.05±0.7)	33.6±2.6 (0.15±0.7)	30.9±4.2 (-0.21±0.6)
Fat mass (FM), kg	11.3±7.1	8.2±3.9	13.7±8
Percentage FM, %	19.9±7.9	16.3±4.9	22.6±8.7
Number of patients with:			
Normal FM%, >2 & <85th centile	20	9	11
Overweight/ Obese FM% ≥85th centile	7	2	5
Underweight FM% centile ≤ 2nd centile	3	2	1
Muscle to Fat ratio (SMMa/FM)	1.9±1	2.3±0.9	1.7±0.9
CPET measurements			
Peak oxygen consumption, VO2peak, mL/min/kg	30.4±7.3	35.5±3.4	26.5±7.2
Percent predicted VO2peak, %	72.7±15.2	80.6±9.5	66.6±16.1
Peak Work rate (WR), watt	109±42	120±39.4	100±43.8
Percent predicted Peak WR, %	63±17.8	71±5.14.3	57±18.3
Peak Heart rate (HR), bpm	177±14.2	183±10.2	172±15.4
Percent predicted peak HR, %	94±7.5	97±5.2	92±8.2
Peak oxygen pulse, ml/beat	9.2±2.5	9.6±2.5	8.9±2.6
Percent predicted oxygen pulse, %	77.1±12	78.3±9.9	76.2±13.7
VO2 at VT1/AT, mL/min	1040.6±248.9	1125.3±298.1	975.9±188
Percent predicted VO2 at VT1/AT, %	47.3±9.9	52±6.4	43.7±10.9
VE/VCO2	37.1±6.2	37.93±7.1	36.5±5.8

PP-253

Rare case presentation of familial thoracic aortic aneurysm and aortic dissection in adolescent – case report

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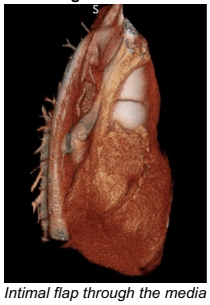
Background and Aim: Familial TAAD (fTAAD) is a genetic disease encoding the major proteins in the smooth muscle (SMC) contractile filaments, of alpha-actin "ACTA2" and myosin heavy chain "MYH11", in an autosomal dominant pattern. "MYH11" mutation is a rare cause of fTAAD and is identified primarily in families with non-syndromic TAAD, inherited in association with patents ductus arteriosus (PDA).
Method: We present very rare case report of a 17-year-old girl with TAAD of ascending aorta and "MYH11" gene mutation confirmed respectively.
Results: At the first year of age, the patient underwent PDA interventional closure. Due to suspicion of coarctation recatheterisation was performed at the age of 4, without confirmation. She was followed up by a pediatric cardiologist and reffered to the consultation because of non-specific precordial feelings. Physical examination showed no sign of connective tissue abnormalities. Transthoracic echocardiography revealed severe dilation of ascending aorta (Z sc +5.3) with new onset of aortic regurgitation, and dissection of ascending aorta was suspected. CTA was performed and confirmed the diagnosis of Standford A dissection with the entry at the level of sino-tubular junction and re-entry at the level of ascending aorta before epiaortic branches of the proximal aortic arch. The patient underwent surgical procedure with resection of the damaged part of aorta ascendens and implantation of tube graft (Vascutek 24 mm). Genetic analysis focused on genes associated with vasculopathies and connective tissue abnormalities was performed and patogenous variant of 4599+1G>A "MYH11" gene mutation was confirmed (Twist Human Core Exome kit analysis). There was no history of sudden cardiac death in the family nor other cardiovascular diseases. The parents and the younger brother underwent genetic testing, and other members of the family are planned to be tested. We have to focus on this mutation result in various aortic and occlusive vascular pathology in our patient in the future.
Conclusions: Early presentation of ascending aorta aneurysm and dissection confirmed "MYH11" mutation in non-syndromic TAAD/PDA phenotype patient. The risk of progressive character leads to intense follow-up of the patient, with possible repetitive imaging of other parts of the aorta, and magnetic resonance angiography of the cerebral arteries.

Keywords: Familial thoracic aortic aneurysm and aortic dissection, "MYH11" gene mutation

Computed tomography angiography in patient with aneurysm and dissection of ascending aorta



Intimal flap through the media and subadvential tissue results in the possibility of 3D model

PP-254

Pulmonary atresia with intact septum associated with ebstein's anomaly: Case report

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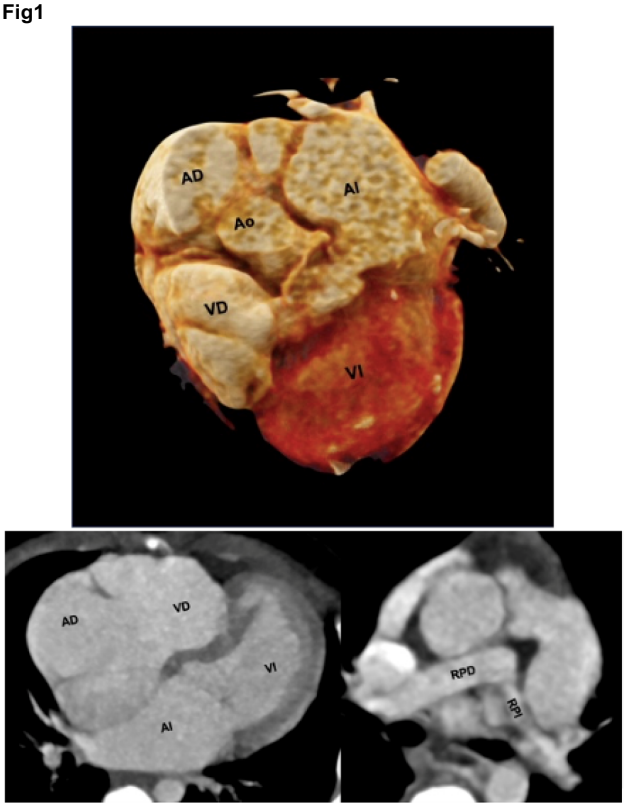
Background and Aim: Pulmonary atresia with intact septum is a rare heart disease, whose frequency is estimated at less than 1% in all congenital heart disease. It presents with a variable spectrum in its clinical presentation. The association with Ebstein's anomaly is rare. Ebstein's anomaly continues to be the first congenital cause of tricuspid regurgitation and its natural evolution conditions heart failure. The treatment will depend on its anatomical characteristics.
Method: Case report of a patient diagnosed with pulmonary atresia with intact septum and its association with Ebstein's anomaly.
Results: A 4-month-old female with cyanosis since birth, sent to our hospital for a diagnostic approach. On admission with respiratory instability. The echocardiogram reports pulmonary atresia with intact septum. Hypoplasia of the left branch of the pulmonary artery. The tricuspid valve with attachment areas that did not allow its morphological characterization. An elongated left ventricle (LV) was observed involving the apical portion of the right ventricle. Systolic and diastolic function preserved. Cardiac computed tomography angiography demonstrated septal and inferior leaflet attachment with fibrous insertions of the anterior leaflet towards the vestigial outflow tract, hypoplasia of the IPR, and LV elongation. Palliation with right pulmonary systemic fistula was decided to improve pulmonary flow and branch growth, with favorable evolution.

Fig1



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Conclusions: This case demonstrates two congenital heart conditions of very rare association. The goal of treatment will be aimed at increasing pulmonary blood flow and allowing the potential growth of the pulmonary artery branches. Multimodal evaluation in patients with complex congenital heart disease is necessary for optimal diagnosis.

Keywords: Ebstein's anomaly, Pulmonary atresia with intact septum, Case report.

Genetics, Basic Science and Myocardial Disease

PP-255
A race against time: A unique case of an embolised gigantic left ventricular myxoma in a pediatric patient

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Background and Aim: The presence of a cardiac tumor is a rare finding at echocardiography. While in adult patients myxomas represent the most common cardiac tumors, in pediatric patients these are very rare. They are mostly localised at an atrial level, with only a handful of cases of ventricular myxomas being reported in literature. That is why, we bring forward the unusual case of a 11-year-old female patient with multiple peripheral embolisations due to a highly unlikely ventricular tumor, with a unique echocardiographic appearance.
Method: Our patient referred to the emergency department due to acute pain and loss of sensation in her legs. From her medical history we found out that she had previously achieved remission from acute lymphoblastic leukemia. Upon physical examination, the patient presented pale-skin and cold lower extremities, with no peripheral pulse present, thus raising the suspicion of bilateral arterial occlusion and ischemia. An arterial doppler ultrasound was performed and confirmed this suspicion, as the right lower extremity suffered from total occlusion of the superficial femoral artery. Meanwhile, in the left lower extremity, the occlusion was localised in the proximal tibio-peroneal trunk. Cardiac sonography revealed a massive left ventricular intracavitary mass. Aside from its large dimensions (6.3 by 3 cm), its aspect was striking as well as it had very mobile and friable edges. Due to its superior extremity that was protruding through the aortic valve during systole, we suspected that a tumor fragment had embolised in the patient's lower limbs' vessels.
Results: Emergency bilateral lower limb endarterectomy and excision of the left ventricular tumor under cardio-pulmonary by-pass were performed alongside a systemic anticoagulant therapy, with excellent results, as no tumoral residual masses could be seen in the left ventricle and the arterial blood flow being restored completely in both lower extremities. The histopathological aspect of the excised masses confirmed a myxoma with multiple systemical emboli.
Conclusions: What started as a bilateral acute lower extremity ischemia, proved to be a systemic tumoral embolisation from a massive left ventricular myxoma. Tumors of this size, aspect and localisation in left ventricle are scarcely represented in literature, with only a handful of cases reported in children.

Keywords: myxoma, ischemia, tumoral embolus

PP-257
Cardiac abnormalities in sudden infant death syndrome

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Background and Aim: Sudden infant death syndrome (SIDS) is the leading cause of death in infants under one year of age in Western countries. The Triple Risk Model, introduced in 1994, proposes acknowledging SIDS as a multifactorial syndrome, suggesting genetic predisposition or structural pathologies, in combination with environmental triggers in a vulnerable infant, as the combined cause of these fatal events. Factors such as co-sleeping, prone sleeping and maternal smoking have been established as precursors for SIDS. Newer studies have shown an association between dysfunctions of the cardiac conduction system and SIDS, suggesting cardiac pathology as an important risk factor. This review aims to analyze, compare, and present the most recent research on SIDS and its association with heart disease.
Method: We have performed a review of the published literature concerning SIDS and cardiac disease, with literature searches in PubMed and SCOPUS. We followed the Preferred Reporting Items for Systematic Reviews and Meta-analyses (PRISMA) guidelines. Full-text articles have been evaluated according to strict inclusion and exclusion criteria by two independent reviewers, and any discrepancies have been resolved through discussion and consensus.
Results: Preliminary results indicate cardiac channelopathies, in cardiac sodium channels, to account for a large portion of cases. Data extraction from the included studies is currently ongoing and is expected to be ready for analysis during December of 2023.
Conclusions: Understanding the causes of SIDS is an important step towards reducing the events. This systematic review combining data from population-based cohort studies, will summarize the most significant genetic mutations and structural risk factors in SIDS.

Keywords: sudden infant death, cardiac abnormalities, crib death, congenital heart disease, heart defects

PP-258
TTN mutation-related supraventricular rhythm disorders without cardiomyopathy – a rare entity

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Background and Aim: We present the case of a 16 years old enduro motorcyclist with symptomatic arrhythmias who was found to have a titin mutation described as likely pathogenic. Titin mutations have been documented to be responsible for several cardiac pathologies such as dilated cardiomyopathy and they are a rare cause of hypertrophic cardiomyopathy and of arrhythmogenic right ventricular dysplasia.
Method: Multiple paraclinical diagnostic tools (imagistic, electrocardiogram, genetic testing) were performed in order to establish the therapeutic strategy.
Results: Our patient was diagnosed by continuous ECG monitoring with supraventricular rhythm disorders (atrial fibrillation with variable ventricular rate, typical atrial flutter, atrial ectopic tachycardia, frequent premature supraventricular complexes) and premature ventricular extrasystoles (couplets, bigeminy, trigeminy). Clinically he reported palpitations, dizziness and nausea at school, symptoms that were correlated with atrial fibrillation with a 172 beats/minute ventricular rate on the ECG monitoring. The echocardiographic evaluation was in normal range. Subsequently we performed a cardiac magnetic resonance (CMR) imaging showing no pathologic findings suggestive for a cardiomyopathy. Furthermore, the comprehensive genetic panel identified a heterozygous frameshift variant TTN c.96044del, p.(Glu32015Glyfs*6). This is predicted to lead to a loss of normal protein function, either through protein truncation or nonsense-mediated mRNA decay. Loss of function is an established disease-mechanism in this gene. It is classified as likely pathogenic, based on the variant's rarity in control populations and variant type. The patient underwent a successful percutaneous ablation with radiofrequency for the recurrent symptomatic paroxysmal atrial fibrillation and flutter, becoming asymptomatic. Although sudden death in childhood is rare, it remains the main concern in this case. Further ECG monitoring, echocardiography, cardiac magnetic resonance and close clinical monitoring are necessary.
Conclusions: It is difficult to estimate the individual contribution of this genetic mutation in the development of possible future malignant arrhythmias, especially taking into account that our patient decided to continue practicing enduro motorcycling. Titin truncating variants are a known cause of dilated cardiomyopathy, thus existing studies have focused on the effects of these variants in ventricular cardiomyocytes. The structural and functional consequences of titin truncating variants in atrial cardiomyocytes and how such variants lead to supraventricular arrhythmias are unclear, especially in pediatric patients.

Keywords: titin, arrhythmia, atrial fibrillation, flutter

PP-259
Rasopathies and cardiac manifestations: The diversity of phenotype and genotype

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Background and Aim: RASopathies are a group of multisystemic genetic disorders affecting the Ras/MAPK signaling pathway including frequently cardiac phenotype. Our aim is to describe the prevalence and variety of cardiac manifestations within RASopathies.
Method: Retrospective review of pediatric-aged patients diagnosed with RASopathies in our center.
Results: The study included 18 patients (61% females) with RASopathies: 14 with Noonan Syndrome (NS) with molecular confirmation (seven had variants in *PTPN11* gene, two in *SOS1*, two in *RAF1*, one in *MAPK1*, one in *LZTR1*, and one in *RIT1*), one patient with NS clinical diagnosis, and three patients with Cardiofaciocutaneous (CFC) Syndrome with molecular confirmation (pathogenic variant in *BRAF*). In 5 cases, one parent was diagnosed with NS subsequently. Medium age for diagnosis in this cohort was 3 years (ranging from pre-natal diagnosis to 12 years). Patients maintained cardiological follow up for an average of 4 years. Regarding prenatal history, two had increased nuchal translucency and one had suspected Dandy-Walker malformation. The latter had NS family history and was diagnosed during pregnancy. In this cohort, 13 patients had cardiac anomalies, the most common being pulmonary valvar stenosis (PVS), presenting in 3 cases as an isolated finding and associated with: interventricular septum hypertrophy (ISH) in 2 cases, hypertrophic cardiomyopathy (HCM) in 1 case, and left ventricular hypertrophy and atrial septal defect (ASD) in 1 case. Two patients presented with isolated severe HCM (one in the neonatal period with a fatal outcome), 2 patients with isolated ISH, 1 with ISH and mitral valve dysplasia, 1 with ASD, and 1 patient with Wolff-Parkinson-White Syndrome and mitral anomaly. Two patients underwent cardiac surgery: ASD closure and pulmonary valvotomy; ASD closure and septal myomectomy.
Conclusions: In our sample, 76% of patients had a cardiac phenotype, displaying that NS has incomplete penetrance for cardiac features. This percentage will vary among different RASopathies. While sample size does not allow for definite phenotype-genotype correlations, it is noteworthy that the two cases with severe HCM had pathogenic *RAF1* variants. Further investigation is needed to a more accurate picture of the cardiologic landscape in RASopathies and the development of new therapeutics for these debilitating disorders.

Keywords: RASopathies, cardio genetics, Noonan Syndrome

PP-260
Danon disease - first family reported in slovakia

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Background and Aim: Danon disease is a rare Xlinked genetic storage disorder caused by mutations of Xq24 gene encoding lysosomal membrane protein 2 (LAMP 2) that is responsible for autophagic processes in the cell. The deficiency or complete absence of LAMP 2 results in accumulation of glycogen and autophagic content in huge vacuoles of cells. Phenotypic expression is

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heterogenous – hypertrophic or dilated cardiomyopathy, skeletal myopathy, hepatopathy, mental retardation, retinopathy,
Method: We diagnosed Danon disase in 5 members of one family – 3 males and 2 females, mother and her four children. This is the first family with Danon disease reported in our country.
Results: Two asymptomatic brothers, aged 11 and 8, came to cardiologic examination because of positive family history. Their initial findings were similar - left ventricular hypertrophy on electrocardiography, extreme hypertrophy of the left ventricle with maximum thicknes of interventricular septum more than 40mm and extreme obstruction of left ventricular outflow tract on echocardiography. Other examinations revealed hepatopathy, only light skeletal myopathy and light mental retardation. Both brothers underwent implantation of dual chamber cardioverter defibrillator as a primary prevention of sudden death. We recommended genetic testing of the family – 8 children and their mother . . . Danon disease war confirmed in 3 sons, 1 girl and their mother, 2 girls had negative genotype and the youngest boy was not tested. They all had the same mutation in the LAMP 2 gene c.864+3-687+6delGAGT. Both our brothers despite the therapy died of congestive heart failure at the age of 14 years, their older brother died suddenly.Their mother died also of heart failure later. The last member of this family with positive genotype of Danon disease is 17 years old asymptomatic girl with borderline hypertrophic cardiomyopathy.
Conclusions: Cardiac involvement in Danon disease is variable, but together with an early diagnosis, crucial for the prognosis. Males are affected in an earlier age and more seriously than females and usualy have also extracardiac signs.The only effective treatment is ICD implantation and heart transplantation, in the future also gene therapy, Testing of all family members and genetic counselling are necessary.

Keywords: Danon, cardiomyopathy, genetic testing

PP-261

Rasopathies - cardiac manifestations and outcomes in a tertiary center cohort study

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Background and Aim: RASopathies are a clinical and genetic spectrum of disorders defined by germline mutations in components or regulators of the RAS/MAPK pathway. This work aimed to provide a comprehensive assessment of cardiac manifestations, morbidity, and mortality in a population of individuals with RASopathies in pediatric age.
Method: Retrospective analysis conducted on medical records of patients with genetic and/or clinical diagnosis of RASopathies who received care at a Pediatric Cardiology tertiary center over a period of 40 years (1982-2022).
Results: Thirty-nine patients were included, the majority being male (64%). Noonan Syndrome (NS) was identified in 27 patients (69,2%), NS Multiple Lentigines in 4, Cardiofaciocutaneous syndrome in 2, Costello Syndrome and Noonan syndrome-like disorder with loose anagen hair 1 were present in 1 patient each. Four patients had overlap syndrome between Type 1 neurofibromatosis and NS. Pathogenic variants in the PTPN11 gene were the most frequent (17,9%). The median age at the initial evaluation was 3 years (4 days - 18 years), and the median follow-up duration was 14 years. Cardiac involvement was present in 84,6% of patients: the most common was pulmonary valve (PV) stenosis (59%), followed by hypertrophic cardiomyopathy (28,2%) and atrial septal defect (25,6%). PV stenosis was the most frequent cardiac lesion in NS while hypertrophic cardiomyopathy was more prevalent in the other groups. During follow-up, 36% underwent cardiac surgery (48% of the procedures included pulmonary valvuloplasty; 19% left ventricular outflow tract relief); 26% underwent interventional cardiac catheterization (9 out 10 for pulmonary valvuloplasty). Two patients died: one at 8 months, with PV stenosis, biventricular hypertrophy, due to multi-organ failure after pulmonary valvuloplasty and Morrow myomectomy; the second with moderate PV stenosis, at 10 months, due to respiratory insufficiency within the context of a polymalformative syndrome.
Conclusions: RASopathies have an important prevalence of cardiac disease with a spectrum that extends from mild to severe and potentially fatal disease. Overall, this study highlights the high prevalence of cardiac involvement in these patients with PV stenosis being the most common form. Due to the heterogeneity of clinical presentation and incomplete penetrance of phenotypes, genetic testing and careful long-term follow-up is essential.

Keywords: RASopathies, Cardiomyopathy, Pulmonary Valve Stenosis, Genetic Variants

PP-262

Treatment with rapamycin for progressive hypertrophic cardiomyopathy in infant with noonan syndrome with multiple lentigines. 2-years follow-up

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Background and Aim: Noonan syndrome with multiple lentigines (NSML) is frequently associated with hypertrophic cardiomyopathy (HCM) with severe left ventricle outflow tract (LVOT) obstruction resistant to conventional treatment. Moreover, the concomitant occurrence of HCM and congenital heart defect in patients with RASopathies has previously been reported as associated with worse clinical outcome. Recently, it was demonstrated that mTOR inhibitors may reverse or slow the progression of cardiac hypertrophy. Therefore, we present our two-years long experience with rapamycin therapy in 2.5 years old child with NSML and HCM with LVOTO accompanied by severe pulmonary stenosis.
Method: The patient was diagnosed with HCM prenatally and in the first days of life conventional treatment with propranolol was introduced. Rapid progression of symptomatic hypertrophy in echocardiography was observed (IVSd 12 mm z-score:+12.24, LVPWd 11 mm z-score:+13.52). Progressive coexisting pulmonary stenosis (RV-MPA systolic pressure gradient max 79 mmHg, mean 42 mmHg) forced balloon pulmonary valvuloplasty at the age of 1 month resulted with temporal reduction of gradient to max 50 mmHg, mean 25 mmHg. At the age of six months, the diagnosis of NSML was established by genetic testing showing a heterozygous mutation in the PTPN11 gene. At the age of 6.5 months, because of clinical status worsening, increasing LVOT and RVOT pressure gradients (max 70 mmHg, max 95 mmHg respectively) and NT-proBNP levels exceeding 80,000 pg/ml, rapamycin (sirolimus) therapy was started. 3 months later disopyramide was added.

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Results: Heart failure improved from NYHA stage IV to II and NT-proBNP values decreased gradually to 4,000 pg/ml. Echocardiography showed decrease of IVSd 10 mm z-score: +7.45, LVPWd 8 mm z-score:+4.95, and stabilization of max LVOT pressure gradient 80 mmHg, max RVOT pressure gradient 95 mmHg. No hypertrophy progression over 23 months was observed. Side effects included hypercholesterolemia and endometrial hyperplasia.
Conclusions: Our findings show that rapamycin treatment could be beneficial for neonates and infants with NSML and stop progression of HCM even when complete reversal of hypertrophy may not be possible. The prolonged therapy seems to be safe. Further studies, starting therapy from neonatal or even foetal period would be of a great value.

Keywords: Noonan Syndrome, Hypertrophic Cardiomyopathy, Rapamycin, RASopathy, PTPN11

PP-264

Multidisciplinary management in vascular ehlers-danlos syndrome: Cardiovascular and multisystemic features in a pediatric cohort

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Background and Aim: Vascular Ehlers-Danlos Syndrome (VEDS) is a rare autosomal dominant (AD) connective tissue disease (CTD) due to likely pathogenic o pathogenic variant in COL3A1 and characterized by variable multiorgan involvement. The main signs are generally cardiovascular with possible progressive dilation and rupture of all vascular districts, in particular aortic and cerebral. Other possible manifestations include gastrointestinal perforation and uterine rupture during pregnancy in women. Potentially any other organ or system with connective tissue background can be affected. This disorder has typically an age-related penetrance. The pediatric population of VEDS undoubtedly represents a real challenge for clinicians not only in terms of diagnosis but also in terms of management. In fact, few studies in literature are available regarding multisystemic involvement of VEDS in children.
Method: Our tertiary academic paediatric represents the referral point for the center and south of Italy for the multidisciplinary management of patients with CTDs. In this study we include data from 10 patients with VEDS who had multisystemic clinical assessment, multiorgan screening and genetic study through (Next Generation Sequencing) sequencing.
Results: We focused on VEDS manifestations in children, reporting the main multisystemic signs and symptoms to be considered at diagnosis and during follow up and considering data related to clinical and instrumental screening and surveillance (electrocardiogram, echocardiography, contrast-enhanced magnetic resonance angiography, spine radiography, dual-energy X-ray absorptiometry, etc.)
Conclusions: In the management of VEDS pediatric population it is important to consider not only the major signs classically associated with the disease such as cardiovascular but also the minor aspects that can have a significant progressive impact on the health in children. We emphasize the importance of the multidisciplinary, personalized, and longitudinal in time approach to children with suspected or confirmed VEDS. Early VEDS diagnosis at multisystemic level can prevent acute and chronic complications, offer tailored and improve the quality of life.

Keywords: connective tissue disease, Vascular Ehlers Danlos Syndrome, progressive multiorgan involvement, pediatric multidisciplinary management, personalized approach

PP-265

Navigating complexity: a comprehensive analysis of arrhythmogenic cardiomyopathy in a 16-year-old adolescent

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Background and Aim: Arrhythmogenic cardiomyopathy (ACM) is a rare cardiac condition marked by fibrous or fibro-fatty infiltration of the myocardium, resulting in arrhythmias and progressive cardiac dysfunction. Pediatric prevalence remains uncertain due to limited reported cases. Initially perceived as exclusively impacting the right ventricle (RV), recent studies revealed the involvement of the left ventricle (LV), leading to the replacement of the term "arrhythmogenic right ventricular dysplasia" with ACM. Arrhythmogenic cardiomyopathy often manifesting in the second to fourth decades of life, poses a risk of sudden cardiac death, emphasizing the importance of risk stratification for implantable cardioverter-defibrillator (ICD) implantation.
Method: We present a case of a 16-year-old teenager who was admitted to the hospital following a syncopal episode accompanied by clonic seizures on the right side of the body. The patient exhibited gradually progressive exercise intolerance subsequent to a respiratory infection two months prior to hospitalization. Diagnostic assessments revealed dilated cardiac cavities with severe biventricular systolic dysfunction. The surface electrocardiogram depicted inverted T waves in V1-V4 precordial leads. Heart failure treatment was initiated, associated with inotropic treatment with Dobutamine, with a slightly favourable evolution.
Results: We performed several tests in order to determine the cardiomyopathy aetiology. Holter electrocardiogram monitoring

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revealed ventricular extrasystoles with left bundle branch aspect and episodes of non-sustained ventricular tachycardia. Magnetic resonance imaging identified a dilated RV with accentuated trabeculae in the medio-apical segment with no fibrotic lesions and severely compromised systolic function of LV with basal subendocardial fibrotic lesions. Genetic testing further revealed a heterozygous desmocollin mutation. Considering the diagnosis of ACM with biventricular involvement, the patient underwent implantation of a primary prophylactic ICD.
Conclusions: The presented case underscores the complexities associated with the diagnosis and management of this rare cardiac condition. The decision to implant a primary prophylactic ICD reflects the imperative nature of risk stratification in preventing potential life-threatening arrhythmias and sudden cardiac death. ACM in pediatric patients, while presenting with recurrent myocarditis-like episodes, demands adapted diagnostic criteria due to unique manifestations. Acknowledgement: "This work was supported by the University of Medicine, Pharmacy, Science and Technology "George Emil Palade" of Târgu Mureş, Research Grant number 511/5/ 17.01.2022"

Keywords: arrhythmogenic cardiomyopathy, pediatric cardiomyopathy, implantable cardioverter-defibrillator

PP-266
Role of immunostaining methods in the diagnosis of mitochondrial cardiomyopathy

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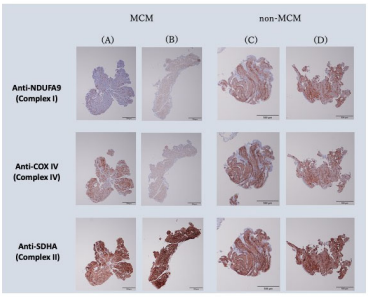
Background and Aim: Mitochondrial cardiomyopathy (MCM) is a complex, heterogeneous condition that may affect infants, children, and adults and is frequently overlooked (E.Braunwald, EHJ 2023). This cardiomyopathy is diagnosed by pathology, genetics, and biochemistry, but until now the pathological approach has only been suspected from electron microscopy. We have developed a pathological diagnosis of MCM using immunostaining histochemistry (IHC) with respiratory chain enzyme antibodies (A.Takeda, J Clin Path 2020). In this report, we describe a case of MCM diagnosed pathologically by this method and discuss this disease's genetics and clinical aspects.
Method: Of the 23 cases diagnosed with mitochondrial cardiomyopathy by IHC, 13 cases in which either genetic testing, tissue biochemistry, or EM could be performed were retrospectively reviewed. To investigate the correlation between respiratory chain complex enzyme activity and immunohistochemistry, 13 patients (7 with mitochondrial cardiomyopathy, the rest with other cardiomyopathies) were examined in myocardium where both were available for testing. For IHC, anti-NDUFA9 for complex I, anti-SDHA for complex II, and anti-COX IV for complex IV were used. DAB-positive areas were measured. The staining area of complexes I and IV was expressed as a ratio of complex II; thus, Area (Complex I/II) and Area (Complex IV/ II) were calculated. Data are expressed as a ratio of the area of positive staining for the specific complex antibody compared with the control area. Those of myocardium with less than 30% of normal control was defined as negative staining with pathologically diagnosed MCM.
Results: Three were negative staining for Complex I only, 3 were negative for Complex IV only, and the remaining 7 had negative staining for both Complex I and Complex IV. Seven patients performed respiratory chain complex enzymatic assay and 5 patients were found to be defective. The results of the enzymatic assay and IHC were significantly correlated.Ten patients had genetic examination and all the patients had pathogenic genetic variants of MCM. Five patients had m.3243A>G and the other had m.4300A>G, m.14453A>G, m.12264C>T, MT-ND6, GARS, GTPBP3, and TOP3A. All the patients had abnormal mitochondrial findings in EM.
Conclusions: IHC is an excellent method of pathologically confirming the diagnosis of MCM.

Keywords: mitochondrial cardiomyopathy, immunostaining methods

Figure



Immunohistopathological study on biopsied myocardium in patients with mitochondrial cardiomyopathy(MCM) and non-mitochondrial cardiomyopathy(non- MCM) (A) Signal intensity of complex I compared with complex II was low (16% of normal) (B) Both complex I and complex IV compared with complex II were low (4%, 0% of normal, respectively) (C) Signal intensity of both complex I and complex IV compared with complex II was normal (99%, 58% of normal, respectively) (D) Signal intensity of both complex I and complex IV compared with complex II was normal (140%, 43% of normal, respectively).

PP-267
Atypical case of hypertrophic cardiomyopathy with sever right ventricle outflow tract obstruction in a 14 years old patient

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Background and Aim: Hypertrophic cardiomyopathy(HCM) is a primary genetic disease of the myocardium, characterized by asymmetric hypertrophy of the interventricular septum(IVS), most frequently associated with dynamic obstruction in the ejection pathway of the left ventricle(LVOT) and with different degrees of mitral regurgitation(MR). It is *rare* in the pediatric population, unlike the adult population, with a prevalence estimated at 1.2 per 1,000,000 and with an incidence of 1.3 per 100,000.
Method: We present the case of a 14-year-old female patient, who presented for dyspnea on moderate exertion. Echocardiography revealed: severe biventricular HCM, predominantly septal (basal

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IVS=21 mm, medioventricular IVS = 34 mm, basal LV posterior wall(LVPW) = 12.7 mm, Medioventricular LVPW = 20 mm, free wall of the right ventricle (RV) = 8 mm), with significant obstruction in the RV outflow tract (RVOT), maximum gradient = 74 mmHg, *without obstruction in LVOT*; moderate LV systolic dysfunction, mild RV systolic dysfunction, biatrial dilatation and grade II MR. ECG: sinus rhythm, ventricular rate = 75 bpm, intermediate QRS axis, wide QRS complexes (0.14 sec), hypervoltage, left bundle branch block-like appearance, negative T waves and ST segment depressions in V3-V6. ECG holter: normal. Propranolol treatment was initiated, 10mgX3/day, with good tolerance. Genetic testing: heterozygous mutations in the MYH7 and ALPK3 genes.
Results: Periodic evaluations: aggravation of the signs of heart failure (fatigue at low efforts) was found, and echocardiographically, mild increase of RV dysfunction and an RVOT gradient of 80 mmHg, promoted the decision to perform the surgery approximately one year after the diagnosis. The intervention involved infundibulectomy, resection of a septal band and RVOT plasty. Postoperative echocardiography: moderate biventricular systolic dysfunction, no RVOT obstruction. In the cases reported so far, the *ALPK3 heterozygous mutation* is usually associated with *HCM with onset in adulthood*, predominantly with IVS and apical hypertrophy, without LVOT obstruction. Although concomitant RV involvement is common (may occur in 28-44% of adult patients with HCM), significant RVOT obstruction due to a hypertrophied right ventricle is *very rare*. RV involvement is associated with an increase in the incidence of supraventricular and ventricular arrhythmias, with severe dyspnea and pulmonary thromboembolism.
Conclusions: The particularity of the CASE: association of HCM with severe RVOT obstruction in a child who required surgical intervention, without ventricular/supraventricular arrhythmias. Genetic testing revealed heterozygous mutations that are not usually involved in such cases.

Keywords: severe hypertrophic cardiomyopathy, right ventricular outflow tract obstruction, no left ventricle outflow tract obstruction, genetic mutations

PP-268
Development of atrial myocardial architecture: The question of homology of pectinate muscles and ventricular trabeculae

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Background and Aim: Ventricular trabeculae and pectinate muscles are morphological hallmarks of chamber myocardium differentiation. Here we sought to identify the factors governing pectinate muscles formation and probe to which extent they are homologous to trabeculae.
Method: Chick embryonic hearts between Hamburger-Hamilton stages 19-29 were studied by whole mount confocal microscopy and histology. Impulse propagation through the atria was monitored by optical mapping. Proliferative history of atrial myocardium was evaluated using label-dilution technique. Hemodynamic loading was modified using either left atrial ligation or isolated heart culture.
Results: Pectinate muscles start to form between stages 23-24. The ratio of pectinate muscles to free wall increased steadily between stages 24 and 29. Complete mechanical unloading in whole heart culture inhibited their formation, which was not rescued by injection of silicon oil droplet. Left atrial ligation caused slowing of the impulse propagation and induced ectopic pacemaking activity in the excluded portion of the atrium. Unlike ventricular trabeculae, which showed considerably slower proliferative activity than the compact myocardium, the proliferative structure of the atrium was homogeneous.
Conclusions: Thus, while there is some structural and functional homology, there are also differences between the atria and ventricles in myocardial morphogenesis.

Keywords: atrial morphology, pectinate muscles, hemodynamics

PP-269
Correlation of nt-probnp to exercise capacity in children with single ventricle physiology

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Background and Aim: Children with single ventricle (SV) physiology may have reduced exercise capacity despite preserved systolic function. We hypothesised that SV patients with reduced exercise tolerance would demonstrate an elevated NT-proBNP on blood analysis.
Method: 19 children with SV physiology underwent exercise stress testing (EST) to exhaustion using Bruce protocol on treadmill. Echocardiograms were performed on a Phillips machine with standardised protocol and ejection fraction (EF) calculated using Simpson's biplane. Bloods were obtained prior to exercise testing and analysed for NT-proBNP using Roche's assay. Daily symptom questionnaire completed by participants/parents.
Results: Participants aged 7-16 years (mean=10.7years). 13 females and 6 males. All participants were in Class I (No heart failure) using the Modified Ross Score and 11 (58%) participants reported at least mild limitations in daily exercise capacity compared to their peers. Length of EST ranged from 6-15 minutes (mean=9minutes). EST time was compared to healthy children of the same age and sex (1). 7 participants' time were <10th percentile for healthy children, 2 participants were in the 10th-25th percentile, 2 participants in 25th-50th percentile, 2 participants in 50th-75th percentile and 6 participants >90th percentile. Of the 8 participants without perceived exercise limitations, 7 (88%) performed better than 50th percentile for age/sex. Participants' mean EF=55.84% (33-69%). Mean NT-proBNP levels=88.8ng/L (42-189ng/L). Using Paired T-testing there was no statistically significant correlation between total EST time and EF (R=0.074 p=0.762) or NT-proBNP values (R=-0.133 p=0.586). There was no statistically significant correlation between age and total EST time (R=0.45 p=0.855). Four participants had ST segment changes on ECG during exercise. One had ectopy in recovery. All participants were asymptomatic. Of those with ECG changes, 3 participants exercised <10th percentile time for age/sex and 1 participant exercised <25th percentile for age/sex.
Conclusions: Participant's perceived levels of exercise capacity were reflected by their age/sex adjusted EST time. Marginally higher NT-proBNP levels demonstrated in participants with poorer

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exercise capacity, but this was not statistically significant. This small study suggests there may be little benefit in using NT-proBNP to determine exercise capacity in children with single ventricle physiology and their perceived daily exercise capacity is a more useful indicator of EST performance.

Keywords: NT-proBNP, exercise, single ventricle

PP-271

The evaluation of FCGR2A gene polymorphism in multisystem inflammatory syndrome in children

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Background and Aim: Multisystem Inflammatory Syndrome (MIS-C) is a new syndrome with hyperinflammation in the pediatric population exposed to SARS-CoV-2 that the pathophysiology and consequences of which have not been elucidated. FCGR2A polymorphism is associated with increased susceptibility to autoimmune and infectious diseases. The aim of our study is to search the relationship of FCGR2A rs1801274 polymorphism with the development and severity of MIS-C.
Method: The study was conducted in a single center with 35 MIS-C patients and 36 healthy children. The relationship between the participants' clinical and cardiac imaging data, severity of the disease, and FCGR2A rs1801274 polymorphism was evaluated.
Results: Although homozygous FCGR2A rs1801274 gene polymorphism was more common in MIS-C patients, no significant difference was found compared to controls. There was no significant association between this polymorphism and cardiovascular complications in MIS-C patients. However, severe cardiac dysfunction developed in those with homozygous FCGR2A rs1801274 gene polymorphism, and they required immune modulatory agents other than IVIG. The average age of patients with severe MIS-C was significantly higher than that of mild MIS-C, and systolic dysfunction was evident ($p<0.05$).
Conclusions: This study is the first to analyze the relationship between the development of MIS-C and the FCGR2A gene rs1801274 polymorphism. Since our study was conducted in a single center and in a rare and new disease group, evaluation was made with a limited number of patients. Multicenter studies in different ethnic groups are needed to examine the relationship between differences in the FCGR2A rs1801274 gene and MIS-C severity.

Keywords: multisystem inflammatory syndrome, gene, polymorphism, children

PP-272

Does intraventricular enzyme replacement therapy (ICV-ERT) with cerliponase alfa affect cardiac function in patients with cln2 disease?

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Background and Aim: Intraventricular enzyme replacement therapy (ICV-ERT) with Cerliponase alfa has shown positive effect on CNS degeneration in neuronal ceroid lipofuscinosis (CLN) 2. However, the impact of therapy itself and prolonged life expectancy on cardiac function remain unclear. Though CLNs are primary neurodegenerative diseases affected patients show an increasing amount of co-morbiditis. One of these may be a decline in cardiac function which is already reported in CLN3. The purpose of this ongoing longitudinal prospective study is to analyze cardiac function in pediatric patients with CLN2 receiving ICV-ERT with Cerliponase alfa.
Method: Forty-six patients with CLN2 with a median age of 106 months (range 41-214 m) were investigated (m = 27, f = 19). All patients received ICV-ERT. Comprehensive assessments were conducted including echocardiography, electrocardiogram (ECG), and cardiac serum biomarkers proBNP and high sensitive troponin I (hsTropI). To this date, 10 patients had a follow up at six months, three patients at twelve months, one patient at fifteen months.
Results: Echocardiography showed normal left and right systolic function in 46 (100%) patients, characterized by LV ejection fractions (EF) within the range of male 52-72% and female 54-74%, myocardial strain values of male -16.7% and female -17.8% and for the right ventricle TAPSE measurements exceeding 17mm. No patient had relevant myocardial hypertrophy. Diastolic function parameters demonstrated borderline normal function in 11 patients (24%). 43 patients had sinus rhythm, three had coordinated atrio-ventricular rhythm. Non pathologic incomplete right bundle branch blocks were observed in 8 patients. One patient showed complete right bundle branch block. Cardiac biomarker levels remained within standard limits, with hsTrop I values below 34.2 ng/l for males and 15.6 ng/l for females. Follow-up assessments after six, twelve and fifteen months showed no substantial deviations in parameters.
Conclusions: So far, our data suggest that ICV-ERT related longer life-expectancy does not lead to any relevant cardiac impairment in pediatric CLN2 patients. More important we did not see any negative effect caused by therapy at short to midterm follow-up. However, occurrence of borderline diastolic function should be followed. Whether longer life expectancy leads to impaired cardiac function in older age has still to be evaluated.

Keywords: myocard, cardiomyopathy, diastolic function, ceroid lipofuscinosis

PP-273

Clinical characteristic and outcome of barth syndrome: results from french cohort of barth syndrome evaluation (FRENCOHBASE)

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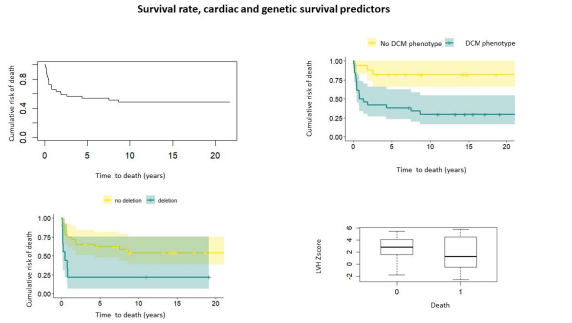
Background and Aim: Barth syndrome (BTHS) is a rare X-linked disease caused by defects in TAFAZZIN gene, responsible of cardiolipin remodelling. BTHS is characterized by cardiomyopathy, skeletal myopathy, neutropenia and growth abnormalities. Following a national series of 22 cases, published in 2013, we extended both the enrolment and the follow-up of our cohort till 2023 to describe the natural history of the disease. The second aim was to define cardiac and non-cardiac prognostic factors and to evaluate potential genotype-phenotype correlations.
Method: The medical records of all patients with BTHS, defined a TAZ variant (Class 3/4 ACMG) were identified in multiple sources and reviewed.
Results: We identified 37 BTHS pedigrees that included 44 patients. The median age at presentation was 0.12 years (range, 0–4.1 years). 21 deaths were observed at the median age of 0.5 years [min 0.06-max 8 years) while the median age at last follow-up was 14 years (range 3–52 years years). The 5-year and 20-year survival were 56% [95%CI 41-70%] and 51% [95%CI 35-65%], respectively. Two deaths occurred after heart transplantation (one immediate and the second one 7 years later from sepsis) while the other 19 deaths were related to the cardiomyopathy. The rate of failure to thrive and muscular impairment were respectively 32%, 37%. The main event during early period of follow-up was hospitalisation for heart failure. Survival correlated with different prognostic factors: age at diagnosis, type of genetic mutation as deletion type, dilated cardiac phenotype at diagnosis, grade of left ventricle (LV) hypertrophy, and LV ejection fraction. The presence of others than cardiac impairments such as muscular or neurodevelopmental impairment did predict outcomes.
Conclusions: Cardiomyopathy is the leading cause of death in BTHS and the main cause of major events in these patients. Longer-term outcomes in our cohort suggest a better prognosis after the first 5 years of life. Factors such as greater LV dilatation, reduced LV hypertrophy, and LV dysfunction were strong predictive factors of worse survival in this population. Our survey found for the first time a correlation between genotype and phenotype in BTHS.

Keywords: Cardiomyopathy, pediatric heart failure, Barth, genetic cardiomyopathy, mithocondrial genetic disease



The survival rate is worst in the early period of life with a better outcome and stabilisation after 5 years old. The DCM phenotype at diagnosis was a negative pronostic factor compared to HCM or LVNC phenotype. Moreover a greater left ventricle hypetrophy was a "protective " factor. As far concerns the genotype-phenotype correlation wa have found a greater risk of death in case of deletion compared to frame-shift or others mutations. Our survey found out for the first time a specific genetic-phenotype pattern in Barth syndrome.

PP-275

Syndromic congenital heart disease associated with novel *TBX1* and *CHD7* variants

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Background and Aim: 22q11.2 deletion syndrome (22q11.2DS) and CHARGE syndrome are two complex genetic disorders showing overlapping phenotypes, including congenital heart disease (CHD), immunodeficiency and developmental delay. The aim of our study was to ascertain genetic variants among patients with phenotypes resembling 22q11.2DS.
Method: Patients were retrospectively collected between 2005 and 2023 from 3 Hungarian pediatric centers. All patients had CHD, minor anomaly pattern resembling 22q11.2DS, and various other suggestive findings (e.g. thymus hypo/aplasia, hypoparathyroidism, hypocalcaemia, hearing impairment, coloboma, neurodevelopmental deficits). Karyotype analysis and fluorescent *in situ* hybridization (FISH) or multiplex ligation-dependent probe amplification (MLPA) were performed for all patients. Whole exome sequencing (WES) was completed for patients lacking the typical 22q11.2 deletion. Validation of identified variants and segregation analysis was run via Sanger sequencing.
Results: WES was performed in 94 unrelated patients and revealed pathogenic and likely pathogenic variants in 29 patients (30.8% diagnostic yield). Furthermore, we identified variants of uncertain significance (VUS) in 7 additional patients (7.4% of the cohort). Two novel likely pathogenic variants were identified in *TBX1* gene: a paternally inherited canonical splice variant (*TBX1*:c.438-2A>T) in Patient 1, and a nonsense variant [*TBX1*:c.612C>G (p.Tyr204Ter)] in Patient 2. Four patients carried *CHD7* alterations. Two novel likely pathogenic nonsense variants were identified: *CHD7*:c.3310_3311del (p.Ile1104Ter) in Patient 3 and *CHD7*:c.4505del (p.Ser1502Ter) in Patient 4. Patient 5 is a carrier of the previously described pathogenic *CHD7*:c.6157C>T (p.Arg2053Ter) nonsense variant. Additionally, we identified a missense VUS: *CHD7*:c.3327G>C (p.Arg1109Ser) in Patient 6.
Conclusions: This is the first study conducted on a large Hungarian patient cohort with 22q11.2DS resembling phenotypes. The WES approach is an effective diagnostic tool in syndromic CHD. *TBX1* and *CHD7* genes were affected in multiple patients (2.1% and 4.2%, respectively), strengthening the differential diagnostic relevance of these genes regarding 22q11.2DS. Importantly, we identified four novel variants, thus our results extend the genotypic spectrum of syndromic CHD-associated *CHD7* and *TBX1* genes.

Keywords: 22q11.2 deletion syndrome, CHARGE syndrome, whole exome sequencing

PP-276

Cardiac rhabdomyomas: A truly benign entity? A single-centre retrospective study in a tertiary institution

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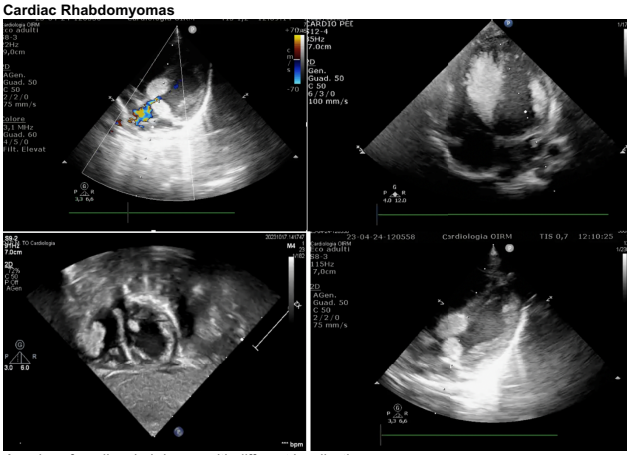
Background and Aim: Rhabdomyomas are the most common fetal heart tumour and are well-established prenatal markers of TSC (tuberous sclerosis complex). Few longitudinal studies in the literature explore the hemodynamic and electrophysiological impact of these formations over time. The aim of the study is to describe the characteristics of paediatric patients with rhabdomyomas from a genotypic, cardiovascular and arrhythmological phenotype point of view.
Method: In this retrospective study, we examined all pediatric patients in the database of a single tertiary institution with echocardiography-based diagnosis of cardiac rhabdomyoma from January 1, 2010 to August, 1st, 2023. Fetal echocardiography when available was reviewed for all cases. The presence of rhabdomyomas on postnatal echocardiography, the occurrence of arrhythmias, postnatal diagnostic and genetic work-up was reviewed using electronic medical records.
Results: During the 12.7-year study period, 41 patients were diagnosed prenatally or postnatally with cardiac rhabdomyoma at our Center. In 21 cases (55%) the diagnosis was prenatal; among patients with prenatal diagnosis, in 5 cases (24%) the family opted for voluntary termination of pregnancy. The survival rate of cardiac rhabdomyoma cases was 95%(34/36); the two deaths recorded in our case series were not related to cardiac causes. Among the 36 cases, partial or complete spontaneous tumor regression occurred in 19 (55 percent). In 3 cases, rhabdomyomatosis resulted in hemodynamic impact defined as obstruction of the LVOT, RVOT, or epiaortic vessels. Thus, in 9% of cases, rhabdomyomas resulted in significant hemodynamic alteration, requiring interventional or medical treatment. In 8 of 36 cases (22%), arrhythmias were found during follow-up (mean follow-up time of 7 years). Antiarrhythmic therapy was required in 3 cases. A clinical diagnosis of TSC according to international criteria was made in 35/36 (97%) patients. A pathogenetic variant on TSC1 was present in 38.9% of patients with positive genetic testing (61,1% on TSC2). Epilepsy was found in 64% of cases, CNS lesions in 86%, and skin, kidney, and retinal lesions in 79%, 47% and 11% respectively.
Conclusions: Rhabdomyomas can have a relevant hemodynamic impact or favor arrythmias in a significant proportion of patients with TS. A tailored cardiologic follow-up, and especially arrhythmologic surveillance, is needed in these patients.

Cardiac Rhabdomyomas



A series of cardiac rhabdomas with different localisations.

Keywords: Cardiac rhabdomyomas, Tuberous Sclerosis Compex, genotype-phenotype correlation, arrhythmological surveillance

PP-278
The influence of early-life thymectomy on immune responses and clinical manifestations in preschool children with congenital heart disease

Friederike Thiele[1], Fridrike Stute[2], Annika Boxnick[1], Sarah Jolan Bremer[1], Finja Beer[1], Daniel Biermann[2], Julia Pagel[3], Simona Murko[3], Christina Gebauer[1], Alica Krause[3], Jakob Olfe[2], Marc Lütgehetmann[4], Gwendolyn Gramer[3], Friedrich Haag[1], Michael Hübler[2], Rainer Kozlik Feldmann[2], Jörg Siegmar Sachweh[2], Eva Tolosa[1] and Anna Gieras[1]
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Background and Aim: Congenital heart disease is the most common birth defect worldwide. Depending on complexity of the CHD, around 50% of these children require cardiovascular surgery, which may involve thymectomy for a better access to the heart. Given the role of the thymus in shaping a functional adaptive immune response, thymectomy may have detrimental health consequences.
Method: We clinically and immunologically characterized 17 children (23% female, 77% male), aged 3 to 5 years, with CHD who had undergone thymectomy in their first year of life. Of these, 65% had acyanotic, and 35% cyanotic CHD. Our assessment included a total blood count, proBNP and TroponinT levels, plasma cytokine levels and comprehensive immunophenotyping. We designed a questionnaire to evaluate the frequency of atopic and autoimmune diseases and measured IgE and autoantibody levels as early markers of allergy and autoimmunity. Finally, we analysed the patients' thymic output by investigating TREC numbers and recent thymic emigrants.
Results: Our study shows that thymectomized children exhibit lymphopenia accompanied by an altered T-cell profile. These changes include a decrease of naïve CD4 and CD8 cells, concomitant with an elevated percentage of memory T-cells. We observed a significantly increased percentage of Tfh and CCR4+ CD4 cells among thymectomized children when compared to controls. Additionally, irrespective of CMV infection status, thymectomized children exhibit an increased percentage of exhausted PD1 + T-cells. Children born with a cyanotic heart defect showed a significantly higher percentage of CD8+ cells expressing the IL-2R, but a lower percentage of total CD8+ cells. Half of the children in our study displayed low TREC numbers. Analysis of plasma cytokines relevant for the maintenance of memory T-cells revealed significantly reduced IL-15 levels in thymectomized children. There was no difference in total IgE between thymectomized children and controls. No relevant autoantibody levels were detected. As expected, proBNP and TroponinT levels decreased significantly after corrective heart surgery.
Conclusions: Our research reveals the immediate immunological impact of thymectomy in early-life, including premature immune aging, reduced thymic output, and signs of lymphopenia-induced

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T-cell proliferation, potentially elevating the risk of autoimmune development later in life. These observations underscore the importance of monitoring the long-term consequences of early-life thymectomy.

Keywords: Congenital Heart Disease, Thymectomy, T cell lymphopenia

PP-279

The influence of early-life thymectomy on immune responses and clinical manifestations in preschool children with congenital heart disease

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Background and Aim: Congenital heart disease (CHD) is the most common birth defect worldwide. Approximately 50% of CDH cases, depending on the complexity, require cardiovascular surgery, which may involve resection of the thymus for a better access to the heart. Given the pivotal role of the thymus in shaping a functional adaptive immune response, thymectomy may have detrimental health consequences.
Method: We clinically and immunologically characterized 17 children (23% female, 77% male), aged 3-5 years, who underwent thymectomy in their first year of life. Of these, 65% had acyanotic CHD, and 35% cyanotic CHD. Our assessment encompassed physical and laboratory examinations, including a total blood count, cardiac biomarkers, plasma cytokine levels and comprehensive immunophenotyping. We designed a questionnaire to evaluate the frequency of atopic and autoimmune diseases and measured IgE and autoantibody levels as early markers of allergy and autoimmunity. Finally, we analysed the patients' thymic output by investigating TREC numbers and recent thymic emigrants.
Results: Our study shows that thymectomized children exhibit lymphopenia and an altered T cell profile. These changes include a decrease of naïve T cells, concomitant with an elevated percentage of memory T cells. We observed a significantly increased percentage of Tfh and CCR4+ CD4 cells among thymectomized children when compared to healthy controls. Additionally, irrespective of CMV infection status, thymectomized children exhibit an increased percentage of exhausted PD1+ T cells. Half of the children in our study displayed low TREC numbers. Analysis of plasma cytokines relevant for the maintenance of memory T cells revealed significantly reduced IL-15 levels in thymectomized children. There was no difference in total IgE between thymectomized children and healthy controls and no relevant autoantibody levels were detected. As expected, both proBNP and TroponinT levels decreased significantly after corrective heart surgery.
Conclusions: Our research reveals the immediate immunological impact of thymectomy in early-life, including premature immune aging, reduced thymic output, and signs of lymphopenia-induced T cell proliferation, potentially elevating the risk of developing autoimmune diseases later in life. These observations underscore the importance of monitoring the long-term consequences of early-life thymectomy. This work was supported by the YAEL Foundation and the German Heart Foundation.

Keywords: Congentinal Heart Disease, Thymectomy, T cell lymphopenia

PP-280

Exploring the 12-lead ECG features of pediatric carriers of plakophilin-2 gene variants

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Background and Aim: The PKP2 gene (locus 12p11.21) encodes the desmosomal protein Plakophilin-2, vital for cellular adhesion between cardiomyocytes. It is associated with classical Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC), a cardiac disorder that can cause malignant ventricular arrhythmias and sudden cardiac death in the young. The 12-lead electrocardiogram (ECG) is an important tool for diagnosis of ARVC but ECG patterns in paediatric carriers of PKP2 variants have not been previously systematically assessed.
Method: The retrospective study enrolled paediatric (<18 years) PKP2 gene carriers referred to Great Ormond Street Hospital between 2005 and 2023. Their ECGs were analysed and compared with an age and gender-matched healthy population using paired two-tailed $\chi 2$-test ($\chi 2$, p-value<0.05).
Results: 28 PKP2 variant carriers (54% females) were identified, of which 2 (7%) were diagnosed with ARVC at the time of the ECG, with a mean age of 13.6 +/- 4.8 years. These were compared to 28 healthy controls (mean age of 12.7 +/- 4.4 years). 17 carriers (60%) had low QRS voltages in the lateral leads, compared to 4 controls (16%). Fractioned QRS complexes were seen in the lateral leads in 19 (67%) and in the septal leads in 20 (71%), compared to 1 and 2 controls (3.5% and 7%, respectively). Abnormal T wave inversion (TWI) for age was found in 54% of the patients (29% in lateral leads) and flat T waves in 22 patients (78%); but these were not statistically-significantly different to controls. Both 2 patients with ARVC phenotype showed fractioned QRS, low voltages and widespread inverted or flattened T waves.
Conclusions: Paediatric carriers of PKP2 gene variants are more likely to have low QRS voltages and QRS fractionation in the lateral and anterior leads than normal controls, but the presence of abnormally inverted or flat T waves did not discriminate between gene carriers and controls. Our findings suggest that paediatric-specific ECG features may represent early phenotypic expression of PKP2 gene variants.

Keywords: arrhythmogenic cardiomyopathy, PKP2 mutation, electrocardiogram, paediatric

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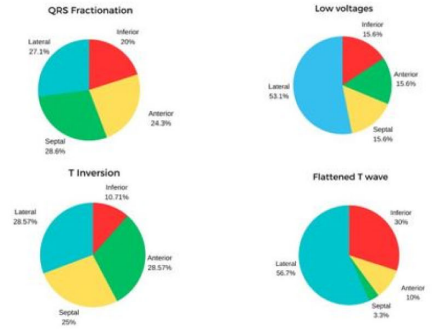
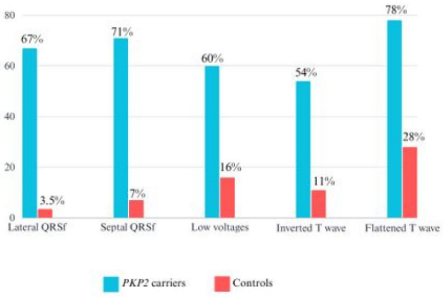
graphs



1 ECG findings in PKP2 carriers



2 Comparison of carriers-controls

PP-281

A pediatric case of dilated cardiomyopathy with inflammatory cell infiltration on histological examination

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Background and Aim: Dilated cardiomyopathy (DCM) is relatively common in pediatric cardiomyopathy, and histological examination is not mandatory for diagnosis. However, we would like to report a case that demonstrates the potential usefulness of histological examination.

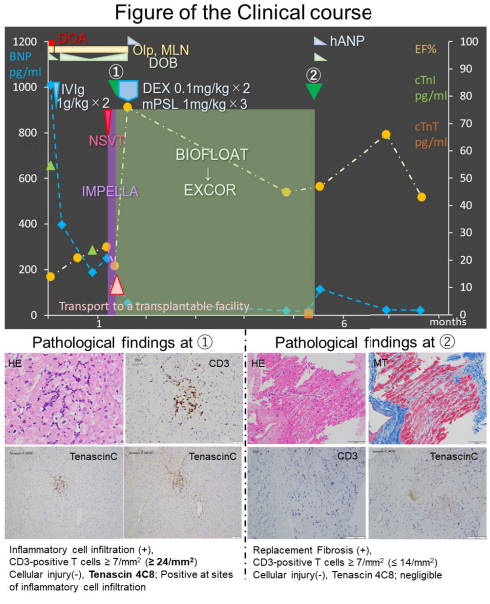
Method: We report a case in which DCM was initially diagnosed and managed, but histological examination during the disease revealed findings of myocarditis.

Results: The patient, a 12-year-old female, initially presented with gastroenteritis-like symptoms and was referred to a general hospital. An electrocardiogram showed negative T waves. Myocardial troponin I increased to 102 pg/ml. Echocardiography showed a marked decrease in ejection fraction (EF) to the 10% range and marked enlargement of left ventricular end-diastolic diameter without wall thickening. Late gadolinium enhancement on MRI findings were characteristic of DCM. However, no mapping was performed. There was no pericardial effusion. The patient was diagnosed with worsening heart failure in DCM; after more than 30 days of medical therapy, BNP improved, but EF did not. Then, the circulation could not be maintained due to ventricular tachycardia and was shifted to the ventricular assist device (VAD) in preparation for heart transplantation. Initial histological examination showed inflammatory cell infiltration, but no cellular injury. This led to the suspicion of inflammatory DCM included in Chronic Inflammatory Cardiomyopathy. In addition, CD3-positive cells exceeded 24/mm^2, and tenascin 4C8 was also stained in the inflammatory foci, suggesting a relatively active inflammation. After 4 months, the patient was able to wean off the VAD due to unloading and steroid therapy. A second histological examination revealed replacement fibrosis and still inflammatory cell infiltration, but CD3-positive cell counts were below 13/mm^2. The disease improved as did the histological findings, and EF improved to the 40% range.

Conclusions: Recently, DCM with inflammation has attracted attention as a potential target for immunosuppressive/immunomodulation therapy, while likely requiring mechanical support. Histological examination may also be considered in DCM with even the slightest suspicion of inflammatory involvement or in DCM with a poor course.

Keywords: dilated cardiomyopathy, CD3, inflammatory dilated cardiomyopathy, chronic inflammatory cardiomyopathy

Clinical course and Microscopic findings



The clinical course of the patient and microscopic findings at the time of VAD implantation and weaning off are shown.

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PP-282

Case report: Heart transplantation in a child with very long-chain acyl-coa dehydrogenase deficiency (VLCADD)

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Background and Aim: Very long-chain acyl-CoA dehydrogenase deficiency (VLCADD) is a rare autosomal recessive disorder of fatty acid metabolism. Its clinical presentation is variable and includes hypoketotic hypoglycemia, rhabdomyolysis, and potentially life-threatening multi organ dysfunction including cardiomyopathy. Treatment is based on strict dietary measures including regular feeding intervals and complete avoidance of long chain fatty acids. Heart transplantation of VLCADD patient due to severe early-onset cardiac failure has not been described in the literature.
Method: A 17-month-old male with dilated cardiomyopathy on the basis of VLCADD was referred for evaluation of heart transplant. His sister had passed away at 8 years of age due to dilated cardiomyopathy. The siblings carried the same mutation L225P, which has not been previously described in VLCADD. Both children developed cardiac disease despite optimal management of the underlying metabolic disease. Our patient became symptomatic at a younger age, indicating that his prognosis under standard treatment was poor. After careful evaluation and interdisciplinary discussion in our transplant meeting, the boy was listed for heart transplant.
Results: He received a suitable offer after a wait list time of 13 days. In cooperation with our metabolic team, heart transplantation was performed without metabolic crisis during the intraoperative and early postoperative course. Perioperative management included trigger-free anaesthesia and avoidance of propofol. Metabolic stability was sustained under perioperative glucose infusion with 17–21 g carbohydrates/kg/d (+ insuline if blood glucose exceeded 150 mg/dl). Immunosuppression was achieved with thymoglobulin, tacrolimus, mycophenolatemofetil and methylprednisolone. The patient recovered quickly and showed excellent weight gain. During the late postoperative course (Week 5 and 6), he experienced two metabolic crises with moderately elevated creatine kinase due to infections. They were treated with i.v. fluids and high doses of i.v. glucose and resolved without sequelae. Graft function remained stable at last follow-up in week 16. Biopsy after 6 weeks showed regular heart muscle histology without signs of rejection.
Conclusions: We present a novel case of heart transplantation for cardiomyopathy with a primary diagnosis of VLCADD. This case report describes the successful perioperative and postoperative management, requiring close collaboration between cardiology, cardiac surgery, metabolic medicine, anaesthesia, and intensive care.

Keywords: VLCADD, Heart transplantation, metabolic disease, dilated cardiomyopathy, genetic disease

PP-283

Outcome of irish children admitted to picu with severe myocarditis in the last decade, a single centre experience

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Background and Aim: This study describes the course and the outcome of children who were admitted with severe myocarditis requiring PICU level of care.
Method: A 10-year retrospective data review of children admitted to PICU in CHI at Crumlin with a primary diagnosis of myocarditis between 2012 and 2022. Demographic data, ICU length of stay, requirement for invasive ventilation, Extracorporeal life support (ECLS), renal replacement therapy (RRT), cardiac function parameters, cardiac enzymes, kidney and liver functions, brain imaging, mortality and requirements of a heart transplant were collected
Results: During the last decade, 22 Irish children were admitted to PICU with severe myocarditis. Eleven males and eleven females, median age 3 years (IQR:1-8). All of these children were well with no comorbidities. A causative virus was isolated in 18/22 patients (82%). Initial Echocardiographic evaluation showed severe cardiac dysfunction, Median ejection fraction (EF) of 23% (IQR16-30), and median fraction shortening of 11% (IQR:8-15). These parameters improved after Balloon Atrial Septostomy which was required in 11/22 patients (50%) and inotropic support required in 22/22 patients (100%). 9/22 patients (41%) required ECLS, median duration 8 days. 19/22 patients (86%) required invasive ventilation median duration 10 days (IQR:8-25). Cardiac enzymes (troponin, BNP, and CK) were also markedly elevated. 18/22 patients (80%) of patients had elevated markers of kidney and liver injury secondary to poor perfusion, and 5/22 patients (23%) required haemodialysis median duration 7 days (range 3-10). 3/22 patients (14%) died during their PICU stay. 4/22 (18%) patients required cardiac transplant, and one patient required temporary LVAD. The remaining 14 patients were discharged alive from PICU. Median PICU length of stay was 14 days (range10-31).
Conclusions: Myocarditis in children is a serious disease with high morbidity and mortality. A high index of suspicion is required for diagnosis. Early referral to PICU is necessary to improve the outcome.

Keywords: Myocarditis, PICU, ECLS, Heart Transplant

PP-284

Validation of drug delivery and functional activation to mitochondria in cardiomyocytes

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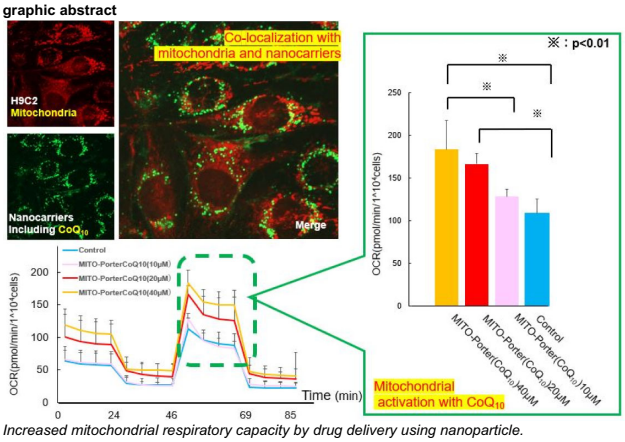
Background and Aim: In recent years, it has become clear that impaired mitochondrial function is involved in the progression of cardiac dysfunction associated with muscular dystrophy, and mitochondria-targeted therapy may be promising in the future. However, mitochondria-targeted therapy has not been developed due to difficulties in drug delivery. In this study, we firstly attempted to activate mitochondrial function in normal rat cardiomyocytes using MITO-Porter, a nanocarrier that specifically delivers drugs to mitochondria.
Method: Normal rat Cardiomyocytes (H9C2) were used as model target cells and a MITO-Porter was used as a nanocarrier. The drug to be delivered was coenzyme Q10 (CoQ10). CoQ10 plays diverse roles in mitochondria, including ATP production by respiratory chain complexes and antioxidant activity. Therefore, CoQ10 has the potential to exert a therapeutic effect on mitochondrial dysfunction, but because it is insoluble in water, it is difficult to deliver it to mitochondria simply by administering it as is, and it therefore cannot exert a therapeutic effect. Fluorescently labeled MITO-Porter was added to H9C2, and the amount of fluorescence of the intracellular nanocarriers was measured using a flow cytometer to confirm the intracellular uptake of the MITO-Porter. Using laser scanning microscopy, we confirmed that MITO-Porter was co-localized with mitochondria. Mitochondrial respiratory capacity was evaluated by measuring the oxygen consumption rate (OCR) after the addition of CoQ10-MITO-Porter using an extracellular flux analyzer.
Results: The results confirmed that the addition of CoQ10-MITO-Porter increased oxygen consumption rate (OCR) compared to the control group (Empty MITO-Porter and PBS (-) group). These results suggest that CoQ10 can be deliver to cardiomyocytes mitochondria by using MITO-Porter and CoQ10 can activate mitochondrial function in H9C2.
Conclusions: It was suggested that coenzyme Q10 delivery to mitochondria improves cardiomyocyte mitochondrial function. This could be one potential treatment for cardiomyopathy. We are particularly interested in its application to the cardiomyopathy of Duchenne muscular dystrophy, in which impaired mitochondrial energy production has been reported.

Keywords: Cardiomyocytes mitochondria, Drug delivery system, Nanoparticle, CoemzymeQ10, Cardiomyopathy, Muscular dystrophy

graphic abstract



Increased mitochondrial respiratory capacity by drug delivery using nanoparticle.

PP-285

Diving deeper into conotruncal cardiac anomalies: The diverse clinical spectrum beyond the cardiac realm

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Background and Aim: Conotruncal cardiac anomalies (CCA) often coincide with aberrations beyond cardiac disease. For instance, in 50 - 60 % of patients with neonatal diabetes mellitus (NDM) due to pancreatic aplasia (PA), causal variants in *GATA6* are found. GATA-binding factor 6 (*GATA6*) is a homeobox containing transcription factor involved in the early development of the pancreas and heart. More than 90 % of these patients also have a congenital heart defect such as truncus arteriosus communis (TAC), double outlet right ventricle (DORV) or tetralogy of Fallot (TOF). To illustrate the multisystem spectrum that accompanies CCA, we compare five patients from three independent pedigrees with pathogenic variants in *GATA6*, CCA and in three cases PA.
Method: We report the clinical, laboratory, imaging, and genetic findings of five patients from three independent pedigrees with CCA and pathogenic variants in *GATA6*.
Results: A-II.1 was prenatally diagnosed with TAC. He was born at 26 weeks of gestation. Postnatal echocardiography confirmed TAC and revealed an interrupted aortic arch (IAA) type B. Prostaglandin E1 was started to maintain ductal patency. Sonography showed pancreatic and gall bladder aplasia. Postnatal blood glucose levels confirmed NDM, and insulin therapy was initiated. He died of NEC at the age of two months. The mother (A-I.1) was born at 32 weeks of gestation with DORV and NDM due to PA. She underwent Rastelli surgery and later required transcatheter pulmonary valve replacement due to stenosis of the RV-PA conduit. B-II.1 was born at 31 weeks of gestation with DORV and died of right-sided heart failure at the age of 2 years. B-II.2 underwent ASD, VSD, and PDA correction and was also born with supravalvular pulmonic stenosis. He exhibits polydipsia, polyuria and hyperglycemia. Their sister and father who also carry the *GATA6* variant only have mild cardiac defects: PFO, ASD and VSD respectively. Family B emphasizes the strong intrafamilial variability ranging from mild septal anomalies to fatal CCA. C-II.1 was born preterm with TOF and NDM. He underwent complete intracardiac repair and later required stenting of the superior vena cava and the left pulmonary artery and is now 14 years old. Imaging later confirmed pancreatic aplasia.
Conclusions: CCAs are often associated with a broad clinical spectrum beyond the cardiac realm including endocrinological and gastroenterological aberrations. Therefore, comprehensive diagnostics including an early genetic workup are imperative for a holistic management of these patients.

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Keywords: conotruncal cardiac anomalies, truncus arteriosus communis, neonatal diabetes mellitus, pancreatic aplasia, GATA6

PP-286

Progression of qrs fragmentation in children with duchenne muscular dystrophy: A 12-LEAD ECG longitudinal study

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Background and Aim: Duchenne muscular dystrophy (DMD) is a rare genetic disorder characterized by a lack of dystrophin, leading to various morbidities, including progressive myocardial fibrosis as evidenced by CMR, cardiomyopathy and early onset heart failure, often in early adulthood or teenage years. Regular cardiac monitoring through yearly ECG and echocardiography is recommended since diagnosis, although their prognostic efficacy is not fully established. This study was conducted to evaluate the presence and progression of QRS fragmentation in 12-lead ECG tracings of children diagnosed with DMD.
Method: A cohort 84 of DMD patients, with a total of 125 ECGs was analysed at two distinct stages in the disease: early i.e. ≤9 years of age and late (>9y). The presence of QRS fragmentation was evaluated in various all 12-leads. The statistical significance of changes in between the stages was assessed using chi2 test with $p<0.05$ threshold.
Results: We analysed 63 recordings in the early stage and 59 in the late stage of DMD. There were 50.9% of early recordings featuring fragmentation in at least one lead and 69.4% late ones, p=0.034. Significant increases in QRS fragmentation were observed in several leads from the early to the late stage of the disease. Specifically, leads I (from 1.6% to 10.0%, p=0.042), II (from 1.6% to 13.6%, p=0.022), III (from 12.9% to 33.9%, p=0.005) and V5 (from 1.6% to 10.0%, p=0.042) showed notable increases in fragmentation prevalence. Lead V4 also demonstrated a considerable increase (from 4.8% to 16.7%, p=0.052). Other leads also showed increases in fragmentation, but these were not statistically significant, possibly due to sample size.

QRS Fragmentation in Early and Late Stages of DMD in Children

	early	late	p	
n	63	59		
I [fr]	1.6%	10.0%	0.042	*
II [fr]	1.6%	13.6%	0.022	*
III [fr]	12.9%	33.9%	0.005	*
aVL [fr]	19.0%	26.7%	0.289	
aVF [fr]	15.9%	27.1%	0.130	
avR [fr]	3.2%	1.7%	0.598	
V1 [fr]	25.4%	35.0%	0.304	
V2 [fr]	14.3%	16.7%	0.685	
V3 [fr]	9.5%	15.0%	0.335	
V4 [fr]	4.8%	16.7%	0.052	
V5 [fr]	1.6%	10.0%	0.042	*
V6 [fr]	1.6%	8.3%	0.079	

Table 1: comparative analysis of QRS fragmentation in early and late childchood stages of Duchenne Muscular Dystrophy across different ECG leads

Conclusions: This study highlights a significantly high prevalence of QRS fragmentation in DMD patients and an increase in QRS fragmentation in certain ECG leads in patients with DMD as the disease progresses. Notably, leads I, II, III, and V5 demonstrated a significant increase in fragmentation, suggesting a potential utility of these ECG findings as indicators of disease progression in DMD. Further research is warranted to explore correlations with echocardiography and CMR findings and understand the implications of these observations for ECG as a tool for monitoring disease severity and progression in DMD.

Keywords: Duchenne Muscular Dystrophy, 12-Lead ECG, cardiomyopathy, heart failure, QRS fragmentation

PP-287

Severe dilated cardiomyopathy in a child with PITT-hopkins syndrome

Jelena Hubrechts[1], Eric Olinger[2], Claire De Halleux[3], Alain Poncelet[4], Yves Sznajer[2] and Stéphane Moniotte[1]
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Background and Aim: In contrast to other types of cardiomyopathies, dilated cardiomyopathy (dCMP) is more rarely of genetic origin. In young patients, associated clinical signs may present later than the cardiac features, etiological diagnosis being challenging.
Method: A 6-months old girl presented with dCMP with on initial echocardiography a very dilated LV (Z-score +9,8) and severe dysfunction with ejection fraction (EF) of 10%. Pro-NT-BNP level was 20142pg/ml. In absence of improvement after 12 days of conventional intensive treatment, pulmonary artery banding was performed, with successful weaning of ventilator despite persistent cardiac dysfunction (LVEF 24%). There were no extra-cardiac symptoms at that stage. Metabolic screening was negative. LV dilatation and function worsened over time. She developed marked global developmental delay (DD), stereotypical hand and head movements, and growth retardation, initially thought to be related to the persistent cardiac condition. SNP-array and panel testing for cardiomyopathy and lysosomal storage diseases were noncontributing. Trio exome sequencing (ES) revealed a de novo likely pathogenic variant (NM_001243226.3: c.638_642delCTTAT; p.(Ser213Ilefs*11)) in TCF4 establishing the molecular diagnosis of PTHS.
Results: This genetic finding explained the non-cardiac features and helped us to rule out invasive approaches such as heart transplantation. This was also of major importance for the parents to accept palliative care. Currently, at the age of 3,5 years, LV dilatation is still very important (+ 18,4 Z-score) as is the dysfunction (LVEF 10%). Palliative care with chronic heart failure therapy is continued at home. As far as we know, this is the first patient described with dCMP and PTHS. TCF4 encodes an ubiquitous transcription factor mainly involved in cellular differentiation, proliferation and lineage commitment. We did not detect other known genetic etiologies for dCMP on targeted panel analysis nor in the exome. Systematic unbiased assessment of infantile dCMP patients in the future will be crucial to confirm a possible relationship between TCF4 variants and dCMP.

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Conclusions: Multidisciplinary approach in patients with dCMP contributes to novel insights. With a genetic diagnosis, precision medicine approaches including major therapeutic decisions such as heart transplantation are clearly considered differently. This makes an important difference for the patient, parents and medical team.

Keywords: dilated cardiomyopathy, heart failure, developmental delay, whole exome sequencing, Pitt-Hopkins syndrome

PP-288

Innate immune signaling in hearts and buccal mucosa cells of patients with arrhythmogenic cardiomyopathy

Joanna Jager[1], Carlos Bueno Beti[1], Alessandro Tafuni[2], Stephen P Chelko[3], Mary N Sheppard[1], Ella Field[4], Jennifer Tollit[4], Imogen K Heenan[4], Annabelle Barnes[4], Matthew R Taylor[5], Luisa Mestroni[5], Juan Pablo Kaski[4], Jeffrey E Saffitz[6] and Angeliki Asimaki[1]
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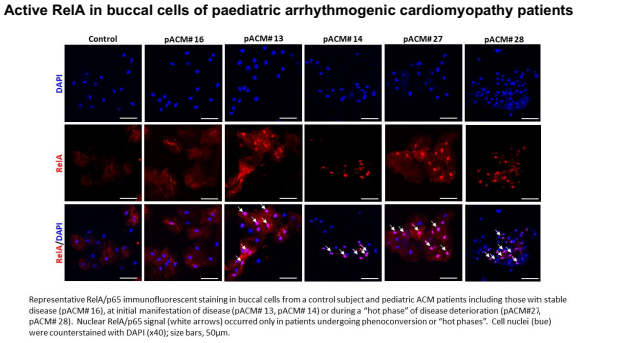
Background and Aim: Despite decades of research, the diagnosis and management of arrhythmogenic cardiomyopathies (ACM) remain challenging, mostly owing to poor understanding of disease mechanisms. Heretofore, inflammation in ACM was regarded as a consequence of cardiac myocyte (CM) death. Recent studies, however, show that inflammation is in fact a driving force of the disease. The major inflammatory pathway nuclear factor kappa B (NFκB) is activated in CMs and causes disease in a mouse model of ACM by mobilizing C-C chemokine receptor type 2 (CCR2)-expressing macrophages, which promote both myocardial injury and arrhythmias. We have previously shown that pathological processes occurring in the hearts of ACM patients are mirrored by equivalent abnormalities in the buccal epithelium; the cheek's inside. Herein, we sought to determine if persistent innate immune signalling via NFκB occurs in CMs in patients with ACM and if this is associated with myocardial infiltration of pro-inflammatory cells expressing CCR2. We also sought to characterize NFκB signalling in buccal mucosa cells from children bearing ACM-causing mutations.
Method: We analysed myocardial samples from ACM patients who died suddenly (n=29), required cardiac transplantation (n=6) or had an endomyocardial biopsy (n=1). We also analysed buccal mucosa cells from children with inherited disease alleles via immunocytochemistry and confocal microscopy (n=28). The presence of immunoreactive signal for RelA/p65 in nuclei of CMs and buccal cells was used as a reliable indicator of active NFκB signalling. Myocardial samples were also examined for the CCR2-positive macrophages.
Results: NFκB signalling was active in CMs in 34/36 cases of ACM but none of 19 aged-matched controls ($p<0.0001$). Cells expressing CCR2 were significantly increased in patient hearts in numbers directly corelated with the number of CMs showing NFκB signalling. NFκB signalling was active in buccal cells only in those children showing initial clinical signs of ACM (phenoconversion) or undergoing clinically unstable phases of the disease (hot phases).
Conclusions: Patients with clinically active ACM exhibit persistent innate immune responses in CMs and buccal mucosa cells reflecting both a local and systemic inflammatory process. Our results indicate that patients undergoing phenoconversion or deteriorating phases of the disease may benefit from anti-inflammatory therapy.

Keywords: arrhythmogenic cardiomyopathy, innate immune signaling, nuclear factor κB, pro-inflammatory macrophages, buccal mucosa cells



Representative confocal immunofluorescence images showing activation of the NFkB pathway only in paediatric ACM patients undergoing active phases of the disease (onset or deterioration)

PP-289

Acute ischemic stroke leading to a diagnosis of restrictive cardiomyopathy in an adolescent

José Luis López Guillén, Joshua Griesman, Mike Seed, Alyssa Power, Aamir Jeewa and Anne Dipchand
Labatt Family Heart Centre, Hospital for Sick Children and University of Toronto, Toronto, Canada

Background and Aim: Pediatric acute ischemic stroke (AIS) is a challenging diagnostic entity. The clinical presentation is often atypical, leading to delayed diagnosis and significant morbidity, and reinforcing the need for a thorough workup for etiology. Here we present the case of an adolescent with AIS who was found to have restrictive cardiomyopathy (RCM).
Method: Following informed consent and IRB approval, the medical chart of a 15-year-old female who presented with AIS was reviewed.
Results: A 15-year-old female presented with left-sided paresis, hemi-neglect, and facial paralysis. Neuroimaging demonstrated a right middle cerebral artery (MCA) territory infarct which was confirmed by angiography and mechanical thrombectomy was performed (Figure 1). Testing for etiology for AIS included a CXR which showed cardiomegaly, pulmonary congestion and pleural effusions, and an ECG which demonstrated biatrial enlargement, intraventricular conduction delay, and ST depression and T-wave inversion in the inferior and anterolateral leads (Figure 2). Screening for thrombophilia disorders and prothrombotic conditions was negative. An echocardiogram (Figure 3) revealed severe biatrial enlargement and abnormal diastolic function in keeping with RCM, providing support for a cardioembolic source of AIS. Cardiac MRI findings were consistent with RCM and severe bilateral lymphangiectasia. Neurologic deficits improved and she received a heart transplant one month later. Genetic testing ultimately identified a heterozygous pathogenic variant in MYH7.
Conclusions: Approximately 15% of AIS in adolescents have been shown to be cardioembolic in origin. Thromboembolic events occur in 12-33% of pediatric RCM patients, half of which are

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cerebrovascular events. This case illustrates the rare presentation of RCM following an AIS and underscores the importance of investigating for a cardiac etiology.

Keywords: cardioembolic stroke, RCM, echocardiography, acute ischemic stroke

All figures

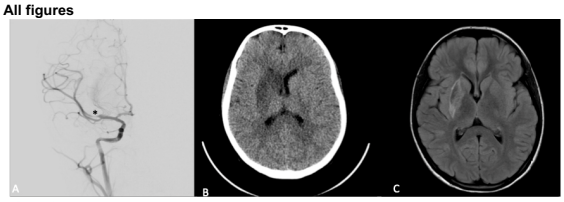

Figure 1.

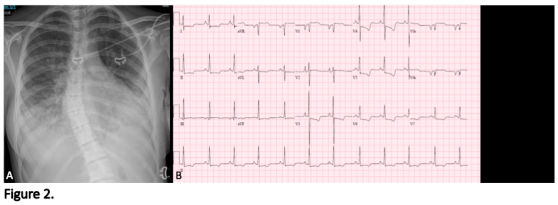

Figure 2.

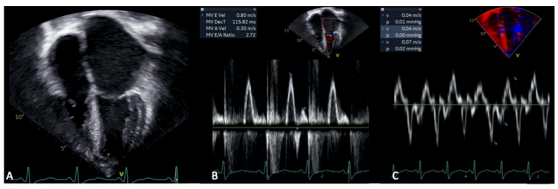

Figure 3.

Figure 1. (A) Cerebral angiogram showing occlusion of the M1 segment of right middle cerebral artery (*). (B) Non-contrast CT head and (C) MRI axial diffusion-weighted imaging showing area of infarction involving the right basal ganglia, anterior and posterior limbs of the internal capsule, and partial obliteration of the adjacent anterior horn of the lateral ventricle. Figure 2. (A) Chest x-ray with moderate cardiomegaly, small bilateral pleural effusions and pulmonary edema. (B) Electrocardiogram showing biatrial enlargement and diffuse repolarization abnormalities. Figure 3. (A) Apical four chamber view demonstrating a massively dilated left atrium with normal left ventricular chamber size. (B) Mitral inflow doppler tracing with E:A ratio > 2.5 and short deceleration time, suggesting reduced ventricular compliance. (C) Tissue doppler of left ventricle lateral wall demonstrating reduced e' velocity and increased E/e' ratio.

Interventional Cardiology

PP-290

Percutaneous aortic valvuloplasty for severe congenital valvar aortic stenosis in children: choice than alternative

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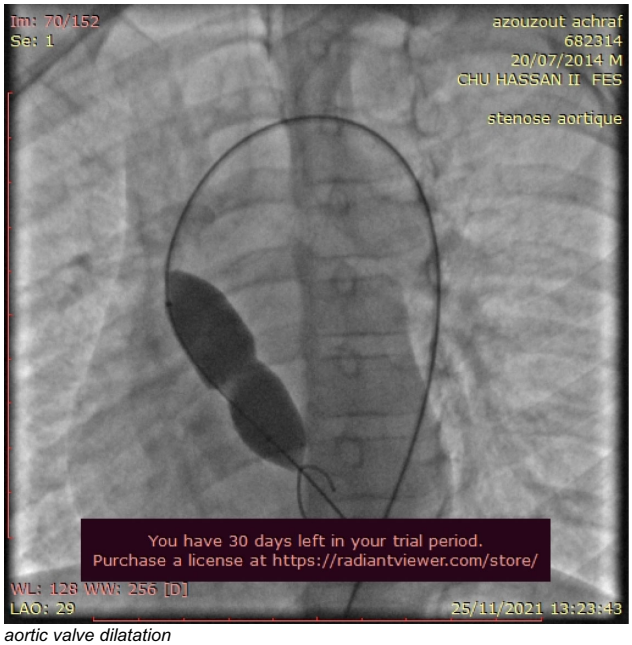
Background and Aim, Objective: evaluate the efficacy and the risk of percutaneous balloon aortic valvuloplasty (PBAV) for severe congenital aortic valve stenosis in infant and children
Method: Between June 2018 and March 2023; we have conducted retrospective study to analyze Fifteen child underwent PBAV for congenital and severe aortic stenosis. The age of our patient ranged from 4 days to 16 years. The procedure was performed when the peak-to-peak gradient VG-Aorte is greater than 45 mmhg). The result was considered as good when the residual gradient was inferior to 25 mm hg with trivial or moderate. three parameters were evaluated to assess the efficacity of this procedure: aortic regurgitation (AR), repeat aortic valve dilatation and occurrence of a serious complication as death or neurologic accident.
Results: Thirty-seven aortic valve dilatation was performed. seven patients had less than 5 years and 25 older than 5 years. The mean gradient was 60 ± 25 mmhg. LVEF was normal in all cases except in two neonates. post dilatation gradient varied from 30 +/- 16 mmhg. The aortic annulus diameter ranged from 7.0 to 20 mm. The ratio of balloon to aortic annulus diameter ranged from 0.8 to 1.00. Moderate aortic insufficiency was reported in one case and only trivial in 5 cases. Severe arterial injury which require surgical intervention was occurred in one patient. transitory bradycardia, arrhythmia was encountered during the procedure in 3 patients. we deplored two dying patients all of them during neonate period. One from severe aortic regurgitation with severe LV dysfunction, one by nosocomial infection, and one later after unluckily numerous attempts. All of our patients are free of novel dilatation by 6 to 72 months. except Just one patient who was needed one more aortic valve dilatation after three years with good result.
Conclusions: percutaneous Balloon dilatation is a feasible palliative procedure for child with isolated aortic valve stenosis. The ratio of balloon to aortic annulus diameter ranged from 0.8 to 1.00 seem to be the most parameters for an effective result. Prospective and multicentric studies are mandatory to improve our experience.

Keywords: percutaneous balloon aortic valvuloplasty,children, Aortic regurgitation, Gradient

Aortic stenosis dilatation



aortic valve dilatation

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PP-291

Anomalous systemic venous return with bilateral SVC AND RSVC to left atrium, unexpected finding in a cyanosed baby

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Background and Aim: Anomalous systemic venous connection with the left atrium (LA) is an unusual congenital cause of a right-to-left shunt. The most common variant is persistent left superior vena cava (SVC), which is estimated to occur among 2.1% to 4.3% of the people with congenital hearts defects. An earlier report by Nazem et al. showed that RSVC to LA caused brain abscess in a 26 years old lady.

Method: We are presenting a child with bilateral SVC and RSVC to left atrium. He presented at age of 7 months with low saturation during routine check up with no respiratory cause and normal chest X-ray. He is a term baby with uneventful antenatal history. Referred to cardiology for assessment.

Echo showed Small high ASD with left to right shunt, however the RSVC was interestingly draining to the left atrium, Pulmonary veins were normal with no PAPVD. There is LSVC to dilated coronary sinus to right atrium.

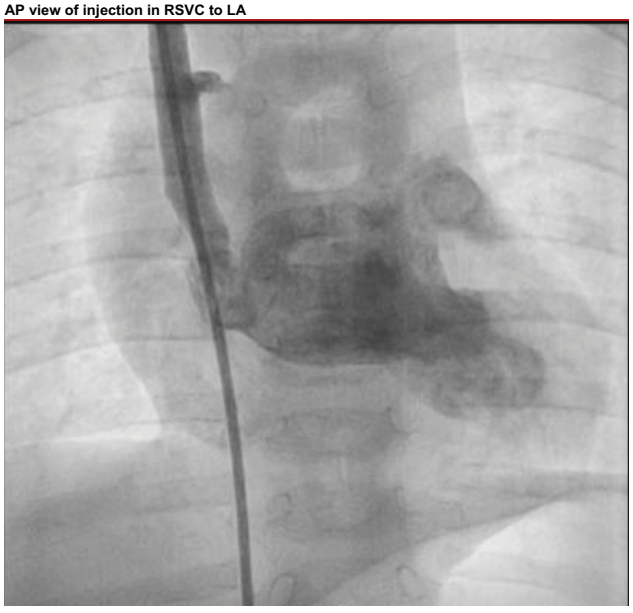
So he was referred for diagnostic cardiac catheter and trans esophageal echo which conformed the above findings. Hemodynamics assessment revealed Qp:Qs 0.9:1. The family was counseled and patient started on Aspirin and advised against any cannulation or injections in the right arm. Cardiac CT was done at age of 3 and half years and confirmed the diagnosis. Patient was continued on follow up in the cardiology clinic and the plan is to prepare him for surgery to route the RSVC to the right atrium.

Results: Anomalous RSVC to LA is a right to left shunt. Proper family counseling about avoiding cannulation of the right arm and jugular seems a sensible approach. Antithrombotic can prevents right to left emboli similar to approach to PFOs with right to left shunting to prevent transient ischemic attacks and strokes. Addressing this anomaly surgically by repositioning the RSVC to RA is important

Conclusions: In cases of hypoxemia echocardiographic assessment is important to exclude such rare anomalies. The right Superior vena cava to left atrium is rare anomaly that can be delineated accurately using multiple modalities of imaging like TTE/TOE and Cardiac CT. Cardiac catheterization is an important tool to gather hemodynamic data in this entity.

Keywords: Right superior vena cava to left atrium, cyanosis, right to left shunt

AP view of injection in RSVC to LA



PP-292

Left ductal stent to disconnected LPA, RVOT stent and right duct closure in the context of severe form of TOF, double aortic arch and disconnected LPA

Ahmed Adel Hassan, Mahmoud Alsoufi and Mohamed Kasem

Pediatric Cardiology Department Heart Center of Excellence, Al Jalila Childrens Specialty Hospital, Dubai Academic Health Corporation, Dubai, United Arab Emirates

Background and Aim: We are presenting this case to highlight the importance of pre planning of complex cath procedures, the safety of intervening in double arch ducts. The importance of optimizing the hemodynamics after stenting unilateral duct.

Method: We are presenting complex congenital heart disease presenting with duct dependent pulmonary circulation with low birth weight that was palliated in cath. Echo showed TOF with narrow LPA origin, L PDA supply the LPA, TOF, severe RVOTO, and right duct from double arch. Cardiac CT showed Double aortic arch with dominant left arch, LSCA and LCA arise from the left arch, and RSCA and RCA arise from right arch. MPA is small connects only to RPA. Right duct arise from the RCA and connect to RPA. Vertical Left duct arise from the left arch and connect to the LPA. Second duct from the right arch which connect to RPA. There is discontinuity between the LPA and MPA.

Results: We took patient to the cathlab at age of nine days, 2.9 KG. The rational was the low saturation, disconnected LPA. we stented the Left PDA to keep flow to LPA using 3/15mm DES, but diastolic pressure was low 19mmHg, saturation still was below 85%. Steal from right duct was the main drive for the low diastolic pressure. We confirmed by test balloon occlusion of right duct. RVOT was stented to improve saturation, using 5/16 DES, saturation went up to 90%. We occluded right PDA to reduce diastolic steal and improve diastolic blood pressure. using MVP 7Q plug via RAA access. Flow across the double arch to the head and neck vessels was intact. No difference between the pressure of the RAA and abdominal aorta. Diastolic pressure came up from 19mmHg to 25mmHg. We routinely use Verapamil for arterial access of neonates. USS of arterial sites was normal.

Conclusions: The baby was extubated the following day, and discharged after few days on dual anti-platelets. Patient was then observed for 2 weeks in the NICU and was doing well so discharged and are following in out out patient cardiology department. Full surgical repair was done at age of 9 months

Keywords: Disconnected left pulmonary artery, Double aortic arch, RVOT stenting, Ductus stenting, Duct occlusion

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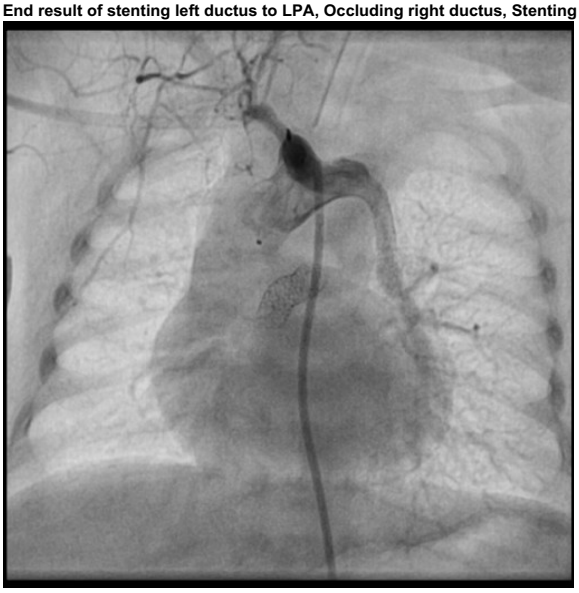
End result of stenting left ductus to LPA, Occluding right ductus, Stenting RVOT



PP-293

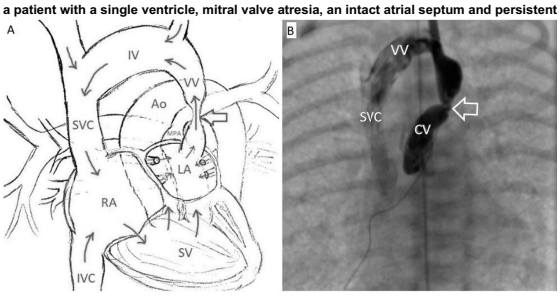
Palliative stenting of critical systemic vein obstruction in newborns with total anomalous pulmonary venous return

Artem Gorbatykh, Aigul Mingalieva, Igor Averkin, Evgeny Grekhov, Alexey Prokhorikhin and Dmitriy Zubarev
Department of Pediatric Cardiac Surgery. V.A. Almazov National Medical Research Center, Saint-Petersburg

Background and Aim: To present two cases of palliative treatment in newborns with critical obstruction of the venous outflow in terms of total anomalous pulmonary venous return.
Method: In 2022, in V.A. Almazov NMRC, two endovascular interventions were performed for critical obstruction of the systemic veins in patients with a total anomalous pulmonary venous return. Both patients were newborns, gestational age 29 and 41 weeks, their weight being 2,400 and 3,160 kg. Newborns were in critical state due to respiratory failure, hypoxemia (SpO2 60%) and signs of pulmonary edema and were transferred to mechanical ventilation. Concomitant intracardiac anomalies were observed. In the first newborn, a single inlet double outlet right single ventricle with moderate subpulmonary stenosis. In the second newborn, non-obstructive double outlet right single ventricle with intact atrial septum and obstruction of the persistent vertical vein. In both cases, the stenotic systemic vein was the only drain from the pulmonary venous collector. The systolic gradient in the constriction area in a patients was more than 14 mm Hg, and the blood flow velocity reached 3 m/s. In the first case, stenting was performed through the femoral vein peripheral stents 10x18 mm via the 4 Fr sheath; in the second case, the stenotic systemic vein was accessed through the left jugular 8x15 mm via the 4 Fr sheath.
Results: Newborns underwent palliative stenting for systemic vein obstruction with good clinical outcome. After the procedure, the pressure gradient during invasive measurement (PV collector / SVC) was less than 2 mm Hg. The pressure in the PV collector in both cases was 12 mm Hg. According to the ECHO data, the blood flow velocity in the stented area was 1.2 m/s. Saturation increased SpO2 90%. In the patient with a persistent vertical vein, narrowing of the PA was performed.
Conclusions: Stenting of critical systemic vein obstruction with total anomalous pulmonary venous return as a primary intervention is possible and can be used as a bridge for the next stage of surgical correction.

Keywords: palliative endovascular treatment, single ventricle, stent.

a patient with a single ventricle, mitral valve atresia, an intact atrial septum and persistent cardinal vein



PP-294

Isolated major aortopulmonary collaterals- a rare cause of hemoptysis in a child

Akriti Gera, Amit Misri, Pankaj Bajpai and Shefali Yadav
Department of Paediatric Cardiology, Medanta Hospital, Gurugram, Haryana, India

Background and Aim: Major aortopulmonary collateral arteries (MAPCAs) are abnormal systemic-to-pulmonary collateral vessels originating from the persistent segmental arteries. Symptomatic MAPCAs are usually found in patients with cyanotic congenital heart disease with decreased pulmonary blood flow. Isolated MAPCAs, are usually seen in preterm newborns, are asymptomatic and resolve spontaneously. Here we report a case of symptomatic isolated MAPCAS in a teenager.
Method: We retrospectively reviewed a single case and collected data from records. Review of literature on isolated aortopulmonary collaterals was done.
Results: A previously healthy 15-year-old female presented to respiratory medicine with 3-4 episodes of hemoptysis and shortness of breath. General and systemic examination was unremarkable. She was being treated for left lower lobe pneumonia. Cardiology opinion was sought in view of persistent symptoms and non-contributary bronchoalveolar lavage. 2D echocardiography showed retrograde flow in left pulmonary artery (LPA) and dilated left atrium (LA) and ventricle (LV). CT pulmonary angiography showed dilated LPA with abnormal branching which formed a mesh of collaterals, and drained into the left subclavian vein (LSCV) and internal mammary veins (LIMV). During cardiac catheterization, branch PA angiograms showed competitive flow into LPA and normal RPA. No pulmonary AV malformation (PAVM) was seen. Aortogram depicted two significant collaterals draining into LPA after forming a mesh. Left subclavian artery angiogram showed one significant collateral forming a mesh and draining into LPA. No collateral seen from right subclavian angiogram. Intervention Vascular plugging and coiling of significant

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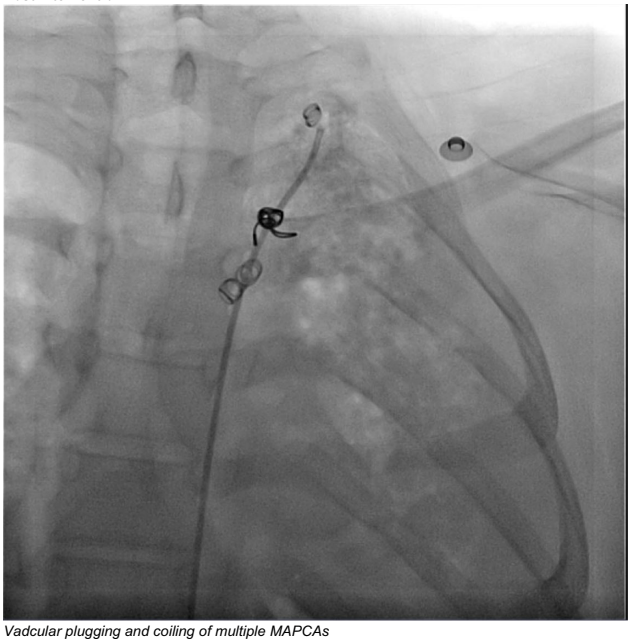
collaterals from descending thoracic aorta to LPA, descending thoracic aorta to left lung, left subclavian artery to LPA was done. There was significant reduction in flow after coiling and device closure. Patient's hemoptysis resolved and she is now asymptomatic.
Conclusions: Isolated MAPCAs are usually a benign anomaly, presenting without any clinical finding and requiring no specific treatment. MAPCAs leading to hemoptysis is a rare occurrence, but timely intervention has good results.

Keywords: Major aortopulomary collaterals, Isolated MAPCAs, hemoptysis

Post intervention



Vadcular plugging and coiling of multiple MAPCAs

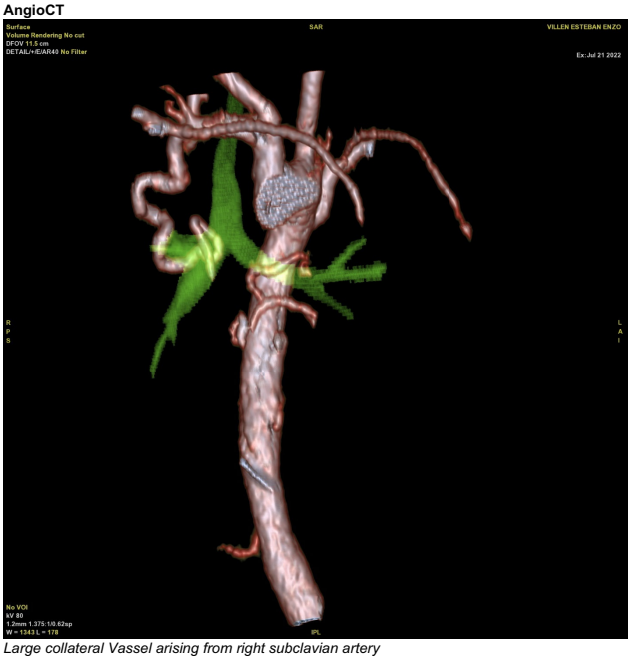
PP-295
What is wrong with this newborn? extubation failure after arterial switch operation

Alex Irurzun Rodriguez[1], Begoña Loureiro Gonzalez[4], Roberto Blanco Mata[2], Javier Ayala Curiel[3], Sonia Blazquez Trigo[3], Maria Teresa Luis Garcia[3], Erika Rezola Arcelus[1] and Jose Miguel Galdeano Miranda[3]
[1]Pediatric Cardiology. Hospital Universitario Donostia. San Sebastian. Spain; [2]Hemodynamic Cardiology, Hospital Universitario Cruces, Barakaldo, Spain; [3]Pediatric Cardiology, Hospital Universitario Cruces, Barakaldo, Spain; [4]Neonatal Intensive Care Unit, Hospital Universitario Cruces, Barakaldo, Spain

Background and Aim: Arterial switch operation is nowadays the preferred treatment for D-transposition of great arteries (D-TGA). Some factors, like hypoxemia, may help develop aortopulmonary collateral arteries. These collateral arteries can be a cause of extubation failure and hemodynamic instability in these newborns.
Method: We report a case of a newborn with a successfully corrected D-TGA at 7 days-of-age, with repeated extubation failures due to pulmonary haemorrhage (at 74 and 86 days-of-age). Previously, at 36 days-of-age, extubation was tried but failed due to left main bronquial obstruction and vocal cord paresis. He also suffered a persistent chylothorax due to a large innominate vein thrombus that was repermeabilized with balloon angioplasty at 61 days-of-age.
Results: A large aortopulmonary collateral artery was detected in a CT scan at 88 days-of-life and was embolized with microcoils at 89 days-of-age. Afterwards, extubation was successful at 91 days-of-age.
Conclusions: Aortopulmonary collateral arteries can be a reversible cause of extubation failure in patients with D-TGA after arterial switch operation and must be suspected. Other causes of chronic hypoxemia in the newborn (extremely preterm, chronic pulmonary desease) can cause the development of these aortopulmonary collateral arteries.

Keywords: D-TGA, Arterial switch operation, aortopulmonary collateral arteries, embolization

AngioCT



Large collateral Vassel arising from right subclavian artery

PP-296
Challenges and opportunities in translating a novel autologous pulmonary valve replacement – an interim report and guidance for translation

Alexander Breitenstein Attach[1], Marvin Steitz[2], Jonathan Kiekanap[3], Jordi Modolell[2], Felix Berger[1] and Boris Schmitt[1]
[1]Department of Pediatric Cardiology and Congenital Heart Disease, German Heart Centre Charité; [2]Department of Pediatric Cardiology and Congenital Heart Disease, Charité University Medicine Berlin; [3]German Centre for Cardiovascular Research (DZHK)

Background and Aim: Each year ~300.000 babies are born with a congenital heart valve defect worldwide; however, a regenerative and durable solution remains yet to be found.
Method: To tackle this medical need, a Procedure Pack (PP) has been developed which enables the intra-operative fabrication of an autologous heart valve replacement. The PP consists of a stent, catheter delivery system, crimping tool, crosslinker and mould. Functionality and translatability were shown over 3 design and 1 concept iteration in-vitro as well as in-vivo.

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Results: The first 3 design iterations utilized in-house-developed nitinol stents and delivery systems. In sheep, iteration 1 and 2 did not achieve acceptable results over 12 months due to unfavourable valve design and crosslinking degree, resulting in median regurgitation fraction (RF) of 43 % measured by MRI underlined by moderate to severe insufficiency by intracardiac echocardiography (ICE). Adjustments in valve shape and crosslinker in iteration 3 allowed fabrication of fully functional valves with median MRI RF of 8 % and trivial to moderate insufficiency by ICE up to 20.5 months. These results were verified in-vitro at certified laboratory AC-Biomed. During all follow-up evaluations, no stenosis was present. Histologically, all valves showed endotheliazation and favourable CD163+ macrophage infiltration. 1 valve showed early signs of degeneration. Though the third iteration was fully functioning, cost and time to maintain CE certification as per Medical Device Regulation of all five PP components made translation impossible. This urged the need to replace stent, delivery system and crimping tool with commercially available products, limiting the in-house developments to the core intellectual property, which does not yet exist on the market, namely mould and crosslinker. An acute trial in minipigs (n=3) demonstrated the feasibility of the modification with mild insufficiency and no stenosis, measured by ICE.
Conclusions: The novel autologous pulmonary valve implant performed well up to 20.5 months, with successful adaptation to commercially available medical devices. Also, regulatory and certification costs should be considered early on through risk assessment following ISO 14971 guidelines. This paper provides insight into the development of medical devices from a pediatric cardiology department to overcome forced widespread off-label use of devices.

Keywords: Transcatheter Heart Valve Prosthesis, Autologous Heart Valve Replacement, Tissue Engineering

PP-297
Evaluation of fenestrated fontan patientS: When to consider closure?

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Pediatric Cardiology, University of Health Sciences, Istanbul Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Training and Research Hospital, Istanbul, Turkey

Background and Aim: In Fontan procedures, a surgically created baffle fenestration has been shown to reduce postoperative morbidity at the expense of systemic desaturation. However, leaving the fenestration open poses the risk of desaturation and systemic embolism due to the created right-to-left shunt. In follow-up examinations, in patients who have experienced suspected or confirmed embolism, or in cases where saturation levels are intolerably low, fenestration closure can be performed using a transcatheter method. Here, we present our patients who underwent fenestrated Fontan procedures, with the subsequent closure of fenestration using a transcatheter approach.
Method: A retrospective evaluation was conducted on 15 patients at our center who underwent transcatheter closure of Fontan fenestration between the years 2015 and 2023 due to desaturation and suspected systemic embolism, with no major complications. All patients were intubated before the procedure. Prior to fenestration closure, balloon occlusion tests were performed, demonstrating an increase in saturation and appropriate pressure.
Results: Of the patients, 53.3% were male, with a mean age of 10.6 ± 5.2 years. Thirteen patients underwent the procedure due to desaturation, while 2 were considered for suspected systemic embolism. No major complications were observed in any patient during or after the procedure. The average length of hospital stay post-procedure was 3.9 ± 4.6 days. The pre-procedural Fontan pressures were 12 ± 3.1 mmHg, and post-fenestration closure pressures were 12.6 ± 2.7 mmHg. Patients showed an 11% increase in saturations and improvement in clinical status. The mean duration between fenestration opening and closure was 22.6 ± 13.7 months (Table 1). In one patient, the fenestration was reopened using a transcatheter approach four months after closure due to recurrent and prolonged pleural effusion. After detecting residual flow through the fenestration in one patient, a covered stent was deployed in the Fontan baffle ten days post-procedure.
Conclusions: In high-risk Fontan patients, the opening of fenestration through surgical or postoperative transcatheter interventions can result in acute clinical improvement. However, in some of these patients, undesired situations such as intolerable desaturation or systemic emboli may occur. In such conditions, the fenestrations of patients can be safely closed using a transcatheter approach.

Keywords: interventional cardiology, fenestration, Fontan circulation, transcatheter closure

Table 1. Demographic data and procedure characteristics of the patients

Patient no	Diagnosis	Anatomy of Fontan connection	Fontan age (year)	Time period fenestration opening& closure (month)	Indication	Balloon occlusion size	Device Size
1	DILV-VA discordance, dextrocardia	Extracardiac conduit	14	11	desaturation	8x20 mm Ovypka balloon	18x18mm Amplatzer PFO occluder
2	DIRV- DORV	Extracardiac conduit	8	23	desaturation	7x20 mm Ovypka balloon	18mm Amplatzer multimesh ASD occluder
3	DIRV- DORV	Extracardiac conduit	11	36	desaturation	7x20 mm TyShak II balloon	18mm Amplatzer multimesh ASD occluder
4	DORV- VSD, dextrocardia	Atrio-pulmonary connection	10	12	desaturation	7x20 mm TyShak II balloon	18mm Amplatzer multimesh ASD occluder
5	DILV- VA discordance	Atrio-pulmonary connection	2	144	desaturation	7x20 mm TyShak II balloon	6x4mm Amplatzer ductal occluder-II
6	Mitral atresia- DORV	Atrio-pulmonary connection	10	19	desaturation	8x20 mm Ovypka balloon	18mm MemoPart multimesh ASD occluder
7	TA-VA discordance	Extracardiac conduit	5	40	desaturation	8x20 mm Ovypka balloon	18mm Occlutech multimesh ASD occluder
8	TA-VA discordance	Extracardiac conduit	6	11	desaturation	7x20 mm TyShak II balloon	7.5mm Occlutech flexi ASD occluder
9	LAI- Mitral atresia- DORV	Extracardiac conduit	6	1	embolism	5x15mm NC coronary balloon	18mm Occlutech multimesh ASD occluder
10	TA-VA discordance	Extracardiac conduit	5	1	desaturation	7x20 mm TyShak II balloon	12mm Amplatzer vascular plug-1 & 28mm CP covered stent
11	TA- pulmonary atresia	Extracardiac conduit	4	26	desaturation	7x20 mm TyShak II balloon	18mm Amplatzer multimesh ASD occluder
12	LAI- Mitral atresia- DORV	Atrio-pulmonary connection	5	34	desaturation	5x15mm NC coronary balloon	18mm Amplatzer multimesh ASD occluder
13	TA-VA discordance	Extracardiac conduit	11	5	desaturation	6x20 mm TyShak II balloon	18mm Amplatzer multimesh ASD occluder
14	TA-VA discordance	Extracardiac conduit	5	18	desaturation	5x20 mm TyShak II balloon	18mm Amplatzer multimesh ASD occluder
15	DORV- VSD	Extracardiac conduit	15	38	suspected embolism	6x20 mm Ovypka balloon	16x18mm Occlutech PFO occluder

ASD: atrial septal defect; DILV: double-inlet left ventricle; DIRV: double-inlet right ventricle; DORV: double-outlet right ventricle; LAI: left atrial isomerism; PFO: patent foramen ovale; TA: tricuspid atresia; VA: ventriculo-arterial; VSD= ventricular septal defect.

PP-298
Transcatheter salvage in a pediatric patient with isolated unilateral absence of left pulmonary artery

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Background and Aim: The unilateral absence of a pulmonary artery (UAPA) stands as a rare congenital anomaly within the spectrum of cardiovascular conditions. Its complex nature involves the underdevelopment or complete absence of one of the pulmonary arteries, often presenting either in isolation or with other congenital cardiac abnormalities. Manifestations can vary widely, ranging from asymptomatic cases to a spectrum of symptoms such as chest pain, exercise intolerance, dyspnea, hemoptysis, and recurrent pulmonary infections. Herein, we report a 9-year-old girl with isolated left pulmonary artery absence and succesfull correction after PDA-LPA recanalisation.
Method: CASE: A 9-year-old patient initially diagnosed during pneumonia treatment three years prior, the young girl had a history of recurrent pneumonia, leading to further investigations. Echocardiography revealed a peculiar absence of the left pulmonary artery, while the right pulmonary artery stemmed directly from the main pulmonary artery. This uncommon scenario prompted deeper exploration through various imaging techniques
Results: Contrast-enhanced computed tomography(CT) played a pivotal role in confirming the absence of the left pulmonary artery

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while highlighting the normalcy of the bronchial structures. Subsequent assessments, notably via right heart catheterization and aortic angiography, unveiled additional complexities and revealed mean pulmonary artery pressure15 mmHg. The examination identified a ductus diverticulum originating from the left innominate artery, offering a potential pathway for interventions. The medical team opted for an innovative approach, aiming to reestablish connection to the absent left pulmonary artery through the recanalized ductus. A 0.014 inch extrasupport coronary guidewire was advanced through the 5.2F JR4 catheter from ductus arteriosus to left pulmonary artery. Dilatation was performed with 3.5x15 mm and 4x20 mm coronary balloons and 4.5x16 mm coronary stent delivered through a 5F guiding catheter was placed in the PDA-LPA junction to sustained blood supply. Subsequent evaluations exhibited promising progress in the development of the left pulmonary artery, eventually leading to corrective surgery. The patient successfully recovered and was discharged
Conclusions: Isolated UAPA is very rare and the diagnosis is very challenging if patients are asymptomatic. CT and catheter angiography helped us to demonstrate distinct Anatomy. Therefore, if the patient is not suitable for surgery for the first approach, percutaneous interventions may be life-saving.

Keywords: absence of left pulmonary artery, recanalisation,stenting of ductus arteriosus

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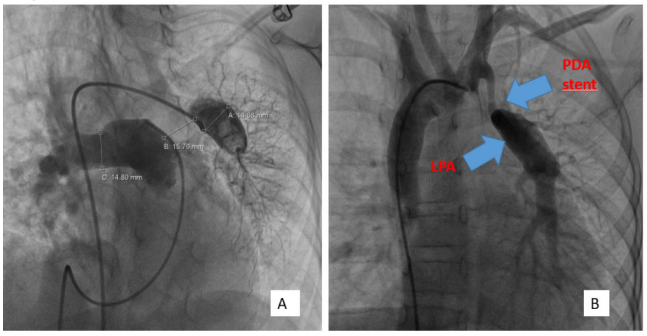

***A:** Distance between LPA-RPA on catheter angiography, **B:** Angioagraphic image after PDA-LPA stenting. LPA: left pulmonary artery, PDA: patent ductus arteriosus, RPA: right pulmonary artery*

PP-299

Venus P-valve implantation in patients with pre-existing stents in the left pulmonary artery branch: A case-series

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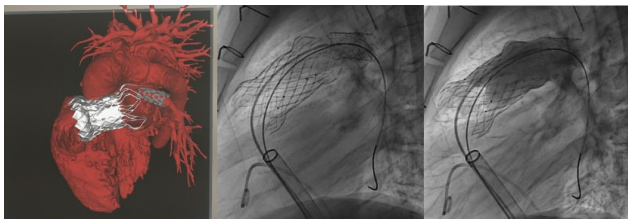
Background and Aim: Venus P-valveTM (Venus Medtech, Shanghai, China) is a self-expandable bioprosthetic valve that can be transcatheter-implanted in patients with severe pulmonary insufficiency and dilated right ventricular outflow tracts (RVOT). Valve implantation is technically challenging because of the high anatomical variability of the RVOT and pulmonary artery (PA), for this reason, in order to guarantee valve stability during and after deployment, the long-sheath delivery system is usually anchored into the left PA (LPA). Consequently, LPA stenting is a relative contraindication to Venus P-Valve implantation. In this case series we describe our experience and the use of a holographic planning of the procedure.
Method: Three patients with a diagnosis of severe pulmonary insufficiency after Tetralogy of Fallot repair with transannular patch and LPA stents underwent Venus P-Valve implantation at our centre. Two patients had a pre-existing LPA stent, while one patient underwent LPA stenting and Venus-P valve implantation in the same sit. All patients underwent pre-operative ECG-gated CT scan, CT images were analysed by Medtech specialists yielding an estimated valve size. The same images were then used to create three-dimensional (3D) holographic models (Artiness, Milan, Italy) of the entire heart with a focus on RVOT and PA anatomy. The holographic models were used for planning valve implantation and to assess the relationship of the valve to the main PA, its branches and the stent (Figure 1). During holography-guided pre-procedural planning it was agreed to deploy the valve by anchoring the delivery system into the right pulmonary artery (RPA).
Results: During the procedure, the valve was deployed starting from the main PA trunk below the LPA stent in order to avoid valve infolding, impingement and dislocation. After valve deployment, stent patency was confirmed with a pulmonary angiography. A post operative transthoracic echocardiography showed a properly functioning valve with no paravalvular leaks.
Conclusions: this case-series underscore the effectiveness of Venus P-Valve implantation in challenging anatomical scenarios such as patients with LPA stents; we also believe that the holographic planning may help the operators.

Keywords: holography, venus p valve, pulmonary insufficiency, pulmonary artery, pulmonary artery stenting

1



Holographic reconstruction and pre-procedural planning of valve implantation and angriographic post-procedural results in the same patient.

PP-300

Stenting of obstructed fontan conduit

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Background and Aim: BACKGROUND: Obstruction of Fontan pathway is a common late complication. Progressive stenosis of Fontan anastomosis accelerates liver fibrosis. Different materials used in Fontan conduit have different mechanisms of obstruction. AIMS: The aim of the study was evaluation of usefulness and safety of obstructed Fontan conduit stenting.
Method: Retrospective, single-center analysis of Fontan patients with conduit obstruction.
Results: Thirty eight patients (28 (74%) male) with extracardiac Fontan conduit stenosis underwent cardiac catheterization with conduit stenting. Mean age during procedure was 16 years

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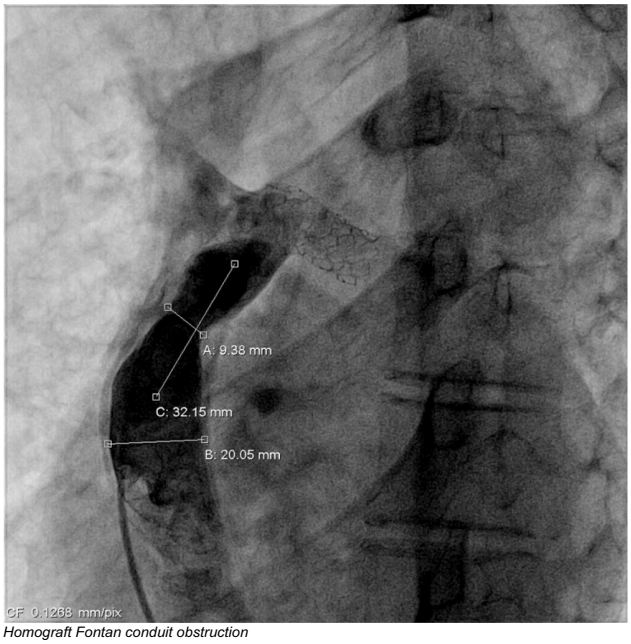
(7-26 years), mean time from Fontan operation was 10 years (1-21 years). Mean age at Fontan procedure was 6 years (2-12 years). In 16 patients homografts (17-22mm), in 20 patients Gore- Tex (18-21mm), were used to create Fontan conduit, two patients had direct cavopulmonary connections. Homografts showed diffuse calcification leading to focal stenosis, Gore-Tex were obstructed by stretching of conduit due to patient's growth.
14 patients had hypoplastic left heart syndrome (HLHS), 12 tricuspid atresia, remaining 12 patients had different forms of univentricular heart. All patient had liver fibrosis, 3 of them had cirrhosis, one patient had history of protein losing enteropathy (PLE), two of plastic bronchitis (PB).
The mean conduit smallest diameter was 11±1,8 mm before stenting.
Covered Cheetham Platinum stents were placed in 37 patients, in 1 patient Bentley Be Graft aortic 18x35 mm was implanted. Stenting was successful in all patient reaching conduit diameter above 18mm with no complications.
Stenting of obstructed Fontan conduit was the most common intervention in adult Fontan patients.
Conclusions:

1. Fontan patients need accurate and methodical follow-up including hemodynamic data.
2. Obstruction of Fontan pathway is common late complication.
3. Percutaneous stenting is safe and effective method relieving Fontan obstruction.
4. Different materials used in Fontan pathways have different mechanisms of obstruction.

Keywords: Fontan pathway obstruction, stenting, Fontan complications



Homograft Fontan conduit obstruction

PP-301

Closure of large patent ductus arteriosus with novel konar tm multifunctional occluder in children less than 3 years

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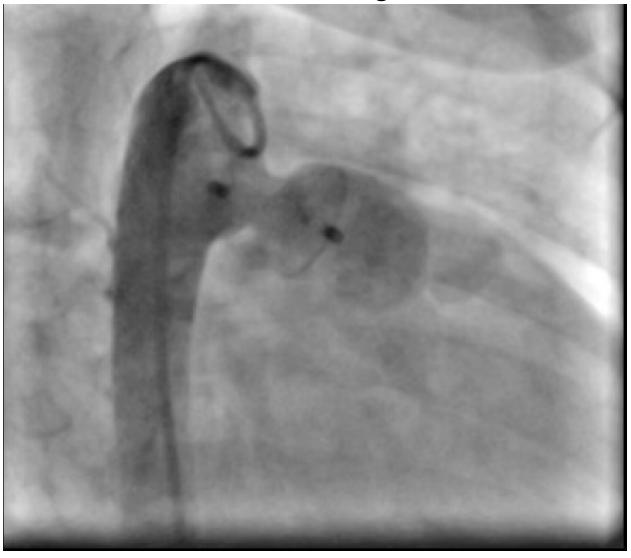
Background and Aim: Incidence of PDA range between 1.5 to 3.5 and 0.5 to 1 per 1000 live births. Infants with large PDA are symptomatic and need early closure. Percutaneous closure of patent ductus arteriosus (PDA) in children is an established procedure. However, few studies are available on usage of MFO for occlusion in large PDA's in children.
Method: A total of 14 children underwent closure of large PDA from March 2019 to March 2023 using MFO. Procedure was done under fluoroscopy and transthoracic echocardiography (TTE) guidance.
Results: A total of 14 children, eight male (57.2%) and 6 female(42.8%) underwent procedure. Mean age was 19.8 ± SD (9.0 - 30.6) months and mean weight 8.8 ± SD (6.4 - 11.4) kgs of patients. Mean ductal diameter was 7.6mm ± SD (6.4 – 8.8mm). Successful closure was possible in 13 (93%) cases. The one failed PDA procedure was due to large window type duct with significant residual flow. Pulmonary artery systolic pressure (PAsP) was 84.7 ± SD (78.5 - 90.9) mmHg which decreased to 40.4 ± SD (34.9 - 45.9) mmHg. A fall in Pulmonary artery mean pressure (PamP) from 44.0 ± SD (37.5 - 51.5) mmHg to 39.6 ± SD (34.4 - 44.8) mmHg was noticed. Significant residual shunts were seen in 5(35%) patients. Complete disappearance of residual shunt was seen in 4 patients after 96 hours. One patient continues to have small residual shunt after 6 months. No haemolysis was noticed in major residual shunt patients. All patients were followed for mean of 16 months ± SD (12.8 - 19.2months). The mean fluoroscopy time for PDA closure was 8.2 mins ± SD (7.5 - 9.1 mins).
Conclusions: MFO device can be safely and effectively used to close large PDA's in children. No serious adverse events were noticed in short term follow-up.

Keywords: Patent ductus arteriosus, transcatheter closure, KONAR-MFO

Post MFO device closure of Large PDA



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PP-302

Percutaneous balloon pulmonary valvuloplasty in congenital pulmonary valve stenosis: A ten-year experience and follow-up

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Background and Aim: Isolated pulmonary valve stenosis (PVS) is found in 8-10% of patients with congenital heart disease. Percutaneous balloon pulmonary valvuloplasty (PBPV) is the interventional procedure treatment of choice. After the procedure most patients have some degree of pulmonary regurgitation (PR). Since few studies have reported long-term results of this technique in the pediatric population, the aim of this study was to characterize PBPV and determine its immediate effectiveness and long-term results.
Method: A retrospective study was performed including patients with isolated PVS undergoing PBPV at a single tertiary center between January/2001 and December/2011. Immediate and long-term results were assessed.
Results: The study included 63 patients, 40 (63%) females and 23 (37%) males. Over the period of review, 65 procedures were performed. The median age at the time of the procedure was 1,5 (range: 0,03–16) years with a median weight of 11 (range:1,9–54,5) kilograms. PVS severity was assessed by echocardiogram in 21 (32%) moderate, 37 (57%) severe and 7 (11%) critical stenosis. The median balloon diameter was 14 (range:5–25) mm. Immediate success was achieved in 56 (86%) procedures. Only 3 (5%) patients presented transient arrhythmia. Median length of hospital stay was 2 (range:1–17) days. Follow-up results were obtained in 63 patients along 10 years. Fifty-eight (92%) developed some degree of PR, 26 (45%) mild, 23 (40%) moderate and 9 (15%) free regurgitation. Twenty-one (33%) patients developed right ventricle dilatation, however only 2 were moderate and none had right ventricle dysfunction. Ten years after procedure, 7 (11%) patients presented moderate PVS. There were no deaths after procedure or during follow-up. Procedures performed at a younger age (≤1 year of age), lower weight (≤5 kg), severe or critical PVS before and moderate PVS after PBVP were all related with statistical significance to moderate and free PR, right ventricle dilatation and moderate PVS. Balloon size was not related to adverse echocardiographic findings.
Conclusions: PBVP is an effective and safe treatment of isolated PVS with only one patient needing reintervention. The majority of patients developed PR, however 10 years after procedure only a few presented with moderate PVS. Patients requiring treatment early in life are more likely to present these changes.

Keywords: pulmonary valve stenosis, percutaneous balloon pulmonary valvuloplasty, pediatric

PP-303

Our perspective on infective endocarditis after transcatheter pulmonary valve implantation: *Can we do better*

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Background and Aim: Nowadays, a widely used interventional procedure of transcatheter pulmonary valve implantation (TPVI) has undoubtedly improved the management of right ventricular outflow tract dysfunction. However, it entailed the development of infective endocarditis (IE) as a possible complication, which may further impair valve durability and deteriorate the final outcome. Our research was conducted with the aim of determing the major risk factors for infective endocarditis along with an overall review of treatment approach in order to improve our current knowledge.
Method: We conducted our retrospective research on 17 patients, with congenital heart defects (CHD) such as Tetralogy of Fallot (TOF), pulmonary atresia (PA), truncus arteriosus and double outlet right ventricle (DORV), who underwent TPVI using the Melody or Sapien valves, in period from 2013-2023. We determined the frequency of IE, its risk factors, Duke criteria, entry sites of bacterial infection and microbiological agents, reintervention procedures, as well as the type of performed treatment. We also took into account the post-implantation ultrasound findings, along with the type and diameter of used prosthetic material.
Results: A total of 17 patients, 14 males and 3 females, mean age 14,1 years at the time of TPVI, 16 of them received Melody valve and only one Sapien valve. The most common CHD in observed patients were conotruncal anomalies: TOF (35,2%), truncus arteriosus (11,8%) and DORV (11,8%), while PA with ventricular septal defect counted 23,5%. IE was confirmed in 23,5%, each of them had mild residual pulmonary stenosis after TPVI and previous infectious episodes (osteomyelitis, respiratory infection) or skin wounds (25%). *Staphylococcus aureus* was the main causative organism (75%), following by *Streptococcus viridans* (25%), confirmed by at least two positive blood cultures. Surgery was performed in 29,4% patients, while transcatheter valve dilatation needed 11,8%, mostly because of IE. In further follow-up, 1 patient died due to the terminal stage of right-sided heart failure with multiorgan dysfunction syndrome.
Conclusions: Our data confirmed that infective endocarditis significantly affects valve durability, the need for reintervention and the success of the final outcome. A multidisciplinary approach is therefore crucial along with improvement of residual transpulmonary gradient and early antimicrobial prevention of the infective endocarditis.

Keywords: Interventional cardiology, Pulmonary valve, Melody valve, Infective endocarditis, Duke criteria, Congenital heart disease

PP-304

Transcatheter bilateral endoluminal pulmonary banding and ductal stenting as bridge to biventricular yasui repair in border line left heart

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Background and Aim: Delaying surgery beyond the neonatal period can mitigate adverse neurological outcomes in complex congenital heart disease. Novel transcatheter approaches using endoluminal pulmonary artery (PA) banding in combination with ductal stenting (DS) may help.
Method: Here, we present a patient (3290g, 53cm) with aortic atresia, hypoplastic aorta, borderline left ventricle (left ventricular

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enddiastolic diameter (LVEDD) 13.9mm, z -2,8; mitral valve (9mm, z -1), restrictive foramen ovale, inlet ventricular septum defect (VSD), duct dependent arch perfusion and left partial anomalous pulmonary venous connection (PAPVC). At day 10 of life, DS (Sinus-SuperFlex-DS, 8x18mm, Optimed, Europe) placement of bilateral modified micro-vascular plugs (MVP 7Q, Medtronic, Europe) for restriction of pulmonary blood flow (endoluminal banding) and Rashkind procedure ensured a balanced pulmonary, retrograde arch and systemic circulation. Five weeks later, balloon dilation (BD) of the right MVP 7Q (3x20mm PTCA balloon) was required due to too much restriction. Coarctation required isthmus stenting (Synergy Megatron 5x8mm) and re-BD of the DS six weeks later. 3D reconstruction of computer tomography (CT) helped to plan surgical biventricular correction. At week 17 (4500g, 57cm) Yasui operation with DKS anastomosis, aortic arch patch expansion, VSD tunneling to neoaorta, correction of PAPVC and implantation of a 12mm RVPA conduit was successfully performed. Subsequent stenosis of right and left PA required additional BD and stenting (Formula 6x12mm, CookMedical, Europe) two weeks later. Ultimatively, extubation was achieved 4 weeks postoperatively. The patient was discharged after 6 weeks. Yasui tunnel revision with Gore-Tex patch placement, VSD patch enlargement, removal of the RPA stent and replacement of the 12mm contegra conduit was undertaken at the age of 7 months.
Results: At follow-up at 18 months of age, the patient was in good clinical status with good neurological development and growth (7100g, 67cm). Echocardiography demonstrated adequate ventricular shaping (LVEDD 23.4mm, Z 0).
Conclusions: Combined interventional and surgical strategies for complex congenital heart disease is nowadays essential for a positive long term outcome. A "No Norwood, no hybrid" approach may be used as bridge to complex biventricular repair beyond the neonatal period. Careful interstage monitoring (echocardiography, CT, 3D reconstructions) of pulmonary and retrograde aortic blood flow is crucial.

Keywords: endoluminal banding, catheter, border line left heart, ductal stenting

PP-305
Impact of rapid ventricular pacing on short- and long-term outcomes of balloon aortic valvuloplasty
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Background and Aim: Maintaining catheter stability is a key consideration in interventional procedures like balloon aortic valvuloplasty (BAV). To achieve this Rapid Ventricular Pacing (RVP) is commonly utilised to ensure that the transcatheter balloon remains stable. Though that practice is routinely applied in BAV, the data supporting its effectiveness in reducing aortic regurgitation (AR) is mixed and often limited to short-term observations. This study seeks to assess the short and long-term outcomes of BAV facilitated by RVP with the aim of clarifying its practical clinical benefits.
Method: Between 1998-2023 total number of 170 patients underwent primary BAV in our centre, of which 76 pts (7.4+/-6.2 years) were included. Inclusion criteria were: primary BAV and good left ventricle (LV) function. Neonates and young infants (under 6 months of age) were excluded from the study. RVP was used in 26 pts (33.8%), the decision whether to use it was operator dependent. Patients with vs. without RVP use did not statistically differ in weight, morphology of AV, preprocedural gradient or regurgitation. Pacing rate was chosen to decrease the LV pressure by >50%, measured hemodynamically. Long term follow-up up to 20 years (mean 6.6 +/- 6.) was available in 54 pts (71%).
Results: Early results of BAV did not reveal a positive effect of RVP. Significant (≥moderate) AR occurred in 6 pts (23.1%) with RVP vs 7 pts (15.2%) without RVP ($p>0.05$). Percentage gradient change was 55.3% with RVP vs 57.7% without ($p>0.05$). Overall 55.1% of patients required reintervention at median 9.2 years (95% CI 7.25 - 12.6). Interestingly, median time to reintervention was 5.3 years (95% CI 2.58 - 8.83) in patients with RVP vs 12.6 years (95% CI 8.3 - 20.7) in patients without. Freedom from reintervention at 5 years was 70.2% overall, 55% in patients with RVP and 81% in patients without.
Conclusions: Our study did not prove a positive effect of RVP on short and long-term outcomes of BAV. Large, prospective and randomised trial in the adult population showed no effectiveness of RVP. To assess the effectiveness of RVP in the paediatric population a larger, prospective trial is needed.

Keywords: Aortic stenosis, Balloon aortic valvuloplasty, Interventional cardiology, Rapid Ventricular Pacing

PP-306
Transcatheter closure of patent ductus arteriosus in infants with weight under 5 kg
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Background and Aim: Persistent patent ductus arteriosus (PDA) in preterm infants is associated with poor respiratory outcomes and an increased mortality. Medical management with non-steroidal anti-inflammatory drugs remains the first line of therapy in closing the PDA, second line treatment is surgical ligation or transcatheter percutaneous closure (TCPC). To evaluate the effectiveness of TCPC of the PDA in terms of feasibility, procedural safety and efficacy in preterm infants and term with major comorbidities, weighing <5Kg. *Method:* Between January2022 and June2023 8 patients with PDA weighting under 5kg were submitted to TCPC in the pediatric cardiology center of Padua. TCPC was performed after clinical finding of hemodynamically significant ductus arteriosus in preterm infants after execution of pharmacological therapy and hemodynamically significant duct in those born to term. Pre-operative evaluation was based on transthoracic echocardiography (TTE), intra–operative evaluation based on both TTE and fluoroscopy, post-operative evaluation (24 hours after), based on TTE took into consideration any residual shunt and the interference of the device with the flow both in the aorta and in the left pulmonary artery branch. Devices used: Amplatzer Piccolo Occluder and Occlutech PDA Occluder. *Results:* The procedure was 100% effective, 24h after TCPC no residual shunt or interference with the blood flow was detected. Patient weight at the time of the procedure: 4:<2kg (average weight 1.37, average G.A.24.95); 4:3-5kg (average weight 3.9, average G.A.36.5). Premature had earlier TCPC (34.7 vs 109.2 days). 100% G.A.<30 pharmacological closure was attempted before TCPC (mean cycles=3). Piccolo was used in 6 patients, occlutech in 2.75% had solo venous approach; 25% both venous and arterial approach. Disk-diameter/duct minimum-diameter ratio was 1,22 for <2kg and 1,96 for the 3-5kg group ($p<0.01$).

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Conclusions: In low weight, the Piccolo allows to use only the venous approach with 4Fr introducer, minimizing the thrombotic risk and vascular damage. In patients with comorbidities (ex-premature, corrected diaphragmatic hernia, broncho-dysplasia), reduced lung compliance may require early correction. In this group, TCPC is preferable to thoracotomy. In patients weighing >2kg, a higher device/PDA diameter ratio is required to obtain adequate stability; as for the entirely intra-ductal implantation technique, it is to be weighted on a case-by-case basis.

Keywords: patent ductus arteriosus (PDA), prematurity, trans-catheter closure, low weight preterm infants, percutaneous closure

PP-307

Advantes of using a torqvue LP catheter for pda stenting in newborn with duct dependent pulmonary circulation

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Background and Aim: Arterial duct stenting is accepted as a reliable alternative to surgical palliation in patients with duct-dependent pulmonary circulation(DDPC) congenital heart disease. In clinical practice, coronary stents are used, requiring a low profile, a 4Fr introducer or a 5Fr guide-catheter. The choice between less support and greater invasiveness of vascular access is crucial; on one hand, the risk of procedural failure increases, on the other, the risk of vascular damage increases, in subjects of low weight. To illustrate the advantages of using TorqVue LP for both the angiographic and the implant phase, to reduce manipulations and complication associated to procedure.
Method: Between May2023 and September2023 in the pediatric cardiology center of Padua 2 coronary stent implants were performed using LP-catheter in newborns with DDPC and complex ductal anatomy. Pre-operative evaluation was based on transthoracic echocardiography (TTE), intra–operative evaluation based on fluoroscopy, post-operative evaluation (24,48h after procedure), based both clinical and TTE evaluation: as clinical was considered the blood oxygen saturation; in TTE evaluation was considered stent positioning in terms of: intra stent blood flow, flow obstruction or presence of turbulences of new onset of blood flow both on the aortic and pulmonary side. The coronary devices were Rebel Stent.
Results: Both patients had a weight of <3.5kg. Vascular approach: first case femoral artery; left carotid artery in the other case Ductal morphology was tortuous in both cases. Stents used: 4x20mm; 3.5x16mm. Post-operative evaluations at 24-48h: TTE showed stents well positioned with laminar flows within them and no obstruction to aortic and pulmonary flows. Blood oxygen values were within normal limits for heart disease.
Conclusions: In our experience, the use of LP-catheter combines supportivity and low-profile of vascular access and for this reason a lower traumaticity on the vessel. A further advantage is the possibility of the contextual execution of angiographs during stent placement which can guarantee fast procedural times and a better control of the position of the stent itself. In addition, in the case of complex ductal anatomies, the use of the LP-catheter and micro-catheter with the "mother-in-child" technique makes implantation possible even in patients where other techniques would not allow it.

Keywords: ductal dependent pulmonary blood flow, congenital heart disease, arterial duct stenting, TorqVueTM LP, PDA stent

angiogram and stent deployment

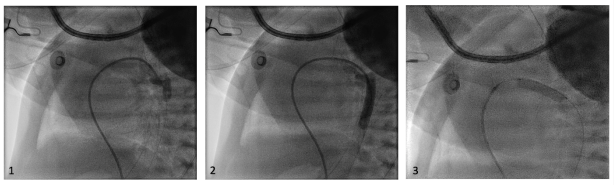

Figure 1 and 2: Angiography performed with 4 Fr TorVue LP catheter
Figure 3: Coronary Rebel stent implanted using 4Fr TorVue LP catheter

PP-308

Use of a mechanical crimper for mounting covered stents in aortic coarctation

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Background and Aim: Despite the continued evolution in use of covered stents in congenital cardiology, no standard technique is available for crimping stents onto balloons. We aimed to describe the use of a mechanical crimper to mount covered stents and its effects on stent integrity, uniformity, and sheath size for intervention.
Method: We present a single-center retrospective review of patients with coarctation of the aorta (CoA) undergoing stent angioplasty with covered stents between January 2019-December 2022. Sheath size used for intervention was recorded and compared to the manufactures' IFU. A bench testing model was then performed to confirm the decrease in sheath size by mechanically crimping two different covered stent models. Percent reduction in profile after crimping, stent uniformity, and PTFE integrity after balloon inflation were recorded.

Figure 1A. Bench test results

Stent	Mean diameter, mm (Uniformity index)		AVG% Reduction	IFU recommended sheath size	Sheath Size attempted	Stent successfully loaded into and passed through the sheath?	Comment on stent and sheath interaction
	Before mechanical crimping	After mechanical crimping					
A#	3.88 (0.07)	.	.	14	12	No	Covering damaged; unable to pass
B	3.52 (0.13)	3.00 (0.13)	14.8	14	12	Yes	Intact- easy
C	4.78 (0.19)	4.12 (0.16)	13.2	14	12	Yes	Intact- easy
D	5.35 (0.13)	4.78 (0.03)	10.3	18	16	Yes	Intact- easy
E*	5.46 (0.02)	4.14 (0.11)	27.1	16	14	Yes	Intact- easy
F*	4.94 (0.04)	3.6 (0.21)	24.2	14	12	Yes	Intact- easy

Figure 1B. Visual changes before and after mechanical crimping

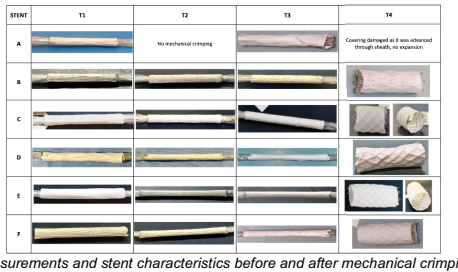

*Figure 1A. Measurements and stent characteristics before and after mechanical crimping. Stent A was a premounted covered CP (8 zig x 3.9 cm) on a 12 mm x 3.5 cm BIB®; Stent B a Premounted covered CP (8 zig x 3.4 cm) on a 22 mm x 3.5 cm BIB®; Stent C a Premounted covered CP (8 zig x 3.4 cm) on a 24 mm x 3.5 cm BIB®; Stent D a Covered G Armor (10 zig x 6.3 cm) on a 34 mm x 5.5 cm BIB®; Stent E a Covered G Armor (10 zig x 6.8 cm) on a 22 mm x 5.5 cm BIB® and Stent F was a Covered G Armor (10 zig x 6.3 cm) on a 22 mm x 5.5 cm BIB®. #Manually crimped control, no mechanical crimping. *Mechanically crimped without prior manual crimping. AVG: average percent reduction in the diameter of the stent after mechanical crimping. Figure 1B. Visual description of stents at determined times: before mechanical crimping (T1), immediately after mechanical crimping (T2), after advancement through a sheath 2 sizes smaller than that recommended by the IFU (T3) and after balloon dilation (T4).*

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Results: A total of 25 clinical events were identified for review. Median age at procedure was 18 years and the most common diagnosis was isolated CoA (80%). 76% (n=19) of stents were mechanically crimped and 24% (n=6) were manually crimped. The median sheath size for mechanically crimped stents was −2 Fr compared with a median of 0 Fr for manually crimped stents (p=0.007). Bench testing revealed a median 12.8% reduction in stent diameter after mechanical crimping. All mechanically crimped stents were successfully introduced through sheaths 2 Fr smaller than the IFU during bench testing, correlating with the retrospective clinical experience.
Conclusions: The use of mechanical crimpers for mounting covered endovascular stents allows the delivery of these devices through smaller profile sheaths facilitating intervention in smaller patients and reducing the risk of vascular access related injuries without affecting stent performance.

Keywords: Congenital heart disease, coarctation of the aorta, mechanical crimping.

PP-309

Konar mf occluder device: versatile application in a single-center experience

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Background and Aim: The LifeTechTM Konar-MF VSD Occluder is a versatile closure device primarily designed for ventricular septal defects (VSD). Despite limited evidence in the literature, our experience showcases its adaptability in diverse cardiac conditions, and the results in the short and long term.
Method: In this retrospective study, outcomes of 71 patients undergoing Konar-MF Occluder implantation for diverse cardiac conditions between March 2019 and August 2023 were analysed. Conditions targeted included VSD, patent ductus arteriosus (PDA), pulmonary, and coronary fistulas, with patients exhibiting signs of hemodynamic impairment on echocardiogram, whether symptomatic or asymptomatic.
Results: Among the 71 patients, 80.3% had VSD (mostly perimembranous), 12.7% had PDA, and 7% had other conditions. Median age at procedure was 15 years, and median weight was 47 kg. The median procedural time was 150 minutes, and the median fluoroscopy time was 63.5 minutes. Implantations, accomplished through a retrograde (51%) or antegrade (22.5%) approach or through an arteriovenous loop formation (22.5%), demonstrated a procedural success rate of 84.5%. Major complications, mainly in VSD closures, occurred in 15.5%, due to unsuitable defect size or embolization, and prompted surgery in 8.5%. In particular, the rate of embolization was 7% but in most cases the device was easily retreated, with only one instance causing tricuspid valve damage. Minor complications included mild tricuspid valve interference and transient rhythm abnormalities (one case of intra-procedural intermittent left bundle branch block, and only one case of transient advanced AV block). Follow-up at a median time of 11.93 months involved 63.4% of patients. In 89% of follow-up cases, procedural success persisted without complications. Complications occurred in 5 cases, including residual interventricular shunts and a coronary fistula aneurysm requiring surgical closure. Notably, only 2 cases of VSD closure had late rhythm complications, with the presence of nocturnal junctional rhythm phases recorded by periodical ECG Holter monitoring.
Conclusions: The LifeTechTM Konar-MF Occluder proves to be a valuable and adaptable device for VSD closure, including the perimembranous type, as well as for addressing PDA and fistulas, as demonstrated in our experience. Follow-up indicates sustained procedural success with a low incidence of electrical and mechanical complications.

Keywords: VSD, coronary fistula, pulmonary fistula, PDA

PP-310

Novel hybrid approach to arterial duct calibre reduction

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Background and Aim: Occasionally complex congenital cardiac cases are encountered where a large arterial duct (PDA) ideally needs to be smaller, but not completely occluded. We report a novel approach to reduce PDA calibre by hybrid banding and stenting in series of 3 cases.
Method: Technique: The procedure is performed via femoral vascular access and median sternotomy. Following angiographic assessment, the PDA is crossed with a coronary guidewire and coronary balloon of the desired size is inflated in the duct. Surgical banding of the duct is then performed, tightening a Gortex ligature around the duct onto the balloon. The balloon is then deflated and removed, followed by PDA stenting to maintain ductal patency. A stent larger than the original balloon is used, which forms a waist at the site of the band corresponding to the original desired size.
Results: Case 1: 1 year old, ex-preterm (28/40) with chronic lung disease and large PDA with bidirectional flow. Concerns regarding PDA occlusion given PHT, although anticipated improvement in lung disease with time and desire to reduce pressure load on pulmonary vasculature. Hybrid ductal reduction undertaken and on last follow-up, ductal shunt is entirely left to right.
Case 2: 32+2-week gestation (birth weight 1.88Kg) with TGA and complex coronary anatomy. Hybrid ductal reduction undertaken to allow growth, whilst maintaining saturations and LV loading off prostaglandin infusion. Has subsequently had successful Arterial Switch Operation (ASO).
Case 3: 35+4-week gestation (birth weight 2.2Kg) with TGA, bilateral superior vena cavae, juxtaposed atrial appendages and complex drainage of the left coronary venous system. Given the complexity of the anatomical set up, hybrid ductal reduction undertaken to again allow growth with subsequent successful ASO.
Conclusions: Hybrid banding and stenting of the arterial duct is a novel technique which has been successful employed in isolated

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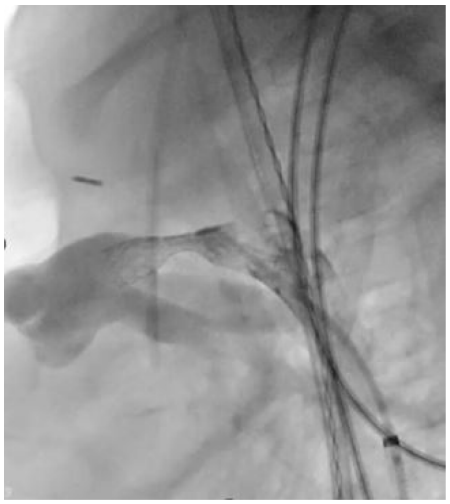
cases where a large PDA needs reduction in size, but not complete occlusion.

Keywords: Arterial duct, Transposition of great arteries, Pulmonary Hypertension

Arterial duct calibre reduction



Fluoroscopy guided image showing banding and stenting of arterial duct.

PP-311
Real-time biventricular pressure-volume loops during percutaneous pulmonary valve implantation in patients with Rvot dysfunction

Heiner Latus[1], Verena Schindler[2], Julie Cleuziou[3], Markus Khalil[4], Christian Jux[4], Christian Meierhofer[2], Daniel Tanase[2], Andreas Eicken[2], Peter Ewert[2] and Stanimir Georgiev[2]
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Background and Aim: In patients with RVOT stenosis and/or pulmonary regurgitation, percutaneous pulmonary valve implantation (PPVI) aims to preserve right (RV) and left (LV) ventricular integrity and function. The best timing for PPVI as well as underlying mechanisms for sufficient ventricular recovery, however, are not yet adequately understood. Our study thought to assess acute changes in biventricular intrinsic myocardial function occurring with PPVI.
Method: Twenty patients with RVOT dysfunction (age 23.0 ± 10.9 years, 9 females, mean peak echocardiographic RVOT gradient 64 ± 25 mmHg) underwent PPVI with biventricular assessment of pressure-volume loops using conductance catheter technique. Load-independent parameters of systolic (Ees) and diastolic (Eed) function as well as pulmonary/systemic arterial elastance (Ea) and V-A coupling (Ea/Ees) were assessed before and directly after PPVI. Cardiac magnetic resonance (CMR) for quantification of biventricular volumes, function and pulmonary regurgitation (PR) was also performed.
Results: With PPVI, both RV Ees (p=0.036) and pulmonary Ea (p=0.0002) decreased significantly while right V-A coupling (p=0.76) remained impaired. LV Ees (p=0.68) and left V-A coupling (p=0.98) were not affected by PPVI although systemic Ea increased significantly (p=0.03). Both RV (p=0.37) and LV (p=0.20) Eed showed no significant change with PPVI. Patients with relevant PR (≥25%, n=10) had lower RV Ees (p=0.03) before and higher LV Eed (p=0.01) after PPVI as compared to patients with minor PR (<25%, n=10) whereas V-A coupling was similar between the two groups.
Conclusions: Acute unloading of the RV by PPVI is accompanied by an instantaneous decline in RV contractility with persistent abnormal V-A coupling. The LV adequately adapts to an increase in pre- and afterload with unchanged LV intrinsic function and V-A coupling. The relevance of these response patterns on long-term biventricular remodeling require further investigation.

Keywords: Percutaneous pulmonary valve implantation (PPVI), systolic ventricular function, diastolic ventricular function, pressure-volume loops

PP-312
Catheter-based rescue intervention for massive life threatening pulmonary thromboembolism in a preterm neonate

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Background and Aim: Massive pulmonary thromboembolism (PTE) is an uncommon condition, presents with sudden cardiorespiratory collapse and is associated with high mortality, reported in up to 65% of the patients and occurring within the first hours after start of symptoms in 70%. Immediate rescue therapy is therefore pivotal. Treatment strategies for adult patients may be extrapolated to children but are limited by feasibility and age-specific adverse effects in neonates.
Method: We describe a case of a preterm neonate with massive pulmonary embolism associated with acute cardiopulmonary deterioration due to absent pulmonary circulation and suprasystemic right ventricular pressure.). Thrombolytic therapy in the preterm neonate is associated with seriously increased risk of massive intracranial hemorrhage and would not restore circulation in time to prevent severe organ damage or death. Surgical thrombectomy has a high mortality rate in preterm infants and carries the risk of embolization and redistribution of thrombus. Rescue extra-circulatory life support in preterm neonates has increased risk of intracranial hemorrhage and is associated with size-related specificities. Catheter-based intervention was therefore deemed the only viable acute therapeutic intervention in this preterm neonate. We performed repeated manual thrombosuction, followed by repeated balloon dilatations in both RPA and LPA, aiming at maceration/compression of the residual thrombus at both sides. Subsequently, we decided to proceed with targeted delivery of recombinant tissue plasminogen activator, followed by subsequent systemic thrombolysis.
Results: The infant fully recuperated without complications and during follow-up of 26 months showed no signs of pulmonary hypertension or chronic thromboembolic vascular disease. Treatment and follow-up regimens for this type of patients are insufficiently defined.
Conclusions: Massive pulmonary embolism is rare in infants and is associated with a very high mortality rate. Due to advanced treatment options in preterm neonates and cumulative risk factors prevalence of massive PTE has increased in this population. As cardiorespiratory insufficiency and death occur rapidly after massive PTE, early recognition and urgent rescue therapy is required. The described case demonstrates that catheter-based rescue

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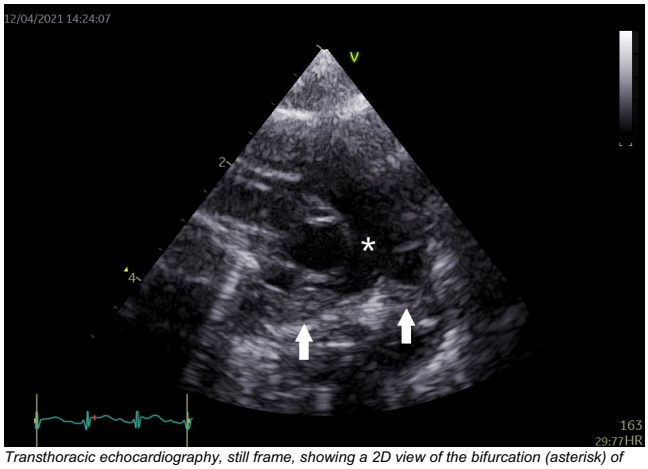
intervention is feasible in small preterm neonates and because of the high risks of other acute management strategies, may be the treatment of choice in such an acutely unstable infant.

Keywords: Pulmonary embolism, catheter intervention, preterm neonate.

Figure 1. Transthoracic echocardiography before intervention



Transthoracic echocardiography, still frame, showing a 2D view of the bifurcation (asterisk) of the pulmonary artery with echodense masses obstructing both pulmonary arteries (white arrows).

PP-314
Ventricular assistance devices in paediatrics as a bridge to recovery

Irene María Romero De La Rosa[1], Pedro Agudo Montore[1], Marta Yagüe Martín[1], Elena Gómez Guzmán[1], María Ángeles Tejero Hernández[1], Jose Joaquín Dominguez Del Castillo[2], Carlos Merino Cejas[2] and Jaime Casares Mediavilla[2]
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Background and Aim: Ventricular assistance devices (VADs) are widely used in paediatrics as a bridge to transplantation. However, their use as a bridge to recovery is an area under development because there are limited data available and because current guidelines does not include exact criteria required for explanting them.
Method: Two case report description and literature review.
Results: Case 1. A 13-year-old boy with no relevant medical history was admitted due to acute cardiogenic shock with severe left ventricular dysfunction (LVD). Due to the refractoriness of the illness with medical treatment for 20 days, it was decided to implant a left pulsatile VAD. A cardiac biopsy compatible with acute myocarditis was received, and the VAD was removed 10 days later because of a significant improvement in function. Currently, he is undergoing follow up in the cardiology clinic with outpatient treatment, and presents moderate LVD with NYHA functional classification I. Case 2. A 20-month-old infant undergoing follow up for dilated cardiomyopathy (mutation in TNNI3), whose sister had a heart transplant for the same mutation, was admitted due to cardiogenic shock after a respiratory infection. The patient showed unsatisfactory evolution, which required continuous assistance, and a left pulsatile VAD was implanted after 14 days. There was significant difficulty in anticoagulation and neurological focality. Moreover, the device was colonised by Serratia marcescens, with no clinical or microbiological response to broad-spectrum antibiotic therapy. It was decided to remove it after 41 days because of the appearance of these serious complications and the function normalization. He continued in follow up with moderate LVD. However, he was admitted in acute cardiogenic shock three months later, after a respiratory infection. A heart transplant was ruled out because of its neurological situation. Finally, he died.
Conclusions: It is vital to highlight there is insufficient data on long-term prognosis and survival, as well as the need for heart transplantation, in these patients. Furthermore, the implantation of large ventricular cannulas could lead to the appearance of fibrosis in the ventriculostomy area, ventricular dysfunction, akinesis or potentially arrhythmogenic areas.
More data are needed regarding the development of a specific protocol for removing them in selected patients.

Keywords: Ventricular assist device, bridge to explantation

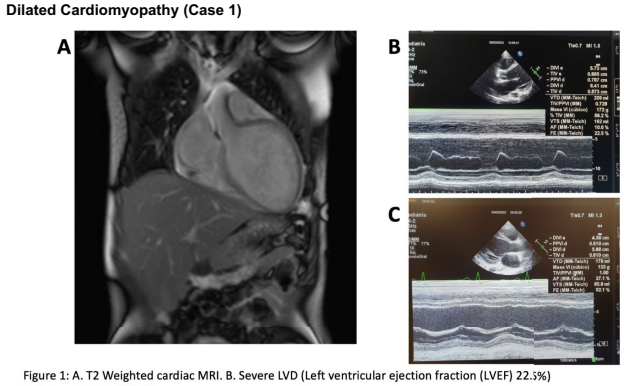

Figure 1: A. T2 Weighted cardiac MRI. B. Severe LVD (Left ventricular ejection fraction (LVEF) 22.3%)
C. Recovery LVEF (52.1%).

MRI cardiac showing dilated, non-hypertrophic left ventricle, with severe dysfunction (LVEF: 14%). Mode M showing the improvement in LVEF.

PP-315
Percutaneous closure of ventricular septal defects with different devices in the years 2002-2008

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Background and Aim: Studies describing long term results of transcatheter closure of ventricular septal defect (VSD) remains scarce. We aimed to evaluate short and long term outcome of this procedure.
Method: In the years 2002-2008 we have attempted to close 29 VSD: 19 patients (pts) with perimembranous (pmVSD) and 10 with muscular (mVSD). In all indication for closure was Qp/Qs ratio > 1,5. In case of pmVSD we applied asymmetrical Amplatzer occluder – asym AO (9 patients - pts with median age of 14 (5-40) years (y)) and Muscular Amplatzer Occluder (MAO) when aortic rim was bigger than 4 mm (10 pts with median age of 10.1 (3,2-40) y). In case of 10 pts with mVSD (median age of 2,25 (0,8-46)y) MAO were used. There were 9 congenital and one posttraumatic mVSD (knife stab). Five children with multiple MVSD previously underwent pulmonary artery banding in infancy.
Results: We were able to close successfully the defect in 24/29 (82,9%) pts. Procedure were abandoned because of severe rhythm disturbances during pmVSD cannulation (2 pts) and impossible cannulation of 1 pt with oblique course of mVSD. In 2 adult

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pts with mVSD embolization of the MAO occurred (probably due to thick IVS). In case of pmVSD treated with asymAO in 2 pts (10 and 13 y) CAVB occurred in the second week post-procedure, which disappeared after steroid therapy in one pt. The second pt required permanent pacemaker implantation (which was active only during first 3 months). In 3 pts with pmVSD treated with MAO moderate tricuspid insufficieny occurred. We did not observe any important complications in long term follow-up (up to 14 y) such as: residual shunt, embolization, hemolysis, infective endocarditis or progressive rhythm disturbances.
Conclusions: Percutaneous closure of pmVSD with asymmetrical Amplatzer device is not recommended because of possibility of rhythm disturbances, whereas with muscular Amplatzer occluder because of possibility of tricuspid insufficiency appearance. Special attention should be paid to closure of mVSD in adult patient with thick IVS (possibility of device embolization). Despite of those finding in long-term follow up of every method was effective.

Keywords: ventricular septal defect, transcatheter closure

PP-316
Percutaneous exclusion of unwanted ventricular-pulmonary connection in patients treated with the fontan strategy
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Background and Aim: Background. Low pressure in the pulmonary vascular bed is crucial for the good functioning of patients treated with a single-ventricle strategy. In some cases, the pulmonary arteries are banded or the pulmonary arteries are recanalized, which results in an increase in pulmonary arterial pressure. and Fontan circulatory failure.
Aim. Evaluation of the results of percutaneous closure of a recanalized or banded pulmonary arteries causing an increase in pulmonary pressure and volume overload of systemic ventricle in patients after Glenn/Fontan procedure
Method: Eight patients (5 after Glenn procedure, 3 after Fontan procedure) aged 2 to 34 (mean- 7,3) years were found to have abnormal ventricular blood flow to the pulmonary bed due to recanalization or incomplete ligation of the pulmonary artery. Cardiac failure was present in all of them, additionally the elevated pulmonary blood pressure was found in 4 pts and "failing Fontan" symptoms in 3 pts. All patients underwent percutaneous inflow closure using different devices: vascular plug, muscular VSD occluder, ASD occluder, ADO II occluder and CP covered stent.
Results: Procedural success (with no residual shunt) was achieved in all patients. Although in one case there was a need for implantation of an additional stent and in another case we had to retrieved MuscVSD and deployed ADOII due to its unsatisfactory position. Follow-up time is 7,4 +/- 3,6 years. We observed improvement and resolution of pleural effusion in the patients with failing Fontan circulation. One patient with cavopulmonary shunt and elevated MPAP six months later was qualified and scheduled for completion of Fontan circulation due to normalisation of MPAP. Two patients had thrombus formation in pulmonary trunk proximally to the device. In one case thrombus formation was detected immediately after the procedure and it resolved with warfarin therapy without complications. Second patient died 6 weeks after the procedure, due to cerebral stroke.
Conclusions: Percutaneous exclusion of ventricle-pulmonary connections is technically possible. This intervention should be considered in patients with cavopulmonary shunt or Fontan circulation with elevated MPAP and volume overload of systematic ventricle. The use of appropriate anticoagulation is crucial for success.

Keywords: percutaneous closure, Fontan procedure

PP-317
Initial experience with implantation of optimus COCR-l stents in children: towards sustainable treatment of vascular stenoses from child- to adulthood
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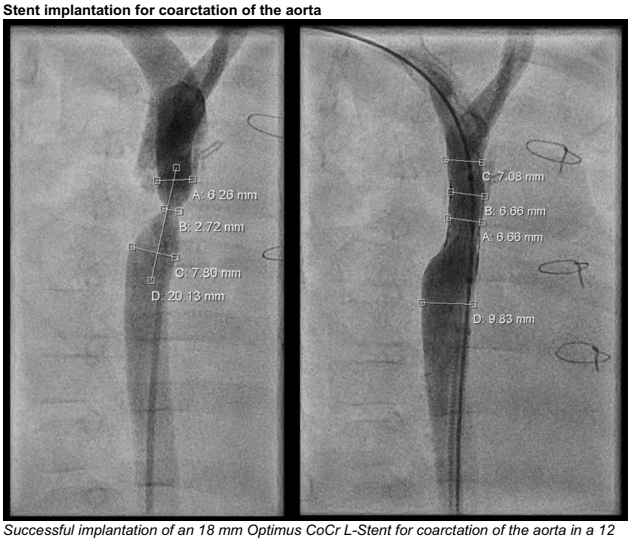
Background and Aim: The search for the optimal stent material to treat vascular stenoses in children remains a challenge. Particularly, small dimensions of the vascular stenoses and/or patients limit the choice of stent material and sizes with acceptable safety. On the contrary, the possibility of appropriate post-dilation needs to be considered to provide a sustainable interventional treatment strategy from child- to adulthood. The Optimus CoCr-L stent is a novel balloon-expandable cobalt-chromium bare-metal stent, which showed promising features in bench test studies in vitro: it could be crimped on an as minimal as 6 mm angioplasty balloon and inserted through 6 Fr sheaths whilst it could be post-dilated up to 23 mm without the occurrence of stent fractures. The stent is intended for use up to 20 mm.
Method: We report our first experience with Optimus CoCr-L stents in the treatment of vascular stenoses in children.
Results: From 6/2023 to 11/2023, 10 Optimus CoCr-L stents were successfully implanted in 10 children (median weight 15.5 kg [9.3-27.4]) due to stenoses of the left pulmonary artery (n=7), superior vena cava/Glenn anastomosis (n=2) or aortic coarctation (n=1). The median diameter of the stenoses was 5.2 mm [2.0-8.3]. For implantation, the stents were crimped onto balloons with diameters of 7 (n=2), 9 (n=5) and 10 mm (n=3), and were inserted through 7 (n=3), 8 (n=4) and 9 Fr (n=3) sheaths. Stent implantation was successful in all patients with elimination of the stenosis (diameter after implantation 8.3 mm [6.7-10.2]). In 3 cases, stents slipped slightly from the balloon during insertion through the hemostatic valve of the sheaths; nevertheless, successful

Stent implantation for coarctation of the aorta



Successful implantation of an 18 mm Optimus CoCr L-Stent for coarctation of the aorta in a 12 months old boy (weight 9.3 kg) crimped on a 7 mm balloon delivered through a 7 Fr sheath

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implantation was still possible. Thanks to improved crimping and insertion techniques, this was no longer a problem with increasing experience. There were no other complications or difficulties with stent implantation.
Conclusions: Our initial findings showed that the Optimus CoCr-L stents could be crimped onto balloons with diameters of 7-10 mm and inserted with 7-9 Fr sheaths, enabling safe and successful use in children with excellent short-term results. However, follow-up studies are required to analyze whether the promising stent capabilities in vitro (post-dilation possible up to 23 mm) can be confirmed in clinical practice.

Keywords: Stent implantation, congenital heart defect, pediatric intervention, coarctation of the aorta, vascular stenosis

PP-318
Utilization of digital subtraction angiography in analyzing pulmonary blood flow with cavo-pulmonary connection - what's old is new again
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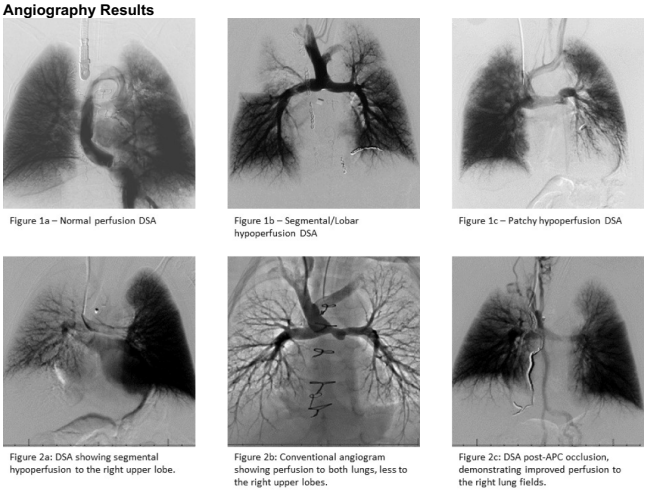
Background and Aim: Digital subtraction angiography (DSA) is an imaging method that allows for high-resolution angiograms with relatively low doses of contrast medium and radiation exposure. There is evidence supporting that quality DSA images can negate the need for further conventional angiographic images, further reducing radiation exposure. Although mainly used by interventional radiologists, its use had been limited to the specific evaluation of certain congenital heart defects (CHDs). However, advances in computing technology have now permitted improved definition of DSA and its use during angiography of CHDs. The aim of this project is to demonstrate the utility of DSA in the evaluation of pulmonary circulation after cavo-pulmonary connection (CPC) and after occlusion of aorto-pulmonary collaterals (APCs).
Method: This is a retrospective review of angiograms performed for evaluation of CPCs including those after the Fontan circulation from 2018 to 2023. The pulmonary circulation based on DSA were categorized into 1). Normal, 2). Segmental/Lobar hypoperfusion, 3). Patchy hypoperfusion (figure 1a-1c). Patients who had undergone occlusion of APCs and had DSA were selected as subjects of this study. The DSA were compared pre- and post-APC occlusion.
Results: Three patients with CPCs and competitive flow were studied during the study period and used as examples for different types of pulmonary circulations demonstrated with DSA (figure 1a-1c). One patient was used as a comparative example of DSA vs conventional angiography: A case study of a 2-year-old boy after CPC with segmental hypoperfusion to the right lung demonstrated by DSA (figure 2a). Compared to the standard angiogram (figure 2b), the extent of competitive flow and hypoperfusion is better demonstrated by DSA. After occlusion of the competitive APC, DSA shows return of flow to the hypoperfused segments (figure 2c).
Conclusions: This case demonstrates how DSA can be used to enhance modern angiography to provide further information regarding the extent of pulmonary perfusion in patients after CPC and can provide additional benefits compared to standard angiography. However, DSA is descriptive and subjective. Future studies with multiple subjects may expand the indications and utility of DSA in the evaluation of CHDs.

Keywords: Angiography, Aorto-Pulmonary Collaterals, Cavo-Pulmonary Connections, Congenital Heart Defects, Digital Subtraction Angiography, Pulmonary Circulation



PP-319
Rapid access patent ductus arteriosus (PDA) closure in extremely low birth weight (ELBW) preterm infants - challenges to clinical pathway development
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Background and Aim: • PDA is the most common cardiovascular condition affecting premature neonates, especially those born with ELBW. Catheter-based PDA occlusion is a revolutionary medical advancement for the very smallest premature infants who require corrective treatment for this condition. It offers clear benefit over the traditional approach via thoracotomy, minimizing secondary complications, and allowing faster recovery, often weaning from inotropic and ventilator supports sooner.
• Historically, PDA closure in ELBW premature infants has represented a 'resource-heavy' intervention, requiring PICU bed/capacity and medical/nursing resources - the facilitation of which can pose a challenge in a tertiary cardiac centre of excellence.
• Preterm infants were also being cared for in PICU versus NICU.
• The demand for a PICU bed combined with lack of PICU capacity (particularly during winter months) can delay treatment for this vulnerable patient cohort.
• Our solution is to create a rapid access service whereby patients are selected by the NICU team and discussed with the interventional cardiologist. Transfer is undertaken by the NICU team directly to the hybrid catheterisation theatre at CHI at Crumlin. The procedure is carried out by the interventional cardiologist and the neonate is transferred back to the NICU within 1-2 hours, eradicating the need for a PICU bed stay. Preterm infants are best managed/cared for in an NICU which can deliver the gold standard of neonatal care.
• The development of this clinical care pathway will contribute to the delivery of neonatal best-practice.
Method: • A pilot programme was developed between CHI at Crumlin and the NICU team in the Rotunda Maternity hospital, to trial this rapid access service.

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• Collaborative meetings were held with members of the MDT and key stakeholders in both CHI at Crumlin and the Rotunda maternity hospital, to troubleshoot and discuss aims and safe execution of the pathway.
Results: • A clinical pathway was developed to ensure safe process for the rapid access PDA device closure service for preterm infants in the NICU at the Rotunda Maternity Hospital.
Conclusions: • Further research into the feasibility and effectiveness of this service will be undertaken with an aim for this service to be introduced to other NICUs in Ireland

Keywords: Clinical Care Pathway, Patent Ductus Arteriosus,

PP-320
Embolization of atrial septal defect occluder device into the abdominal aorta diagnosed as an incidental finding on imaging studies

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Background and Aim: Device embolisation after endovascular closure of atrial septal defects (ASDs) occurs in 1% of cases and is associated with large ASDs, inadequate rims, bidirectional shunt, aneurismatic septum, inadequate device size or placement. Embolized devices usually migrate into the systemic veins, right heart and pulmonary arteries and precise retrieval due to risk of complications, such as perforation, vascular occlusion with ischemia or congestion.
Method: Retrospective single-patient descriptive case report.
Results: We present a male patient congenitally diagnosed with dTGA with intact ventricular septum who during first days of life underwent successful Rashkind procedure and anatomical correction by arterial switch with residual ASD, moderate supravalvular pulmonary stenosis and mild aortic regurgitation. At 15 years of age during systematic follow-up, echocardiography revealed right ventricle overload and catheterization evidenced Qp/Qs 1,8 with normal pulmonary resistance.
Endovascular ASD closure was performed under general anesthesia with TEE guidance evidencing an ASD of 10x10mm, with favorable rims and left to right shunt. A 12-mm ASD septal occluder device (Occlutech Figulla ASD Occluder N) was deployed with fluoroscopic and TEE guidance. After confirming its adequate position with TEE, fluoroscopy control and the "Minnesota technique", the device was deployed.
After an episode of cough during complicated extubation, chest X-ray was performed without evidence of the device in the cardiac chambers, which was later confirmed in ECHO. In the Biplane X-ray the device was visualized in abdominal aorta at the level of renal arteries, without signs of trombosis. Patient was asymptomatic. Successful endovascular retrieval was performed.
Currently at 17 years of age the patient doesn't present complications and is listed for endovascular ASD closure.
Conclusions: Although the exact cause of embolization could not be determined in this case, after excluding common risk factors such as inadequate posteroinferior rim, due to its migration to the systemic arteries, we posit a possible correlation in between Valsalva maneuver during episode of cough, asimetric device and device embolization.

Keywords: ASD, device embolization, endovascular retrieval

Imaging studies

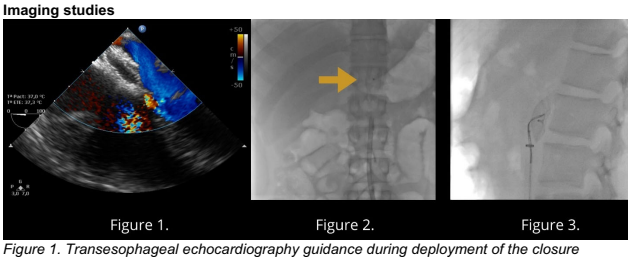

Figure 1. Transesophageal echocardiography guidance during deployment of the closure device. Figure 2. Visualization of the device in abdominal aorta at the level of renal arteries in Biplane X-ray. Figure 3. Endovascular retrieval of the embolized device.

PP-321
Non-contrast closure of patent arterial duct using intracardiac echocardiography

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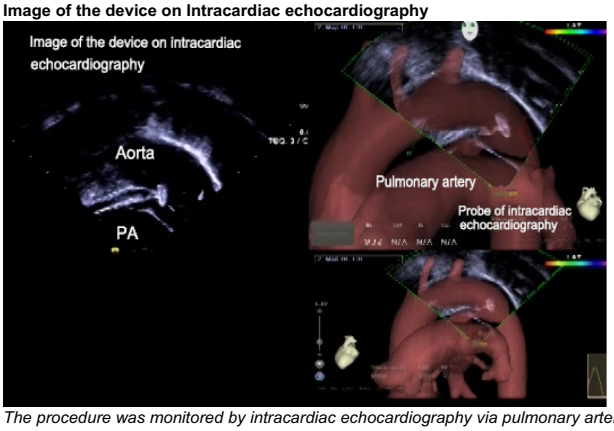
Background and Aim: In transcatheter closure of the patent arterial duct, aortography may not give us a clear image to guide the procedure and precludes in patients with renal dysfunction especially common in elderly patients. This study aimed to examine the effectiveness and safety of intracardiac echocardiography as the guide for this procedure.
Method: Subjects were 9 adult patients, 6 females and 3 males, with a median age of 62 (35-85) years old. Of them, 2 were suffered from renal dysfunction. Krichencko's classification of the arterial duct was type A in 6 and each type B, D, and E in 1 patient, respectively. The minimum diameter of the duct was 3.6 (2.7-11.7) mm and Qp/Qs was 1.9 (1.3-2.4). Pulmonary hypertension was observed in 4 patients.
Results: We have actively used intracardiac echocardiography as the guide for this procedure. Keeping intracardiac echography in the left pulmonary artery, we could have monitored the relationship between the device and the aortic ampulla as well as the pulmonary end, the degree of protrusion of the device into the pulmonary artery, and the residual shunt as in the closure of the atrial septal defect. All patients underwent successful closure

Image of the device on Intracardiac echocardiography



The procedure was monitored by intracardiac echocardiography via pulmonary artery

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using ADO in 8 and ASO in 1 without contrast aortography. No significant complications were observed.
Conclusions: Zero-contrast closure of patent arterial duct can be safely achievable using intracardiac echocardiography, which should be useful for adult patients with renal dysfunction.

Keywords: patent arterial duct, transcatheter closure, intracardiac echocardiography, adult, renal dysfunction

PP-324
Balloon atrioseptostomy and atrial flow regulator implantation for left ventricular failure after rastelli procedure – a case report

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Background and Aim: Complex congenital heart defects (CHD) with borderline left ventricle (LV) remain an important challenge of paediatric cardiology. The aim was to present a case of patient with double outlet right ventricle (DORV), large ventricular septal defect (VSD), transposition of the great arteries (TGA), pulmonary atresia (PA), borderline LV and atrial septal defect (ASD).
Method: The data was collected retrospectively.
Results: A boy with prenatally diagnosed DORV, VSD, TGA, PA, ASD had initially modified right Blalock-Taussig-Thomas shunt performed due to PA. Because of borderline mitral valve (MV) and LV diameters and poor weight gain the decision of final treatment was postponed and patient underwent modified central shunt at the age of 13 months. On reassessment at the age of 20 months echocardiography showed dominance of enlarged right ventricle (RV), MV diameters and LV length within normal values, borderline LV end-diastolic diameter. MRI showed dominant, enlarged, apex forming RV – indexed volume 126ml/m2, LV volume was acceptable - indexed 48ml/m2. The patient was qualified for Rastelli procedure with ASD closure, leaving small foramen ovale. However, he could not be taken off ECMO due to LV failure. On echocardiography LA was enlarged, LV dilated with significantly reduced contractility, RV was compressed by LV. On day 3 mild pulmonary artery banding (on RV–pulmonary arteries homograft) was performed, resulting in deacrease of LA pressure. The patient could be taken off ECMO. During the following days his status remained critical. In order to decompress LV balloon atrioseptostomy was done on day 15. The interatrial communication was enlarged from 1mm to 4mm resulting in decrease of interatrial gradient from 13 to 4mmHg. Following the procedure, the clinical status of the child improved. He could be extubated on day 18 and discharged after another one month of hospitalisation on oral heart failure treatment. Three weeks later the atrial flow regulator (AFR) was successfully implanted due to diminishing shunt. After one month of follow up the patient was in good clinical condition.
Conclusions: Biventricular repair in complex CHD with borderline LV is always favourable, but frequently hard to achieve. Adequate interatrial shunt is crucial in maintaining good LV function and growth.

Keywords: balloon atrioseptostomy, atrial flow regulator, borderline left ventricle

PP-325
Hemolytic anemia in neonate with supravalvular aortic stenosis after arterial switch operation

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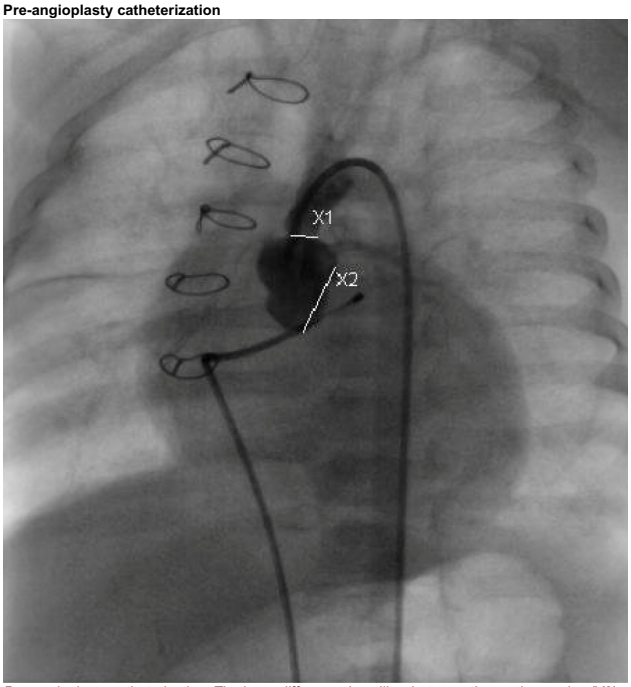
Background and Aim: The arterial switch operation (ASO) is the surgical intervention of choice in dextro-transposition of great vessels (d-TGA). This operation involves major reconstruction of both great arteries including the transfer of the coronary arteries and anterior translocation of the pulmonary artery, known as the Lecompte maneuver. Pulmonary artery stenosis is a well-known complication after the Lecompte maneuver. However, supravalvular aortic stenosis (SVAS) after ASO is uncommon. We present a case report of a patient undergoing ASO who develops severe hemolytic anemia due to SVAS as an early postsurgical complication.
Method: An 4-month-old female patient, prenatally diagnosed with d-TGA without other associated lesions, underwent corrective ASO with the Jatene technique at one week of life. The intervention was incident-free, with good hemodynamic evolution. During follow-up, she developed severe hemolytic anemia requiring weekly transfusions of blood products. An extended study of the anemia was performed, concluding a mechanical cause. Transthoracic echocardiography (TTE) showed severe SVAS (PG 90 mmHg), mild supra-pulmonary stenosis and moderate mitral regurgitation (MR). This was confirmed with CT angiography and catheterization was performed at 4 months of age.
Results: Catheterization revealed a 2.5 mm stenosis at the neoaortic suture level with a post-stenosis aortic caliber of 7.5 mm. Sequential dilatation controlled by TTE was performed with

Pre-angioplasty catheterization



Pre-angioplasty catheterization. The large difference in caliber between the aortic annulus (X2) of 10.75mm and the supravalvular stenosis (X1) of 4mm can be observed.

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Sprinter NC 6/15 mm, Sterling 8/30 mm and Sterling 9/20 mm balloons. After successful SVAS angioplasty, progressive improvement of the supra-aortic gradient was achieved (currently PG 35 mmHg) and improvement of MR, with disappearance of hemolytic anemia.
Conclusions: SVAS is a very rare early post-surgical complication of d-TGA; only several cases have been described, mainly associated with anomalies of the native aortic arch. Furthermore, transcatheter intervention is uncommon as surgical re-intervention has been traditionally done. The destruction of red blood cells as a cause of hemolytic anemia has been described in residual shunts of device closures and in prosthetic valves. We have not found in the literature any study describing mechanical hemolytic anemia associated with SVAS, as in our case.

Keywords: Hemolytic anemia, transposition of great vessels, heart surgery, atrial switch, supravalvular aortic stenosis, postoperative complication.

PP-326
Simulation of healthy patient-specific mitral valves using fluid-structure interaction: validation against in vivo data

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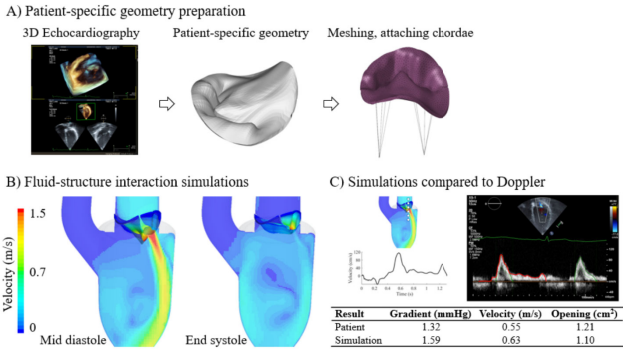
Background and Aim: Atrioventricular heart valve disease is a common primary and secondary cause of heart failure. Atrioventricular valve dysfunction, such as insufficiency or stenosis may be a result of a primary structural valve problem, secondary to ventricular dysfunction, or both. Although techniques for pediatric valve repair exist, it is difficult to predict the outcome of valve interventions and thus tailor the treatment. Patient-specific computational simulations have the potential to provide further insight into the valvular dynamics in different hemodynamic conditions as well as to predict the valve repair outcome. We thus aimed to develop a computational framework to simulate patient-specific mitral valves to investigate differences in trans-valvular velocity, pressure gradient, and valve opening between values obtained from echocardiography (Echo) and simulation.
Method: Four out of ten planned healthy volunteers, aged 28 ± 7.6 years, were recruited. The patient-specific valve geometry was obtained from 3D Echo data and inserted into a generic left heart model (Fig. 1A). The contraction of the heart was mimicked by numerically applying the ventricular volume change as a mass flow to the ventricle, based on 3D Echo measurements. The simulation model uses Fluid-structure interaction (FSI) modeling to simulate the valve motion and blood flow. The valve tissue and chordae tendineae were modeled as linear elastic with Young's modulus of 1 MPa and 22 MPa, respectively. The Carreau model was employed to model the blood viscosity behavior, assuming laminar flow.
Results: Preliminary simulation results (Fig. 1B) show that the valve opening differs by 9 %, and the mean trans-valvular velocity by 16 % compared to Doppler data. The mean pressure gradient, computed using the simplified Bernoulli equation, was subsequentially overestimated by 20 % (Fig. 1C).
Conclusions: The successful implementation of the first patient-specific mitral valve simulation shows reasonable differences between Echo and simulation. Thus, we believe the method has the potential to resolve patient-specific valvular hemodynamics. Further improvements lie in the material modeling of the mitral valve, for improved estimation of the valve opening area and thus the pressure gradient across the valve. Future development entails advancing toward the pediatric population and simulation of valve repair.

Keywords: Simulation, hemodynamics, heart valve repair, fluid-structure interaction



A) the patient-specific mitral valve was obtained from 3D Echo data. B) the velocity through the valve; with peak velocity reached at mid-diastole and a low-velocity field obtained during systole. C) Diastolic Doppler measurements from Echo and computer simulations, from which the mean pressure gradient and the mean velocity are determined.

PP-327
Right disc thrombosis of the new gore cardioform asd occluder

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Background and Aim: The GCA is a well performing device in terms of efficacy despite complex anatomies (aortic rim <5 mm and ASD diameter >17mm) with a good safety profile. To evaluate atrial septal defect (ASD) features impacting on right disc device thrombosis in patients who underwent Gore Cardioform ASD Occluder (GCA) implantation.
Method: A total of 44 consecutive patients undergoing percutaneous ASD with GCA device from January 2020 to September 2022 at our tertiary care Centre were evaluated. The minimum follow-up was 6 months.
Results: The patients were stratified in two groups according to a cut-off value of ASD diameter equal to 20 mm at sizing balloon, derived from ROC analysis (AUC = 0.894; p = 0.024).
Baseline characteristics were comparable between groups in terms of age, sex, weight, height and interatrial septum dimensions. Patients with ASD>20 mm (n=9) had a higher ASD/device dimension ratio, both at echocardiography (p = 0.009) and at

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sizing balloon (p = 0.001), longer fluoroscopic time (p = 0.022), and higher incidence of device thrombosis (0.006). Right disc thrombosis was observed in 3 patients of the ASD>20 mm group, always in the inferior portion of the right disc.
On univariate analysis, ASD diameter at sizing balloon (OR 1.360; p = 0.036) was the only positive predictor of device thrombosis.
Conclusions: Right disc thrombosis of the GCA device may be under-recognized at follow-up, hence deserving clinical attention, especially in those patients with larger ASD diameters

Keywords: GCA, ASD, thrombosis.

PP-328

Percutaneous venus p-valve implantation: preliminary results and six-month follow-up in a single centre

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Background and Aim: Transcatheter pulmonary valve implantation (TPVI) is a recognized alternative to surgery in patients with severe pulmonary regurgitation. The recently developed Venus P-valveTM (Venus Medtech, Shanghai, China) is a self-expanding valve that may be used even in patients with dilated right ventricular outflow tract (RVOT), where other valve systems fail. Since its introduction in the European market, the initial follow-up showed encouraging results, although mid-term follow-up data are still lacking. The aim of our study was to evaluate prolonged outcomes after Venus P-Valve implantation.
Method: Ten adult patients (pts) with a history of surgically-corrected congenital heart disease underwent Venus P-Valve implantation at our Centre from January 2023. All patients had severe pulmonary regurgitation, one of them had an associated moderate valve stenosis. The mean RVOT-PA trunk dimensions, area and perimeter at pre-procedural CT scan are resumed in Table 1.
All procedures were performed under general anesthesia with both left femoral venous and arterial accesses. A complete hemodynamic assessment of the RVOT and PA was performed and the risk of valve-induced extrinsic coronary compression was assessed before deployment. The size of the implanted valve was estimated according to the diameter and morphology of the RVOT and PA trunk obtained via angiography. After valve deployment, repeat angiograms confirmed correct valve function and position.
Results: The Venus P-Valve was implanted successfully in all patients with no procedure-related complications. Transthoracic echocardiography confirmed a properly functioning valve with normal transvalvular gradients, no residual pulmonary regurgitation and paravalvular leaks at discharge. One month follow-up was performed with 24-hours Holter ECG which did not detect significant arrhythmias and echocardiography which confirmed normal valve function. A CT-scan was performed 6 months after the procedure and showed valve leaflet hypoattenuation in seven pts suggesting subclinical leaflet thrombosis. A cardiac MRI (CMR) will be performed at 12 months to evaluate the ventricle function and remodeling.
Conclusions: The self-expanding Venus P-Valve, now available in the CE Market, is a safe alternative to surgical valve replacement in patients with severe pulmonary regurgitation even in the presence of severe RVOT dilatation, however a longer follow-up is needed to assess long term outcomes

Keywords: venus p-valve, pulmonary insufficiency, pulmonary valvular stenosis, right ventricular outflow tract dilatation, Transcatheter pulmonary valve implantation

PP-329

An uncommon natural history of congenital pulmonary vein stenosis disease

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Background and Aim: Congenital pulmonary vein stenosis is a rare condition. There has been a 0.03% incidence in the general population. The timing and severity of symptoms appears to depend largely on the number and severity of PV involved. Magnetic resonance imaging (MRI) has proven to be a useful non-invasive diagnostic method for evaluation of the PV. The natural history of PV stenosis is not entirely clear. Treatment can be expectant, pharmacological, surgical or transcatheter.
Method: A clinical case report describing an uncommon natural history of congenital pulmonary vein stenosis disease.
Results: We present a 17-year-old female, admitted to our hospital due to progressive deterioration of the functional class. Physical examination revealed pleural effusion, pulmonary and tricuspid regurgitant murmur, lower limb edema, hepatomegaly and cyanosis. Echocardiography showed pulmonary artery systolic pressure of 130 mmHg, three dilated pulmonary veins with drainage to a collecting duct connected to the left atrium, a membrane inducing stenosis in this collecting duct, and a persistent ductus arteriosus. MRI revealing stenosis in the left and superior right pulmonary veins. Later on, the angiography confirmed the presence of PV stenosis, a transseptal puncture was performed, the left PV was observed with stenosis at the left atrium, angiography was performed in superior, middle and inferior PV, observing confluence in a single vein and with stenosis at the left atrium. Angioplasty with stent was delivered in the common ostium of the left PV and right PV with adequate opening, observing adequate expansion. Post-treatment, she was discharged in well health conditions, with a management plan involving spironolactone, acetylsalicylic acid, and sildenafil. One year later, cardiac catheterization was performed again, finding a pulmonary pressure at 60% of the systemic pressure and no obstruction at the level of the PV.
Conclusions: Congenital pulmonary vein stenosis is a rare entity which in pediatric age manifests mainly in the early years, however in this case, the natural history of the disease was not as expected so it is important to know that despite the severity of symptoms with which it could manifest. We must know the variants of

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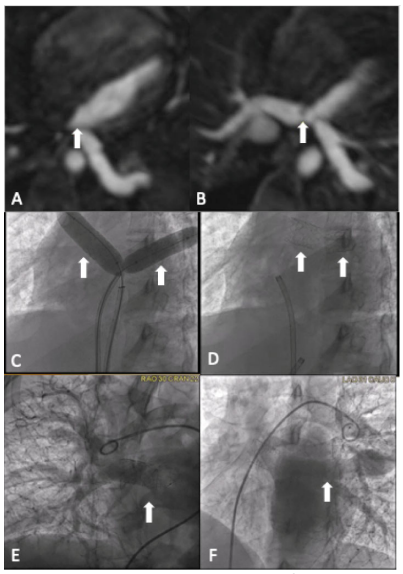
presentation and high clinical suspicion for the appropriate diagnostic and therapeutic approach.

Keywords: congenital estenosis pulmonary veins, interventional cardiology, cardiac imaging.

Figure 1



A. MR pulmonary angiography showing left pulmonary vein stenosis (white arrow). B. MR pulmonary angiography showing right pulmonary vein stenosis (white arrow). C. Angiography showing angioplasty with stent in the common ostium of the left and right pulmonary veins. (white arrow). D. Angiography showing stents without displacement in the left and right pulmonary veins (white arrow). E. Angiography one year after angioplasty showing adequate blood flow through the right pulmonary veins in levo-phase (white arrow). F. Angiography one year after angioplasty showing adequate blood flow through the left pulmonary veins in levo-phase (white arrow).

PP-330

From 4,2% to 1.5% - the role of 3D modeling in reducing complications in transcatheter asd closure

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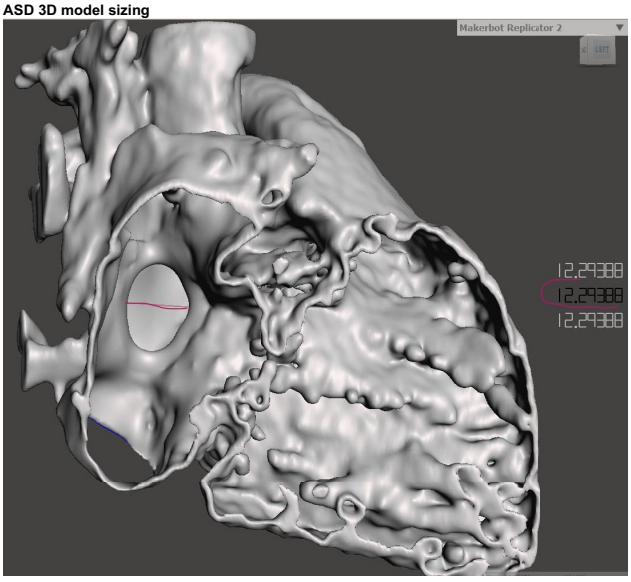
Background and Aim: Percutaneous closure of ASD is known as an alternative treatment to surgery, which provides many benefits (decreased surgical morbidity, no scarring, and shortened hospital stay). However, it is associated with infrequent, but life-threatening complications such as device embolization.The incidence of ASD device embolization varies from 0.55% to 1.1%, according to various sources. Analyzing our results of transcatheter closure of ASD, over a period of 14 years, we found an unacceptably high rate of device embolization of 4.2%. From 164 percutaneously closed ASDs, we have 7 device embolizations - 3 of them were removed transcatheterically, and 4 surgically. We set ourselves the goal of reducing the rate of device embolizations, considering that the main reason for these complications is poor assessment of ASD morphology and wrong size device selection.
Method: Starting in 2019, we reinstated balloon sizing and started a 3D modeling program for all patients referred for transcatheter ASD closure.
Results: Based on the prepared 3D computer models, we evaluated the morphology and size of the ASD. We compared the defect sizes from TTE, TEE, the 3D model and balloon sizing and found that there were significant discrepancies between the defect sizes from TTE, TEE and balloon sizing. Δ (balloon sizing - TTE) p = 0,033 and Δ (balloon sizing - TEE) p = 0,054 We found no statistically significant difference between defect size from 3D models and balloon sizing. Δ (balloon sizing - 3D model) p = 0,33 For the period from 2019 to 2023, we percutaneously closed 67 ASDs and had only one device embolization (1.49%).
Conclusions: We believe that computer 3D modeling can significantly help to estimate the size and morphology of ASD and reduce the number of device embolizations.

Keywords: 3D modeling, transcatheter ASD closure, device embolizations



PP-331

Fast technique: fast atrial sheath traction technique for device closure of atrial septal defects

Mahmoud Alsoufi
aljalila children specialty hospital

Background and Aim: Transcatheter closure of atrial septal defects (ASDs) is well-established. However, this procedure can be challenging, requiring multiple attempts and advanced implantation maneuvers.
Method: From July 2019 to July 2022, patients to whom the fast atrial sheath traction (FAST) technique was applied for ASD device closure were prospectively followed up. The device was rapidly unsheathed in the middle of the left atrium (LA) to let it clamp the ASD from both sides simultaneously. This novel technique was directly applied in patients with absent aortic rims and/or ASD size-to-body weight ratio higher than 0.9 or after failed attempts of standard implantation.
Results: Seventeen patients (64.7% males) were involved with a median age of 9.8 years [interquartile range (IQR), 7.6–15.1] and a median weight of 34 kg (IQR, 22–44). The median ASD size on ultrasound was 19 mm (IQR, 16–22). Five (29.4%) patients had absent aortic rims, and three (17.6%) patients had an ASD size-to-body weight ratio higher than 0.9. The median device size was 22 mm (IQR, 17–24). The median difference between device size and ASD two-dimensional static diameter was 3 mm (IQR, 1–3). All interventions were straightforward without any complications

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using three different occluder devices. One device was removed before release and upsized to the next size. The median fluoroscopy time was 4.1 min (IQR, 3.6–4.6). All patients were discharged the next postoperative day. On a median follow-up of 13 months (IQR, 8–13), no complications were detected. All patients achieved full clinical recovery with complete shunt closure.
Conclusions: We present a new implantation technique to efficiently close simple and complex ASDs. The FAST technique can be of benefit in overcoming left disc malalignment to the septum in defects with absent aortic rims and in avoiding complex implantation maneuvers and the risks of injuring the pulmonary veins

Keywords: Atrial septal defect, device closure

PP-332
Percutaneous closure of ventricular septal defects with different devices in the years 2009-2016
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Medical University of Silesia

Background and Aim: Studies describing long term results of transcatheter closure of ventricular septal defect (VSD) remains scarce. We aimed to evaluate short and long term outcome of this procedure.
Method: In the years 2009-2016 we have attempted to close 22 VSD: 13 patients (pts) with muscular (mVSD), 7 pts with residual post surgery (RVSD) and 2 pts with perimembranous VSD(pmVSD). In all pts indication for closure was Qp/Qs ratio > 1,5. In case of mVSD (pts with median age of 2,25 (0,7 - 10) years (y) we applied Muscular Amplatzer Occluder (MAO). In 5 cases with mVSD hybrid procedure was applied – transcatheter mVSD closure and surgical debanding of pulmonary artery in 4 children and in one infant with 4,9 kg body weight as a single procedure. In case of RVSD (median age of 3,0 (1,5 - 31)y) and 2 pmVSD (2 and 4 years old respectively) different duct occluders (Amplatzer or Chinese Amplatzer-like Amplatzer Duct Occluder type I - ADOI) or ADO type II (ADOII), or ADOII Additional Sizes(ADOIIAS) were used.
Results: We were able to close successfully the defect in 20/22 (91%) pts. Procedure was abandoned because of severe rhythm disturbances during mVSD cannulation in 1 pt. In another patient with inflow type mVSD 3 days after implantation of MAO massive tricuspid valve (TV) regurgitation appeared, caused by TV chorde tendinae cutting (device was removed, VSD surgically closed and TV was successfully reconstructed). We did not observe any important complications in long term follow-up (up to 6y) such as: residual shunt, embolization, hemolysis, infective endocarditis or progressive rhythm disturbances.
Conclusions: Percutaneous closure of inflow MVSD with muscular VSD occluder is not recommended because of possibility of inpredictable tricuspid valve damage. Percutaneous closure of not big pmVSD and postsurgical residual VSD with ADOIIAS is especially suitable because of its soft construction. Untill now there is a lack of ideal device for pmVSD closure.

Keywords: ventricular septal defect, transcatheter closure

PP-333
Leaflet thrombosis after percutaneous pulmonary valve implantation: a case series
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Background and Aim: Hypoattenuating leaflet thickeness (HALT) is a well described phenomenon in biological aortic valves with yet unknown consequences but, to our knowledge, no cases of HALT have been reported after percutaneously implanted pulmonary valves.
Method: Here we are presenting three cases of HALT after Venus P Valve implantation.
Results: Three patients with Tetralogy of fallot and hemodinamically pulmonary valve insufficiency were deemed suitable for pulmonary valve implantation with a Venus P valve. Two patients were implanted with a Venus P Valve 34/25 mm and one patient with a 32/25 mm. Post operative course was uneventful. Two patients were discharged on dual antiplatelet therapy while one patient on anticoagulative therapy because of a change in our center protocol. All the Valves were well functioning at echocardiographic examination at follow up but in all there was a newly appearance of a mild central valvar insufficiency. A thickening and hypo-mobility of the pulmonary valve cusps, suspected for HALT was detected in two patients at CT Scan performed at three months according to our center protocol, while in the last patient CT Scan was anticipated at one month because of a suspicion related to the newly appearance of a mild pulmonary valve insufficiency and it showed thickening and hypo mobility of one of the cusps. All patients are yet on anticoagulative therapy and DAPT was stopped.
Conclusions: We described three cases of HALT after Pecutaneous pulmonary Valve implantation with Venus P valve. In our experience among 10 patients implanted with a venus P valve in the period from July 2022 to September 2023 and undergoing a control CT scan, the incidence of HALT was 33%. All the patients were on DAPT and in all cases echocardiography could not detect any valve malfunctioning or significant alteration except for a newly mild valvar regurgitation. This, in our opinion, could be a red flag for HALT. HALT detection in percutaneously implanted pulmonary valve has never been described but could be an important trigger for valve malfunctioning and endocarditis. Multicenter studies with longer follow up in patients undergoing PPVI are needed in order to clarify causes, consequences and treatment of this complication.

Keywords: HALT, autoexpandable valve

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PP-334

Immediate and long-term results of balloon aortic valvuloplasty for severe aortic valve stenosis in newborns. a single center experience

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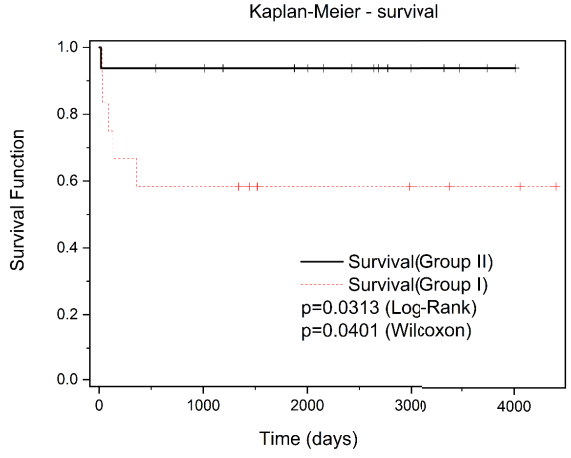
Background and Aim: Neonates with severe isolated aortic valve stenosis (AVS) requiring urgent treatment constitute unique group of patients. In most centers, the treatment of choice is balloon aortic valvuloplasty (BAV). The aim of the study was to assess the immediate and long-term outcome of BAV in newborns including preterm infants with severe/critical AVS
Method: We analysed retrospectively 28 neonates with severe isolated AVS treated with BAV procedure at mean age of 12.1 days and body weight 3213 g. The follow-up was more than 5 years (mean 6.4). The patients were divided into 2 groups: with left ventricle ejection fraction (LVEF) ≤40 % (Gr-I, n=12) and >40% (Gr-II, n=16). There were five premature newborns in Gr-I and two in Gr-II.
Results: The pressure gradient through the aortic valve decreased significantly following BAV in both groups, mean LVEF increased significantly only in Gr-I. Twelve children (42,8%) required re-interventions: a repeat BAV (n=4), surgical valvuloplasty (n=8) and AV replacement (n=1). Re-interventions were more common in Gr-I (50% vs 37.5%, p=0.5083). In long-term follow up six patients died, 5 in Gr-I and 1 in Gr-II, including 3 of 7 premature babies (all from Gr-I). Kaplan Meier survival curve showed significantly higher mortality in Gr-I (p=0.03).
Conclusions: The BAV procedure as primary palliative therapy in neonates with severe AVS shows good initial results, but further follow-up indicates a common need for re-interventions. Overall left ventricular systolic dysfunction and prematurity are risk factors for higher mortality.

Keywords: aortic valve stenosis, balloon aortic valvuloplasty, clinical outcomes, newborns, premature babies

Figure 4.



Kaplan-Meier plots presenting survival in the studied groups were presented in Figure 4

Neurodevelopment and psychosocial care

PP-335

Health-related quality of life in children and adolescents with marfan syndrome or related disorders: a controlled cross-sectional study

Thomas Edouard[1], Fernanda Bajanca[1], Maud Langeois[1], Eric Garrigue[1], Bertrand Chesneau[1], Aitor Guitarte[1], Helena Huguet[2], Philippe Khau Van Kien[3], Yves Dulac[1] and Pascal Amedro[4]
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Background and Aim: This cross-sectional controlled study aims to assess health-related quality of life (HRQoL) of children and adolescents with a molecular diagnosis of Marfan syndrome (MFS) or related disorders and to evaluate the factors associated with HRQoL in this population.
Method: Sixty-three children with MFS and 124 age- and sex-matched healthy children were recruited (NCT03236571). HRQoL was assessed using the Pediatric Quality of Life Inventory (PedsQL™) generic questionnaire.
The correlation between HRQoL scores and the different continuous parameters (age, body mass index, disease severity, systemic score, aortic sinus diameter, and aerobic physical capacity) was evaluated using Pearson's or Spearman's coefficient. A multiple linear regression analysis was performed on the two health summary self-reported PedsQL™ scores (physical and psychosocial) to identify the factors associated with HRQoL in the MFS group.
Results: Except for emotional functioning, all other domains of HRQoL (psychosocial and physical health, social and school functions) were significantly lower in children with MFS compared to matched healthy children. In the MFS group, the physical health summary score was significantly lower in female than in male patients (self-report: absolute difference [95%CI] = -8.7 [-17.0; -0.47], P = 0.04; proxy-report: absolute difference [95%CI] = -8.6 [-17.3; 0.02], P = 0.05) and also negatively correlated with the systemic score (self-report: R = -0.24, P = 0.06; proxy-report: R = -0.29, P = 0.03) and with the height Z-score (proxy-report: R = -0.29, P = 0.03). There was no significant difference in the physical health summary scores between the different genetic subgroups. In the subgroup of 27 patients who performed a cardiopulmonary exercise test, self- and proxy-reported physical health summary scores were highly correlated with their aerobic physical capacity assessed by peak oxygen consumption (VO2max) and ventilatory anaerobic threshold (VAT). In the multivariate analysis, the most important independent predictors of decreased physical health were increased height, decreased body mass index, decreased VAT and use of prophylactic therapy.
Conclusions: This study reports an impaired HRQoL in children and adolescents with MFS or related conditions, in comparison with matched healthy children. Educational and rehabilitation programs must be developed and evaluated to improve exercise capacity and HRQoL in these patients.

Keywords: Marfan Syndrome, health-related quality of life, aerobic physical fitness, peak oxygen consumption, physical well-being, psychological well-being

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Table 2. Predictors of HRQoL self-reported physical functioning in children with Marfan syndrome.

Variables			Univariate analysis		Multivariable analysis (n=21)	
		N	cβ [95%CI]	P-value	aβ [95%CI]	P-value
Age (years)		62	-0.86 [-1.95; 0.25]	0.1258	-	-
Gender	Female	22	8.51 [-17.80; 0.78]	0.0719	-	-
	Male	40	-			
Height (z-score)		62	-0.57 [-3.74; 2.59]	**0.7185**	-4.86 [-8.71; -1.02]	**0.0164**
Body mass index (z-score)		62	0.37 [-2.33; 3.07]	**0.7855**	3.37 [0.86; 5.89]	**0.0117**
Aortic sinuses diameter (z-score)		60	-1.14 [-4.69; 2.41]	0.5213	-	-
Medical prophylactic therapy	No	57	-20.00 [-35.96; -4.04]	**0.0149**	-29.19 [-46.77; -11.61]	**0.0001**
	Yes	5	-		-	
Ectopia lentis	Yes without surgery	37	1.87 [-10.33; 14.08]	0.2998	-	-
	Yes with surgery	14	-7.87 [-19.02; 3.29]			
	No	11	-			
Systemic score		62	-1.30 [-2.86; 0.26]	0.1008	-	-
VO_{2max} (mL/Kg/min)		27	1.01 [0.33; 1.69]	0.0053	-	-
VAT (mL/Kg/min)		27	2.20 [0.82; 3.57]	**0.0029**	2.96 [1.71; 4.21]	**0.0028**
Inheritance	De novo	35	-3.02 [-13.12; 7.09]	0.5522	-	-
	Familial	22	-		-	-
Place among siblings		56	-3.21 [-7.52; 1.09]	0.1406	-	-
Civil status	Divorce / death	41	-10.96 [-21.01; -0.94]	0.0327	-	-
	Together	16	-			
Living area	Urban zone	26	-7.54 [-16.59; 1.51]	0.1006	-	-
	Rural zone	33	-			

cβ: crude β; aβ: adjusted β; CI: confidence interval. Variables associated with PedsQL self-reported physical functioning in both univariate and multivariable analyses are marked in bold.

PP-336

Assessment of the corpus callosum by ultrasound in patients with congenital heart disease: description of the factors influencing its development

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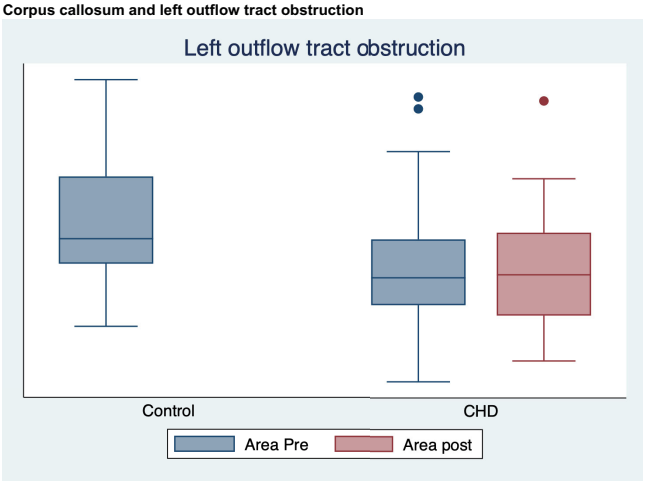
Background and Aim: Congenital heart disease (CHD) is the most common congenital malformation. A high incidence of CNS lesions with consequent alteration of their neurodevelopment has been described in these patients. White matter alteration is the most common lesion. The corpus callosum is one of the most important biomarkers to assess white matter. Oxidative stress has been related to the development of white matter damage.
Our aim is to describe changes in the biometry of the corpus callosum, to identify risk factors and o try to establish how the type of congenital heart disease and its surgery influence in these changes.
Method: Pre- and postoperative cerebral ultrasounds were analyzed in 70 patients who undergo cardiac surgery within the first month of life. The obtained results were compared with those of a control group of healthy patients. Measurements of the total length and area of the corpus callosum were performed off-line using specific software designed for this purpose (Matlab 2009b, The MathWorks Inc., Natick, MA).
Corpus callosum measurements were analysed according to on their CHD. In addition, a correlation between corpus callosum biometrics and different variables was performed.
Results: A significant decrease in the area of the corpus callosum was observed in both before and after surgery studies when comparing patients with CHD to healthy patients. We also observed a statistically significant decrease in total corpus callosum area in patients with left outflow tract obstruction and in patients with cyanotic heart disease. No relationship was observed between oxidative stress biomarkers levels and corpus callosum biometrics.
Conclusions: Patients with CHD have a smaller corpus callosum than healthy patients. Hypoperfusion and cyanosis are two of the main factors affecting corpus callosum development. Early detection of these changes can help to identify high-risk patients, assuring individualized intervention and follow-up programs.

Keywords: Corpus callosum, ultrasound, congenital heart disease, oxidative stress

Corpus callosum and left outflow tract obstruction



PP-337

Developmental and psychological concerns in children with complex congenital heart disease: the need for a screening program

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Background and Aim: Complex congenital heart disease (CHD) is associated with adverse neurodevelopmental and psychological outcomes. Currently, there is limited Australian data regarding the frequency of these outcomes and presence of active developmental or psychological screening and intervention. In South Australia, there is no established screening program for this cohort. This study aims to determine the local prevalence of developmental and psychological concerns in preschool and early primary school aged children with complex CHD, and any current involvement with developmental and psychological programs.
Method: Families of children with complex CHD aged between 3 years and 7 years 11 months, identified through the local departmental database, undertook a series of validated screening questionnaires for development (Parents' Evaluation of Developmental Status
[PEDS], Parents' Evaluation of Developmental Status - Developmental Milestones
[PEDS:DM]), and mental health and behaviour (Strengths and Difficulties Questionnaire
[SDQ]). These were evaluated by a single trained medical practitioner. Demographic data, cardiac history, comorbidities, and current healthcare engagement was determined through standardised patient information questionnaires and medical records. Rates of developmental and psychological concern were determined, with comparison between participants who received initial cardiac surgery before and after one month of age.
Results: Thirty-six percent (55/151) of people identified consented to participate in the study. The most common cardiac conditions were Transposition of the Great Arteries (n=13), Tetralogy of Fallot (n=11) and post-cardiac transplantation (n=5). The majority of participants had at least one predictive developmental domain of concern in the PEDS (57%) and PEDS:DM (70%) screening tools, which exceeds Australian population rates (22%). Psychological concern was identified in 28% of participants, which is high

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relative to Australian population data (10%). Almost half (48%) of the participants identified to have at least one domain of developmental concern had no current general paediatric or allied health team engagement. Similarly, of participants with psychological concerns, 45% had no current psychology or general paediatrics engagement.
Conclusions: Developmental and psychological concerns detected on screening tools in children with complex CHD significantly exceed Australian general population prevalence rates. Without an established screening program, a large proportion of participants did not have active follow-up, indicating the importance of commencing screening programs in this cohort.

Keywords: Developmental, psychological, complex congenital heart disease

PP-338

What are the neurodevelopmental outcomes in patients who receive cardiopulmonary bypass due to congenital heart disease?

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Background and Aim: Neurodevelopmental outcome in patients with Congenital Heart Disease (CHD) has been a growing field of interest, and despite several large studies and publications on the topic, it is difficult to monitor and address this early.
The aim of this study is to assess neurodevelopmental outcomes at 5 years of age in patients who receive cardiopulmonary bypass, within the first year of life due to CHD.
Method: A retrospective study, looking at patients who received cardiopulmonary bypass within the first year of life, due to CHD born from 01/01/2014 to 31/12/2014 under the Leeds Teaching Hospital Trust (LTHT). A selection of parameters were monitored against neurodevelopmental outcomes at age 4-6 to ascertain whether there was an association.
Results: There is an association between CHD and lower neurodevelopmental outcomes, the most significant findings are described below.
Patients with a single ventricle are more likely to have a motor deficit (Chi-squared sig, p-value 0.003) and have a documented neurodevelopmental deficit (Chi-squared sig, p-value 0.006).
Total bypass time and cross-clamp time correlate to hearing impairment (respectively t-test -2.47, p-value 0.015 and t-test -2.0, p-value 0.045) and total circulatory arrest to motor deficit (t-test -2.32, p-value 0.02).
During the post operative period there was a correlation between duration of post operative ventilation and motor (t-test -2.8, p-value 0.006) and hearing impairment (t-test -2.2, p-value <0.028); repeat surgical intervention with motor (p-value <0.001), hearing (Chi-squared sig, p-value 0.031), behavioural (Chi-squared sig, p-value 0.016), vision (Chi-squared sig, p-value 0.031) and any neurodevelopmental deficit (Chi-squared sig, p-value 0.001) and acute neurological event and vision (Chi-squared sig, p-value 0.002).
Conclusions: Despite improvements in surgical techniques, children with CHD who had cardiac bypass surgery between 2014-2015 in this study have an association with poorer neurodevelopmental outcomes, with comparable results to other cardiac centres internationally.
This study hopes to:

1. Increase awareness/information on the link between neurodevelopment and CHD.
2. To provide the patients and families with better prognostication in the initial stages.
3. To highlight areas for closer monitoring and early referral for further assessment/care in the community paediatric set up.

Keywords: Congenital Heart Disease, neurodevelopmental outcomes, cardiopulmonary bypass

PP-339

Altered cerebral growth trajectories from third trimester to neonatal age in patients with congenital heart disease

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Background and Aim: Newborns with congenital heart disease (CHD) often face challenges beyond primary cardiac outcome. During long-term follow up they are at risk for impaired neurodevelopment, which confirms the need for a better understanding of modifiable risk factors influencing their brain development and their clinical outcome. This includes a larger knowledge of their brain development starting during fetal life as hemodynamic alterations due to the CHD are present before birth influencing fetal cerebral perfusion.
Method: In this longitudinal, multicenter study we employed serial (at least two times) fetal cerebral magnetic resonance imaging (MRI) during last trimester and after birth in the perioperative setting in patients undergoing neonatal cardiac surgery for severe type of CHD to analyze brain development. Patients (n=30) were scanned multiple times from third trimester of pregnancy until two months of age. Patients were compared to healthy controls group (n=9) that underwent a fetal and a neonatal scan. Here, we present data from a cohort born between 6/2020 and 12/2022. Volumetric measurements for the total brain volume as well as sub-tissue specific measurements for cortical grey matter, white matter, deep grey matter, brain stem and cerebellum were generated based on the MRI and a gyrification index was calculated.
Results: Using a mixed effects model, that accounted for birth weight, age at scan and random intercept for each participant, our analysis revealed reduced total brain volume in CHD children with a significant interaction effect between group and age at scan (group (CHD vs control) beta estimate: -49.76, p-value <0.001, interaction term group*age at MRI: beta estimate: 6.65, p-value<0.001).

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Conclusions: Our results support the theory, that brain growth trajectories might be reduced in CHD compared to controls and that volumetric differences between CHD and controls become more pronounces at later stages.

Keywords: brain growth, neurodevelopment, cerebral MRI

PP-340

Childhood neurocognitive outcome in patients with fontan circulation

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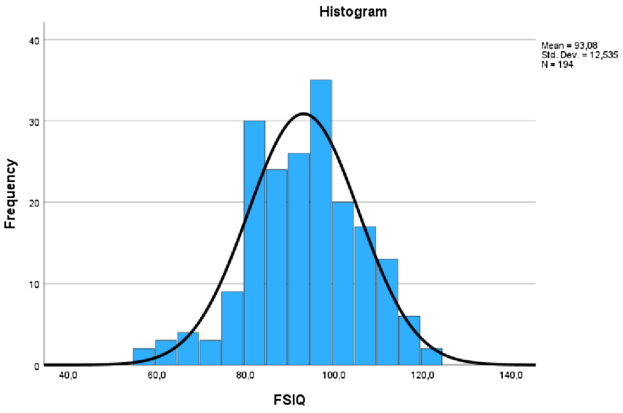
Background and Aim: Within the growing population of Fontan patients, long-term cognitive outcome and potential modifiers are of special interest. Better understanding and characterization of the multifactorial deficits and abilities can have a major impact on our patients in the future. This single-centre study aimed to assess neurocognitive outcome in a large contemporary Fontan cohort.

Method: Single ventricle patients born between 2004 and 2019 who received total cavopulmonary connection (TCPC) and preceding surgical steps in our centre were identified. Patients who underwent neurocognitive testing with age-adapted standardised versions of the Wechsler Preschool and Primary Scale of Intelligence or the Wechsler Intelligence Scale for Children in the current German edition were included. A subgroup of preschoolers was also assessed with three subtests from the German "Kognitiver Entwicklungstest für das Kindergartenalter" (KET-Kid: psychomotor development, visuoconstruction and spatial conception).

Results: Overall, 305 patients were identified. Of those, 203 underwent neurodevelopmental assessment (hypoplastic left heart syndrome: 55.7%; other univentricular heart defects: 44.3%). Median age at TCPC was 2.8 years (range 1.5-9.6). Neurocognitive examination was performed at 5 years (3.1-13.1). A full-scale IQ (FSIQ) could be realized in 194 cases and was in the lower-normal range (mean FSIQ: 93 ±12.5). Severe cognitive impairment (IQ<70) was found in 4.6% of the cases, 21.7% had moderate impairment (IQ: <85-70) and 69.6% showed normal intellectual potential (IQ: 85-115) and 4.1% scored above normal range (IQ>115). Among the subgroup of pre-school children (n=160), 34.4% showed a developmental delay in visual construction. FSIQ was not different between patients with hypoplastic left heart syndrome or other univentricular heart defects (93.1 ±13.2 vs. 93.0 ±11.7, p=0.974). No difference in FSIQ was observed between 137 patients requiring neonatal surgery utilizing cardiopulmonary bypass and 57 cases managed without neonatal cardiopulmonary bypass surgery (93.1 ±13.1 vs. 92.9 ±11.1, p=0.913).

Conclusions: Overall, neurocognitive outcome in this contemporary Fontan cohort was in normal range. However, with a higher proportion of patients with moderate impairment compared to the normal population. Hypoplastic left heart syndrome or the need for neonatal cardiopulmonary bypass surgery were not associated with lower FSIQ scores. All Fontan patients should have neurodevelopmental monitoring throughout childhood to identify the need of support.

Keywords: neurodevelopmental outcome, Fontan circulation, congenital heart disease, intelligence tests, cognitive function



Distribution of FSIQ scores for the entire cohort

PP-341

Educational attainment of children with congenital heart disease in the united kingdom

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Background and Aim: Educational attainment in children with congenital heart disease (CHD) within the UK has not been reported, despite the possibility of school absences and disease-specific factors creating educational barriers.

Method: Children were prospectively recruited to the Born in Bradford birth cohort between March 2007 and December 2010. Diagnoses of CHD were identified through linkage to the congenital anomaly register and independently verified by clinicians. Multivariable regression accounted for relevant confounders. Our primary outcome was the odds of 'below expected' attainment in Maths, Reading and Writing at ages 4-11 years.

Results: Educational records of 139 children with non-genetic CHD were compared to 11,188 age-matched children with no major congenital anomaly. Children with CHD had significantly higher odds of 'below expected' attainment in Maths at age 4–5 years (Odds Ratio 1.64, 95% CI 1.07–2.52), age 6–7 (OR 2.03, 95% CI 1.32–3.12), and age 10–11 (OR 2.28, 95% CI 1.01–5.14). Odds worsened with age, with similar results for Reading and Writing. The odds of receiving special educational needs support reduced with age for children with CHD relative to controls (age 4–5: OR 4.84 (2.06–11.40); age 6–7: OR 3.65 (2.41–5.53); age 10–11: OR 2.73 (1.84–4.06)). Attainment was similar for children with and without exposure to cardio-pulmonary bypass.

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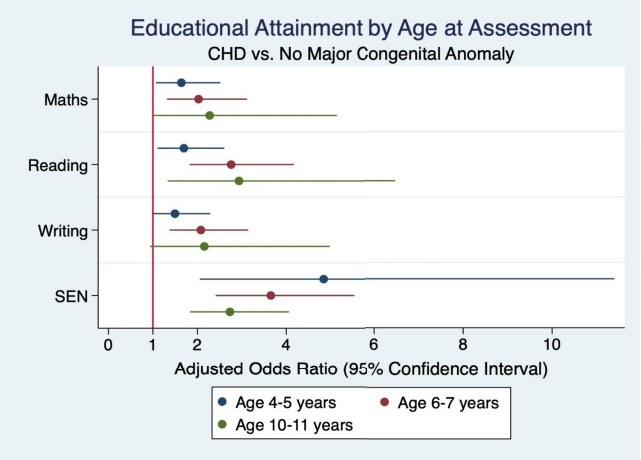
Lower attainment was strongly associated with the number of preschool hospital admissions.
Conclusions: Children with CHD have lower educational attainment compared to their peers. Deficits are evident from school entry and increase throughout primary school.

Educational attainment for children with CHD



Educational attainment at each assessment age for children with CHD vs. no major congenital anomaly. Odds ratios for 'below expected' achievement adjusted for sex, month and year of birth, consanguinity, child ethnicity, Index of Multiple Deprivation score, receipt of benefits and maternal smoking or alcohol use in pregnancy.

Keywords: congenital heart disease, education, school.

PP-343

Quality of life in children with severe congenital heart disease: its association with surgical factors. a systematic review

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Background and Aim: Children with severe congenital heart disease (CHD) often undergo cardiac surgery involving cardiopulmonary bypass (CPB), early in life. Such procedures are suspected to affect neurocognitive development and quality of life (QoL), but the specific factors that underly this association remain unclear. This systematic review aims to delineate the specific perioperative factors influencing long-term QoL in children with severe CHD undergoing early cardiac surgery.
Method: A comprehensive search was conducted in Medline, Embase, and Web of Science Core Collection databases, spanning from 1975 to June 2023, to identify studies that explored the association of various perioperative variables — such as the number of surgeries, CPB duration, cross-clamp time, and length of hospital/intensive care unit (ICU) stay — with QoL in children with severe CHD who underwent early cardiac surgery.
Results: Sixteen studies were included, including eight cross-sectional and six cohort studies, with sample size ranging from 40 to 572. QoL was assessed in subjects aged 1 to 20 years. Predominant perioperative variables studied were CPB time (9 studies), hospital/ICU stay duration (8), and number of surgeries (7). The Pediatric Quality of Life Inventory (4) and Child Health Questionnaire (4) were primarily used for QoL assessment. A consistent finding was a significant negative association between the number of surgeries and QoL scores, particularly in the physical functioning domain. Similarly, prolonged CPB time and hospital/ICU stay also correlated with lower QoL scores. However, heterogeneity in QoL assessment tools and outcome reporting precluded a conclusive aggregated effect size estimation.
Conclusions: This systematic review highlights a clear inverse association between the number of surgeries, CPB duration, and length of hospital/ICU stay with QoL in children with severe CHD. The observed heterogeneity in outcome assessment and reporting methodologies underscores the need for standardization in future research and healthcare pathways. In the context of shared decision-making, QoL assessment can serve as a crucial tool in consultations with medical specialists, functioning as an initial screening for children and parents who might need additional diagnostics or support for developmental delay. Standardization in research methodologies is crucial to accurately determine the perioperative factors affecting QoL.

Keywords: Congenital Heart Disease, Pediatric Cardiac Surgery, Quality of Life, Perioperative Factors, Neurocognitive Development

PP-345

Glial fibrillary acidic protein: a neuromarker of psychomotor developmental impairment in patients with congenital heart defects after cardiovascular

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Background and Aim: Psychomotor developmental impairment is the most frequent association in patients with congenital heart defects (CHD). Early diagnosis and treatment improve long-term quality of life. The aim of this study was to evaluate glial fibrillary acidic protein's (GFAP) role in predicting impairment in psychomotor development of patients with CHD which undergo cardiac surgery.
Method: A prospective, observational study was conducted which included 40 pediatric patients with CHD requiring surgical intervention. Psychomotor development was assessed using Denver Developmental Screening Test II scale few days preoperatory and then at 4-6 months after the cardiac surgery. Samples were collected immediately preoperatory and at 24 hours after the end of surgery. Enzyme-linked immunosorbent assay was used to determine GFAP concentrations.
Results: GFAP had significantly higher values postoperatory than preoperatory ($p=0.0046$). These values correlated with impairment of psychomotor general scores ($p=0.004$) and also in specific domains like personal-social domain ($p=0.04$) and fine motor domain ($p=0.03$). Receiver operatory characteristic analysis showed that GFAP is a potential neuromarker of psychomotor development impairment (AUC=0.75) with a cut off value of 0.688ng/ml.

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Conclusions: GFAP is a neuromarker that could prognose short term psychomotor impairment in patients with CHD which undergo cardiac surgery.

Keywords: congenital heart defect, heart surgery, neuromarker, glial fibrillary acidic protein, psychomotor development

PP-346

Which neuromarker is the best predictor of psychomotor impairment in children with congenital heart defects after surgery?

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Background and Aim: Psychomotor impairment occurs in an important percentage of cases in patients with congenital heart defects (CHD). These patients should be identified and treated as early as possible, because early psychomotor recovery improves long-term quality of life. The objective of this study was to identify the best neuromarker which could predict early psychomotor impairment in children with CHD after surgical intervention.
Method: Pediatric patients under 6 years with CHD undergoing elective cardiac surgery were included. Psychomotor status was assessed using the Denver Developmental Screening Test II (DDST II) which includes items grouped into four domains (personal–social behavior, fine-motor adaptive function, language, and gross motor function), preoperatively and 4–6 months postoperatively. Blood samples necessary for neuromarkers assessment were collected preoperatively and postoperatively.
Results: Forty-two patients were included and dichotomized into cyanotic (13 patients) and non-cyanotic (29 patients) groups based on peripheric oxygen saturation. Although the included patients were clinically considered as having normal neurodevelopment, 19 patients (65.5%) had abnormal developmental scores in the non-cyanotic group and 11 patients (84.6%) in the cyanotic group after DDST II evaluation, with the greatest impairment in the fine-motor and language domains in the non-cyanotic CHD group and the fine-motor and personal-social domains in cyanotic CHD group. The following neuromarkers were assessed: glial fibrillary acidic protein (GFAP), brain-derived neurotrophic factor (BDNF), protein S100 (pS100), and neuron-specific enolase (NSE). Postoperative, statistically significant higher values were seen for pS100 and GFAP, and statistically significant lower values were obtained for BDNF in the cyanotic group. A good predicting model was observed with GFAP and developmental scores in the cyanotic CHD group with an AUC of 0.667. A correlation was found between NSE and developmental quotient scores ($r = 0.09$, $p = 0.046$).
Conclusions: From all four neuromarkers studied, GFAP had a good prognostic value for short-term psychomotor impairment in cyanotic CHD patients and NSE had only a modest prognostic value. This neuromarkers could be used in developing neuroprotective perioperative strategies in order to reduce neurological insult of these patients.

Keywords: congenital heart defect, neuromarker, glial fibrillary acidic protein, neuron-specific enolase, neurodevelopment

PP-347

The hybrid palliation for hypoplastic left heart syndrome: surgical rationale, survival, and neurodevelopmental outcomes at two years of age

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Background and Aim: The rationale for hybrid palliation (HP) for hypoplastic left heart syndrome (HLHS) and its variants varies significantly between centers. The Stollery Children's Hospital is one of the largest Canadian centers for HLHS surgical management. To date, information is lacking on rationale and outcomes of the HP at this center. To describe the surgical rationale, survival and 2-year-old neurodevelopmental outcomes for children undergoing the HP at Stollery Children's Hospital.
Method: Historical cohort study from the Complex Pediatric Therapies Follow-up Program (CPTFP). All children with HLHS and its variants who underwent the HP from 2008-2022 at Stollery Children's Hospital were identified. Pre-intra-post surgical variables and outcomes were prospectively collected, charts were reviewed retrospectively to determine rationale for HP. Surviving children underwent neurodevelopmental assessment (Bayley-3 & ABAS) at two years. Descriptive analyses were performed.
Results: 14 infants underwent HP at a median age of 15 days (IQR 7-20) (64 %male,80% prenatal diagnosis). Rationale for HP: poor candidates for a Norwood procedure either due to small size (n=4, right ventricular dysfunction (n=1), coronary sinusoids (n=1), severe atrioventricular valvar regurgitation (n=2) or neonatal hypoxic ischemic encephalopathy/reluctance to go on cardiopulmonary bypass (n=1). Five infants had HP as a "bridge to decision" for borderline left heart structures to see if they would be a future candidate for biventricular repair. Four infants had an orthotopic heart transplant with two survivors; three required mechanical circulatory support (VAD, n=2 and ECMO, n=1) with one survivor. Three (21.4%) children (all with HP as a bridge to decision for small left heart structures), survived to 2 years of age and underwent neurodevelopmental assessment. Mean Bayley scores: cognitive 90(10), Language 90.7(9.3), Motor 99.3(8.1), ABAS-GAC 96.7(10.4).
Conclusions: At our institution, the HP was used for infants with risk factors who were not candidates for the Norwood procedure, or as a temporizing solution in infants with borderline left heart structures. Survival was low and only among patients with borderline left heart structures who had a biventricular repair or an orthotopic heart transplant. Neurodevelopmental outcomes for surviving children were in the average range for their age. This is relevant information to help guide discussions with families.

Keywords: Hypoplastic Left Heart Syndrome, Child, Canada

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Nursing and Allied Health Professionals

PP-348

Improving appropriate parenteral nutrition prescribing and transition to oral feeding

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Background and Aim: The incidence of necrotising enterocolitis (NEC) is higher in those with congenital heart defects compared to healthy infants. Patients considered to be high risk or have developed NEC often have enteral nutrition withheld for prolonged periods with parenteral nutrition (PN) provided. PN poses an infection risk and may delay establishment of enteral feeds. Evidence shows there is minimal nutritional benefit when PN is provided for less than 5 days. We therefore aimed to assess whether through introduction of feeding and PN protocols we could reduce inadequate PN prescribing and improve feeding outcomes.
Method: The primary objectives were to determine if PN provision could be improved to only provide it to babies who would nutritionally benefit, and to review related feeding outcomes. All babies with a congenital cardiac diagnosis provided with PN from 2017- early 2019 and 2021- early 2023 discharged from a single tertiary unit were included. Feeding and PN protocols were rolled out in 2020. Feeding and PN data was retrospectively reviewed for pre and post-operative periods, including length of time on PN, day from procedure until discharge and feeding route at discharge. Patients provided with PN for <5days were classed as having inadequate PN provision.
Results: 161 infants were included. Pre-protocol there were 39 patients given PN for an average of 26 days. Post-protocol 121 patients were given PN for an average of 11 days. Inadequate PN provision improved post-protocol from 17.9% to 12.4% of patients receiving <5 days of PN. Patients were discharged post procedure at an average of 27 days in the post-protocol group, which was faster than the 44 day average in the pre-protocol group. Patients discharged home on part oral feeds also increase post-protocol at 91%, compared to 69% pre-protocol, and full oral feeds at 71% post-protocol, 56% pre-protocol. Patients who sadly passed away before discharge were not included in these feeding outcome figures.
Conclusions: Feeding and PN protocols can help to reduce the use of PN in patients who will not nutritionally benefit. These protocols also helped to establish feeding quicker, transitioning off PN which led to improved oral feeding rates at discharge.

Keywords: Parenteral nutrition, NEC, Oral feeding, Enteral feeding, Necrotising Enterocolitis

PP-349

How does prescribing of anti-reflux medication in infants with shunt dependant diagnoses affect reflux symptoms and growth?

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Background and Aim: Gastroesophageal reflux disease (GORD) is common in infants with congenital heart disease (CHD) which is linked with poor growth and aspiration pneumonia in this cohort. Symptom management involves optimisation of enteral feeds and provision of pharmaceutical agents to support with gastric emptying and reduction of gastric acidity. Proton pump inhibitors (PPI) and prokinetic agents are often used as the first line treatment, but there are concerns they are commonly inappropriately and/or over-prescribed, which can be detrimental to patient care due to risks of cardiotoxicity.
Method: A retrospective review was conducted looking at shunt dependant infants discharged from a regional cardiac unit between 2020-2023. Growth, presence of reflux symptoms and anti-reflux medication prescribed were recorded at discharge from hospital and 3-months post-discharge. Medication doses were reviewed against the UK national formulary for children. *Results:* 94 patients were included in the discharge data and 71 included in 3-month data, as 23 patients were readmitted into hospital or had died before the 3-month point. At discharge 30% (n=28) had reflux symptoms, with 29% (n=8) of on suboptimal anti-reflux prescriptions. Whilst 70% (n=66) had no reflux symptoms, with only one infant on suboptimal medication (2%). At 3 months 28% (n=20) had reflux symptoms, and 70% (n=14) of these had suboptimal anti-reflux prescriptions. Whilst 72% (n=51) had no reflux symptoms, and 11% (n=6) of these had suboptimal medication. Overall, 24% (n=17) of infants had poor growth across the 3 months, with 35% (n=6) of these having a period reporting reflux symptoms and not on optimised anti-reflux medication.
Conclusions: Treatment of GORD with pharmaceutical agents is common, but there is strong evidence of over-prescribing and under-prescribing, which increased over time post discharge. The lack of optimisation of reflux management could be linked with the poor growth seen, and presents as an opportunity to improve patient care and outcomes. Ensuring regular medication reviews from an experienced practitioner would support both the symptom management of GORD and appropriate provision of potentially toxic medication through appropriate prescribing.

Keywords: GORD, Reflux, PPI, Growth

PP-351

A retrospective review: pre-intervention nutrition support in prostaglandin e2 dependent neonates with critical congenital heart disease

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Background and Aim: The decision to enterally feed neonates with critical congenital heart disease (CCHD) on prostaglandin E2 (PGE2) is controversial due to the risk of necrotizing enterocolitis (NEC). Recent evidence suggests that enteral nutrition (EN) is possible and highlights the clinical benefits of trophic feeds and breastmilk (BM). Guidelines (ESPNIC) recommend EN for neonates on PGE2 infusions in a critical care unit with adequate observation and monitoring. Historically, at Children's Health Ireland neonates were not fed enterally. In 2018 a feeding assessment tool was introduced. Evaluate pre-operative nutrition support (NS) and incidence of NEC. Explore the relationship between NS and outcome measures. Assess the feeding assessment tool.
Method: Design: A retrospective study. Inclusion Criteria: Neonates (<14 days) with CCHD on PGE2 infusions who underwent cardiac catherisation or surgery. Data collected included: demographics, anthropometry, cardiac diagnosis, procedural, pre-intervention NS provision and outcomes variables. Weight for age z-scores (WAZ) were calculated using WHO standards.

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Results: Seventy one neonates were included. Pre-operative NEC occurred in 1.4% (n=1) of neonates. 53.5% received EN, 28.2% received PN and 35.2% received no NS pre-intervention. No NS was provided on 37% (n=93) of admission days (cumulative total). There was no significant difference in pre-intervention NEC, hospital length of stay, days to full feeds and discharge WAZ in those that received NS pre-intervention versus those that received no NS. On discharge, 60.5% had a WAZ <-1 and 30.2% had a WAZ <-2 (moderate malnutrition). The risk assessment tool was used with 49% (n=20) of ward admissions. Pre-operatively, 21% of those with a risk factor for NEC, were fed volumes >24mls/kg enterally. Only 30% received oral feeds pre-operatively. Pre intervention, 44% (n=31) received some BM feeds. On discharge, 71% (n=41) were still receiving some BM, with 7% (n=4) being exclusively breastfed.
Conclusions: Despite the tool being used on 50% of admission, this study showed EN is possible in some neonates with CCHD without increasing the incidence of NEC. The rate of BM feeding is encouraging, however direct breastfeeding rates on discharge is low. The incidence of malnutrition during neonatal admission is high, but not surprising considering rate of NS.

Keywords: Breastmilk, Prostaglandin, Nutrition, Enteral Nutrition, Weight

PP-352

Assessment of a nurse-dietitian led paediatric cardiology clinic in western ireland: a service evaluation

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Background and Aim: Infants diagnosed with complex congenital heart defects (CHD) necessitate consistent nursing and nutrition intervention both before and after surgery to optimise clinical outcomes. Traditionally, families faced the challenge of traveling extensive distances, often hundreds of kilometres, to Children's Health Ireland at Crumlin, Dublin, for all required appointments. Recognising the need for accessible care, in 2022, a nurse-dietitian led clinic was established at Galway University Hospital to provide local support for infants with complex CHD, easing the burden on families and enhancing healthcare accessibility in the region.
This service evaluation sought to investigate how the establishment of a nurse-dietitian led clinic locally has influenced the experiences of families caring for infants with complex CHD.
Method: Caregivers voluntarily filled out an electronic service evaluation questionnaire, achieving a response rate of 78% (n=14).
Results: Caregivers consistently perceived the clinic as "very helpful, informative and supportive." A unanimous 100% reported the convenience of having the clinic nearby. Citing time and cost savings and reduced stress associated with travel. They expressed peace of mind, emphasising the quick reassurance, and prompt adjustments to diet and care when needed. Similarly, 100% found joint consultation with both nurse and dietitian beneficial, streamlining multiple medical visits into one. However, parking emerged as a universal stressor for all caregivers attending the clinic. Respondents indicated a desire for additional support services, including occupation therapy, speech and language therapy, play therapy and a lactation consultant as part of the local multidisciplinary team. While 53% expressed interest in a written nutrition plan, 67% expressed a need for more written information on their child's medical condition. Notably, all caregivers found it easy to access the dietitian and nurse, receiving prompt responses.
Conclusions: The nurse-dietitian led clinic demonstrated a noteworthy overall positive satisfaction among participants. While the feedback was generally favourable, areas for improvement and identified, with a particular emphasis on enhancing written information for delivery. These findings underscore the clinic's commitment to continuous quality enhancement, ensuring a more comprehensive and effective healthcare experience for individuals seeking nutritional guidance and support.

Keywords: Clinic, service evaluation, paediatric cardiology, nurse, dietitian

PP-353

Parental anxiety, depression, and sense of coherence of neonates with congenital heart defect: a cross-sectional study

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Background and Aim: Parental psychosocial stress symptoms shape the cognitive and socio-emotional development of neonates with congenital heart defect (CHD). The Sense of Coherence (SOC) can support health-promoting stress management in burdensome situations. Yet, data on anxiety and depression compared to the SOC among parents of a neonate with CHD is missing.
To assess the frequency, severity, correlations and differences in anxiety, depression, and SOC in parents of an inpatient neonate with pre- or postnatal diagnosed CHD.
Method: A descriptive online cross-sectional study at the university children's hospital Zurich using the Hospital Anxiety and Depression Scale in German (HADS-D), the revised SOC (SOC-R), and sociodemographic and disease-related data. Hypotheses were evaluated descriptively, and the correlations were evaluated with the Spearman's correlation coefficient (Rho).
Results: 23 parents of 15 newborns with severe CHD participated. In the case of HADS-D, twelve parents were above the cut-off (>7) in the anxiety subscale and eight in the depression subscale. In the anxiety subscale, the parents' mean score was 8.39 (SD = 3.96) and the median was 8 (min.-max. = 3-17). The depression subscale had a significantly lower mean of 5.65 (SD = 4.73) and a median of 4 (min. max. = 0-17). The correlation between anxiety and depression in HADS-D shows strong positive effects (Rho =.681; p-value =.000). The SOC-R has a strong negative effect with the HADS-D in the anxiety subscale (Rho = –.436; p =.042) and a medium negative effect in the depression subscale (Rho = –.217; p =.329).
Conclusions: This study group showed a high SOC, which implies healthy promotion of stress management and resources to deal with stressful life situations. A strong SOC supports parents of neonates with congenital heart defects and provides an approach for interventions. It is crucial to develop pre- and postnatal salutogenic family-centred care interventions. This includes early involvement of parents caring for their neonates supervised by skilled nurses, thus promoting multi-professional cooperation and communication. These strategies would

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support parents in caring for their neonates and therefore strengthen the neonate's long-term cognitive and socioemotional development.

Keywords: anxiety, depression, sense of coherence, parents, congenital heart defect

PP-354

Mapping the landscape of paediatric cardiology advanced nurse practitioners (PCANP'S) in the United Kingdom (U.K)

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Background and Aim: The first U.K. PCANP peer network was established in August 2023. Initial aims were (1) provision of support for advanced clinical practice within paediatric cardiac nursing (2) introduction of a speciality specific education programme (3) collaborative research.
The network's initial project is a U.K-wide scoping exercise of the PCANP role. The role was benchmarked against the 4 pillars of the ANP role in the absence of U.K. PCANP implemented standards.
Method: Invitations to the network were extended via email to all PCANP teams in the U.K. and Republic of Ireland (ROI). After an initial qualitative scoping session, snowball sampling was used to identify PCANPs across all 14 U.K. centres and questionnaires distributed.
Results: 7 ANP's from 6 centres attended the scoping session. Questionnaire responses were received from 89% of centres with PCANPs. These 8 centres have 19.5 qualified and 3 trainee PCANPs with a median of 2.4 (range 1-10) qualified PCANP's/centre. 5 respondents were the sole ANP in their department.
Although the majority are in surgical centres, two surgical centres do not have PCANPs and one PCANP is in a non-surgical centre. All respondents spent at least 80% (range 80-100%) of their time delivering direct clinical care. There was variation in specific roles with 95% of PCANP's covering more than one aspect of the patient journey. Each centre included ward cover in their provision. The greatest role variation was seen in outpatient care where roles covered the breadth of the speciality from ANP-led pre-assessment (15%), delivering care to fragile pre-operative infants (5%) and subspecialty care e.g. inherited cardiac conditions (5%), arrhythmia (5%) and single ventricle physiology (5%).
Only one centre provided designated teaching for PCANPs. However, all PCANPs are involved in teaching and training a wide range of professionals.
Four teams have published research in the last 2 years.
Conclusions: A peer network has been used to map the U.K PCANP landscape. The role has been developed in most U.K. cardiac surgical centres, is predominantly clinical, but demonstrates important variation in its specific delivery. This suggests individual services are empowered to developed the role according to their specific needs.

Keywords: anp, advanced nurse practitioner, scope

PP-355

Nurse-led congenital heart defect (CHD) vulnerable patient telephone clinic

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Background and Aim: The Paediatric Congenital Heart Disease (CHD) Clinical Nurse Specialist (CNS) team service has adapted to provide and pro-active service by delivering a tailored telephone call service for our most complex CHD patients with vulnerability to developing symptoms of heart failure based on the Congenital Heart Assessment Tool (CHAT) (Gaskin et al., 2023).
Method: The CNS team researched appropriate methods of creating an improved service to support patients and families with a CHD vulnerable to developing signs of heart failure. The CNS team used CHAT as a basis to create a series of key questions to evaluate CHD patient symptoms via a telephone call. The CNS team also designed an inclusion criteria of patients who are vulnerable to developing heart failure.
The patient families now receive a personalised folder containing information specific to their child's CHD, cardiac red flag sign and symptoms, treatment plan.
The CHAT tool enables professionals to identify the patients clinical condition using an early warning score system.
Results: The eligibility criteria for this service have been tailored as the service has progressed. The CNS team have created a criteria based on patient vulnerability for patients likely to go into heart failure pre-surgery, which has also determined patient requirement of calls such as weekly, fortnightly or monthly.
The CNS team feel that we have recognised early warning signs of heart failure and provided early escalation for intervention – assessment – medication alterations – feeding support, weight gain/loss (referral onto other services) reduced hospital admissions, helped families utilise local services appropriately such as GP/A&E and specialist care as needed.
Conclusions: The implementation of the Nurse-led vulnerable patient telephone consultations has aided identifying early signs of heart failure, expedited assessments, helped avoid frequent hospital visits and when required, facilitating escalation of care. It has been considered by colleagues and families that this level of support has helped ease their anxiety in the pre-operative period.

Keywords: paediatric, cardiology, nurse specialist, telephone clinic

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Paediatric Cardiovascular Intensive Care

PP-356

Representation learning for the prediction of intensive care unit length of stay in infants with congenital heart disease undergoing cardiac surgery

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Background and Aim: Congenital Heart Disease (CHD) is the most common birth defect and it has a significant impact on health, mortality, and healthcare (HC) costs. About 30% of these children require at least one cardiac procedure during their first years of life. An accurate estimation of the intensive care unit length of stay (ICULOS) after cardiac surgery could help HC management to improve resources allocation. While some perioperative risk factors prolonging ICULOS after cardiac surgery have been identified, tools to accurately estimate ICULOS for these patients are insufficient. Thus, this study aims to predict ICULOS in pediatric CHD patients undergoing cardiac surgery by employing explainable Representation Learning (RL).
Method: Demographic (age, sex, main disease) and perioperative data (surgery type and duration, cardiopulmonary bypass (CPB) time, preoperative laboratory data, etc.) from infants (1-24 month old) with CHD undergoing cardiac surgery between September 2020 and March 2023 was collected. We used unsupervised Multi Kernel Learning (MKL) to reduce data dimensionality and position patients based on data similarity. Finally, a binary random forest (RF) classifier was implemented using the low dimensional data to predict if patients would stay in the ICU more than 6 days. Data was randomly split into training and test datasets (75/25). Five-fold cross validation was performed to select the best model (Figure 1A).
Results: 134 patients (54% male, age= 8.22±5.37 months, ICULOS= 3 (2, 6) days) were included. Regarding the low-dimensional space, it stabilizes at the seventh dimension. Age, location before the surgery, surgery duration or STAT score among other variables show a significant correlation with at least one of the seven first dimensions (Figure 1B). In the test subset, the predictive model demonstrated a good performance to predict ICULOS> 6 days with an accuracy of 0.88, an area under the ROC curve (AUC) of 0.87, and an F1-score of 0.82.
Conclusions: Our results serve as a proof of concept that explainable RL can be useful to predict if a CHD infant undergoing a cardiac surgery will stay in the ICU more than 6 days, using demographic and perioperative surgery. Further experiments including new data are warranted to further validate our model.

Keywords: Intensive Care Unit (ICU), Machine learning (ML), Representation Learning (RL), Congenital Heart Disease (CHD), Length of stay (LOS)

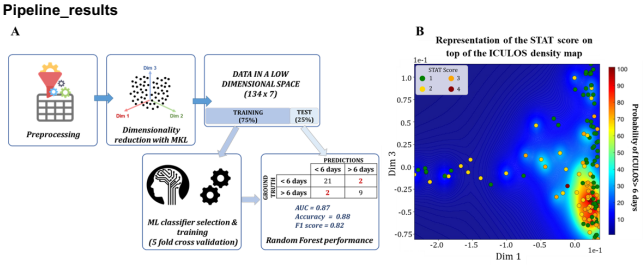

Figure 1. **A.** Schematic of the methodology implemented. **B.** Example of relevant variable (STAT score) scattered on top of the ICULOS density map. Each dot represents the patient STAT surgery score and the background the density of patients with an ICULOS larger than 6 days.

PP-358

Incidence of patent ductus arteriosus (PDA) in premature newborn at the tertiary care rural hospital of central India

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Background and Aim: One of the most common congenital heart diseases among preterm newborn is PDA. There is a higher incidence of PDA with lower birth weights and gestational ages. To investigate the incidence of PDA in premature infants by using echocardiography.
Method: Preterm newborn delivered between January 2021 and December 2022 were included in this study. All preterm newborns had their first echocardiogram at 3 days of age. Echocardiography was repeated at the chronological age of 7 days if PDA was detected.
Results: During the study period, eighty-seven preterm neonates participated in this study and underwent echocardiography. In the first echocardiogram, the incidence rate of PDA was 38 (43.67%). At gestational age of less than 28 weeks, 12 of 18 newborns had PDA. Of 24 newborns with birth weight of more than 1000 grams and less than 2500gms, 14 had PDA. Newborns with birth weight of more than 2500 grams(n=45), 12 had PDA. On the second echocardiography, the incidence rate of PDA was 18% and mostly found in neonates at gestational age of less than 28 weeks and in those with birth weight of less than 1000 grams.
Conclusions: Preterm neonates had an 18% incidence rate of PDA. Those who are born with a low birth weight are more likely to suffer from PDA.

Keywords: Preterm, Neonates, Patent ductus arteriosus, Low birth weight

PP-361

Establishment of pre-operative pediatric cardiac intensive care unit in a tertiary care teaching hospital in an lmic: a quality improvement initiative

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Background and Aim: Survival of children with congenital heart disease (CHD) has improved markedly in high-income countries, as

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against the low- and middle-income countries (LMICs). Many present in critically ill state with malnutrition, respiratory distress, shock and/or pneumonia, and need pre-operative stabilization. Data are scarce on clinical profile and outcome of these pre-operative critically ill ones from LMICs. We assessed impact of a newly commissioned pre-operative Pediatric Cardiac Intensive Care Unit (PCICU) on clinical profile, critical care needs and outcome of these patients.

Method: A new 10-bed unit was commissioned on Nov 2018 to manage pre-operative critically ill children with CHD presenting in Pediatric Emergency of a tertiary care teaching hospital in an LMCI. The unit started in the ward setting with 2 monitors and 1 ventilator, and subsequently shifted to the fully-functional formal ICU (with 10 multiparameter monitors and ICU beds, 5 ventilators, 4 high-flow nasal cannula, echocardiography, point-of-care ultrasound and blood gas machines) on Oct 2021. Janani Shishu Suraksha Karyakaram (JSSK), from April 2022, provided free treatment to infants. Medical records were reviewed retrospectively (Nov, 2018–Sept, 2021) and prospectively (Oct 2021-Oct, 2023). Impact of starting a formal intensive care services and JSSK was assessed on clinical characteristics, critical care needs, pre-operative and post-operative outcome.

Results: Total of 1098 patients with CHD were admitted during the study period. Complete information could be retrieved for 1073 patients [ward setting, n=533; ICU without JSSK, n=67; ICU with JSSK, n=287] and these included non cardiac(70), immediate post op(19),post op follow up(57) and acquired heart diseases(40). Ventricular septal defects was the commonest defect (n=230, 25.9%), followed by d TGA (n=124, 13.9%), TOF (n=93, 10.4%), TAPVC (n=71, 8%), PDA (n=59, 6.6%), double outlet right ventricle (n=51, 5.7%), and coarctation of aorta (n=48; 5.4%). Clinical profile and outcome of patients are shown in the table 1.

Conclusions: With provision of specialised care to critically ill pre-operative children with CHD, significantly higher number of younger babies could be managed. This led to significantly increased utilization of critical care interventions, and thus increased the number of children who could undergo surgery post-stabilization. However, this could not reduce the proportion of pre-operative deaths.

Keywords: Pediatric Cardiac Intensive Care Unit, Critical care needs, Outcome, Quality improvement.

Results table

Characteristics		Ward Setting (n=533) (1)	ICU without JSSK (n=67) (2)	ICU with JSSK(n=287) (3)	p-value
Gender	Male	348	43	180	0.637#
	Female	185	24	106	
	Ambiguous	0	0	1	
Age (days) [Median (IQR)]		108(35,226)	42(13,120)	68(20,180)	<0.001* 1-2:0.0005** 1-3:0.0038 2-3:0.1119
Age Groups	First Week	56	11	42	0.058#
	Post 7-days Neonates	75	22	62	
	Post Neonatal Infants	318	27	144	
	Post-infancy	84	7	39	
Pre-operative Non - invasive mechanical ventilation, n (%)		328(61.5%)	44(65.6%)	208(72.4%)	0.007#
Pre-operative NIMV duration among survivors (days) [Median (IQR)]		5(3,13)	9(5,16)	8(4,13)	0.006* 1-2:0.04** 1-3:0.0073 2-3:0.900
Need of Ventilation, n (%)		204(38.2%)	31(46.2%)	143(49.8%)	0.005#
Pre-operative Ventilation Duration among survivors (days) [Median (IQR)]		11(4,24)	11(4,18)	7(3,13)	0.3*
Need of Inotropes, n (%)		183(34.3%)	21(31.3%)	104(36.2%)	0.718#
Inotrope duration among survivors (days) [Median (IQR)]		3(2,5)	5(3,6)	3(2,6)	0.4*
Maximum Vasoactive Inotropic Score [Median (IQR)]		20(10,42)	20(20,60)	30(20,60)	0.205*
Pre-Operative ICU Stay among preop survivors(days) [Median (IQR)]		24(11;36.5)	11(5;27)	17(9.5;28.5)	0.157*
Pre-Operative ICU Stay among those who underwent surgery (days) [Median (IQR)]		17(9,25)	13(9,22)	12(6,20)	0.006* 1-2:0.45** 1-3:0.002 2-3 :0.50
Pre-operative Outcome	Discharge	318(60%)	41(61%)	185(65%)	0.003#
	Deaths, n (%)	120(22.6%)	18(26.8%)	78(27.4%)	
	LAMA, n (%)	92(17.3%)	8(12.5%)	21(7%)	
	Transfer for surgery among survivors	150(41.4%)	31(62%)	117(54%)	0.003*
Post-operative Outcome among those went for surgery (n=298)	Discharge	106(71%)	21(68%)	82(72%)	0.346#
	Death/LAMA/DOR	43(29%)	10(32%)	32(28%)	
Total Hospital Stay who got operated and survived (days) [Median (IQR)]		43(31,59)	37(26,52)	37(26,50)	0.065*
#: Chi-square test *: Kruskal Wallis test **: Dunn's test					

The below mentioned table consists of observations of critical care needs of the enrolled cases

PP-362

Presepsin may differentiate pge-1 induced systemic inflammation from sepsis: A prospective study

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Background and Aim: Prostaglandin E1 (PGE1) infusion maintains ductal patency in duct-dependent congenital heart disease(CHD) while awaiting surgery. Delay in surgery necessitates prolong PGE1 infusion, and predisposes to healthcare-associated infections. Considering difficulty in differentiating PGE1-induced systemic inflammation from neonatal sepsis, we planned to study presepsin and inflammatory markers in patients on PGE1 infusion for >5 days with & without sepsis.

Method: Baseline demographic, clinical and laboratory characteristics were recorded at enrolment, then every 5 days and when sepsis was suspected, until end points (surgery or death). Sepsis defined by clinical suspicion with positive body fluid culture, and/or pneumonia on X-ray, or improvement in inflammatory markers with antibiotics. Presepsin and inflammatory markers were compared at observation points when patients did and did not have sepsis. Data were expressed as median (IQR).

Results: Thirty patients [male, 18; age, 11 (7, 17.5) days] with CHD were enrolled. PGE1 infusion was started at 5 (3, 12) days of life at 20 (12.5, 50) ng/kg/min. PGE1 was given for 21 (11, 27) days at cumulative dose of 388 (216, 691) mcg/kg. Out of 89 observation points, 78 were non-sepsis, while 11 observation points were during sepsis episodes. Cumulative dose and duration of PGE1 infusion were similar in two observation groups [648 (374, 979) vs 324 (216, 590); p=0.2 and 22 (10, 27) vs 20 (12, 27); p=0.44 respectively], so was TLC and ANC. Platelet count was lower during sepsis episodes [190 (73, 373) vs 360 (260, 480); p=0.001], while CRP (mg/dl) [38 (24, 100) vs 4 (2.5, 6.4); p<0.001], procalcitonin (ng/ml) [2.03 (0.49, 8.12) vs 0.19 (0.10, 0.46); p<0.001] and presepsin (ng/ml) [2.99 (2.79, 3.64) vs 1.91 (1.72, 2.103); p<0.001] were higher. CRP and procalcitonin cut offs to identify sepsis were 19.55 mg/dl (AUC, 0.95; sensitivity, 91.7%; specificity, 92.2%) and 0.29 ng/ml (AUC, 0.95; sensitivity, 91.7%; specificity, 75.3%) respectively. Presepsin of 2.17 ng/ml had high predictability for sepsis (AUC, 0.99; sensitivity, 100%; specificity, 94.7%).

Conclusions: Prespsin and procalcitonin seem to help identify sepsis in patients on PGE1 for >5 days. CRP >20mg/dL also suggests sepsis. Findings need further confirmation.

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Keywords: Prostaglandin E1, Presepsin, Sepsis, Duct-dependent lesion, Systemic Inflammation, Congenital Heart Disease

PP-363

Metabolic effects of prolong pge-1 infusion: a prospective observational study

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Background and Aim: Prostaglandin E1 (PGE1) infusion is used to maintain ductal patency in duct-dependent critical congenital heart disease (CCHD) while awaiting surgery. Delay in surgery necessitates prolong PGE1 infusion, especially in resource-constraint settings. Cases of pseudo-barter syndrome and cortical hyperostosis have been reported with prolong use of PGE1 infusion. However, there is paucity of substantial clinical data. Hence, planned to study the impact of prolong (>5 days) PGE1 infusion on electrolytes and bone metabolism.
Method: Neonates with echocardiographic evidence of duct-dependent CCHD and PGE1 infusion for 5 days were enrolled. Culture-positivity at enrolment were excluded. Baseline demographic, clinical and biochemical parameters were recorded on day 1 of enrolment, and then every 5 days until surgery or death, whichever was earlier. Data are expressed as median (IQR).
Results: Thirty neonates [males, 18; age, 11(7, 17.5) days] with CCHD were enrolled. PGE1 infusion was started at 5(3, 12.25) days of life at 20(12.5, 50) ng/kg/min. Patients received PGE1 for 21(11.25, 27) days. Cumulative dose was 388.8(216, 691.2) mcg/kg, while maximum rate of infusion was 40 (30, 57) ng/kg/min. We had total of 89 observation points, with average of three observation points per patient. At enrolment, serum levels of sodium, potassium, chloride, calcium, phosphorus and ALP were 139 (132, 141), 5.1 (4.4, 6.0), 103 (101, 106), 8.4 (8.2, 8.6), 4.8 (4.2, 5.3) and 128 (114, 154) respectively. A negative correlation was noted between cumulative dose of PGE1 with sodium (R=-0.25, p=0.017), chloride (R=-0.21, p=0.05) and potassium (R=-0.18, p=0.082). Cumulative dose of PGE1 was positively correlated with ALP [R=0.35, p=0.0007] and serum phosphorus [R=0.24, p=0.023]. Calcium did not get affected. Two patients developed clinical and radiological evidence of cortical hyperostosis, while a patient developed pseudo-barter syndrome due to urinary loss of electrolytes (Na+, 110mEq/L, K+, 2.6mEq/L).
Conclusions: Prolong PGE1 affects electrolytes and bone metabolism, manifesting clinically as pseudo-barter syndrome and cortical hyperostosis in some. Patients on PGE1 infusion for more than 5 days need meticulous biochemical monitoring for early identification and intervention.

Keywords: Prostaglandin E1, Congenital Heart disease, duct-dependent lesion, Electrolytes, Bone metabolism

PP-364

Atrial flutter in the newborn, our experience

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Background and Aim: Atrial flutter is a well-known arrhythmia that occurs in the perinatal period, often in the third trimester of pregnancy or within the first two days of life. In infants without congenital heart disease (CHD) or ventricular dysfunction, the recurrence risk is low once flutter has converted to sinus rhythm. However, managing atrial flutter in such scenarios can be challenging.
Method: We present a case of a newborn diagnosed with prenatal hydrops fetalis due to atrial flutter with severe myocardial dysfunction (EF 25%) that was confirmed after birth. Although electric cardioversion was effective, atrial flutter recurred several times even under intravenous amiodarone, leading to refractory cardiogenic shock.
Results: We decided to initiate extracorporeal membrane oxygenation (ECMO) support for clinical stabilization and to administer cardio depressive drugs for heart rhythm control, such as amiodarone and digoxin, achieving continuous sinus rhythm after two days of this treatment. Myocardial function completely recovered on day 7, ECMO was withdrawn on day 7, and amiodarone was swift to propranolol on day 18. Finally, the patient was discharged asymptomatic on day 30 with a left ventricular ejection fraction (LVEF) of 70% and sinus rhythm under digoxin and propranolol. After eight months of follow-up, no recurrences have been detected.
Conclusions: Based on meta-analysis evidence, direct current cardioversion is the most cost-efficient method of cardioverting a neonate with atrial flutter to sinus rhythm. This is especially relevant in cases with severe cardiac dysfunction and critically ill neonates, where the administration of heart rate control medications such as beta blockers or cardio-depressive antiarrhythmic drugs such as flecainide and amiodarone could be deleterious. Furthermore, supporting with ECMO after cardioversion was crucial in our patient, leading to stabilization, normalization of cardiac function, and concomitant administration of such medications.

Keywords: ECMO, auricular taquicardia, flutter, newborn.

PP-365

Cardiac manifestations in multisystem inflammatory syndrome in children

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Background and Aim: Multisystem inflammatory syndrome in children (MIS-C) refers to a new condition that was first described in April 2020. The pathophysiology is still not well understood; however, cardiac involvement is common. The aim of this study was to characterize patients hospitalized in a tertiary hospital with cardiac manifestations of MIS-C.
Method: An observational, cross-sectional and retrospective study was performed including patients with MIS-C and cardiac manifestations hospitalized between April/2020 and June/2022 at a single tertiary center.
Results: The study included 28 patients diagnosed with MIS-C, of which 19 (68%) had cardiac involvement. The majority (74%) were male with a median age of 10 years (range: 3.5-17.8 years). Approximately 79% (n=15) had positive anti-SARS-CoV-2 IgG antibodies. All patients presented elevated cardiac markers: the

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median troponin-I level was 292 (maximum 23449) ng/L, BNP was 528 (maximum 3961) pg/mL and myoglobin was 120 (maximum 1904) ng/mL. Only one had ventricular dysfunction upon admission with an ejection fraction of 40%, 5 developed dysfunction later and 7 presented coronary artery ectasia. The median C-reactive protein (CRP) was 262 (maximum 394) mg/L and procalcitonin was 6 (maximum 308) ng/mL in 16 patients. The majority (95%) received treatment with immunoglobulin, corticosteroids and antiplatelet dose of acetylsalicylic acid. Ten (53%) were admitted to the pediatric intensive care unit (PICU) and required support with aminergic drugs; in this group of patients the median troponin-I level was 1275 ng/L and BNP was 993 pg/mL. The median length of hospital stay was 12 days. There were no fatalities. Being >10 years old, male, with higher values of CRP and cardiac markers and ventricular dysfunction upon admission were all related with statistical significance to the need of PICU care. Also ventricular dysfunction was related with statistical significance to higher cardiac markers values upon admission and hospital stay. Regarding coronary artery ectasia, all patients presented normal coronary diameter in one year of follow-up.
Conclusions: Cardiac involvement in MIS-C can manifest with changes in cardiac markers, ventricular dysfunction and coronary abnormalities. The authors found that patients who presented ventricular dysfunction presented higher cardiac markers values. Part of the treatment involves immunomodulation and antiplatelet therapy.

Keywords: multisystem inflammatory syndrome in children, pediatric

PP-366

Perioperative malnutrition as a risk factor for adverse outcomes after pediatric cardiac surgery

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Background and Aim: Malnutrition has been thoroughly investigated and proven to be a risk factor for surgical outcomes in children with congenital heart disease (CHD). The aim of this study is to analyse the outcomes of surgical treatment of children with CHD and to determine the association of malnutrition with early postoperative mortality, complications and hospital stay.
Method: All patients up to 18 years of age who underwent cardiac surgery for 2 years period (between 2016-2017) were studied. Data of all patients who met the inclusion criteria were analyzed. Patients were divided into 2 groups according to their weight - group 1 underweight and group 2 normal or overweight. The groups were compared in terms of early postoperative mortality, postoperative complications, duration of invasive mechanical ventilation (IMV), and length of hospital stay. Data are presented as medians with interquartile range or means with standard deviation. Non-parametric Mann-Whitney U test analysis integrated into SPSS 19.0 statistical software was used. Values of $P<0.05$ were considered statistically significant.
Results: 384 patients were included in the analysis. Children who were underweight (group 1) were more likely to have prolonged IMV 29.5% vs. 16.8% (p=0.004), systemic infection 13.9% vs. 7.6% (p=0.05), pulmonary complications 15.6% vs. 8.8%(p=0.047), longer ICU stay 5days(8) vs. 4days(5) and in-hospital stay 17.5days(11.25) vs. 14days(10.25). These children also had a higher risk of early postoperative death 16.8% vs 8.6% (p=0.014).
Conclusions: Malnutrition is significantly associated with complications and adverse outcomes after cardiac surgery in children with CHD. Introduction and adherence to feeding recommendations, in an attempt to reduce the degree of malnutrition, could effectively lower perioperative complications and early postoperative mortality.

Keywords: malnutrition, risk factors, cardiac surgery, congenital heart disease, children

PP-367

Highlights of knowledge about modified blalock-taussig shunt: *What we know and what we can improve*

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Background and Aim: The most commonly performed systemic-pulmonary shunt in neonates, with cyanotic congenital heart diseases, is undoubtedly a modified Blalock-Taussig shunt (MBTS). The main aim of this research was to show known risk factors associated with overall morbidity and mortality as well as to emphasize and clarify the importance of seemingly irrelevant details, which often determine the final course of this simple-looking procedure.
Method: We made a retrospective study of 68 severely cyanotic neonates, presented mostly with pulmonary atresia and Tetralogy of Fallot (TOF) with hypoplastic pulmonary arteries, who underwent a MBTS procedure from 2011-2022. Preoperative characteristics were observed, such as body weight, shunt size, thrombophilia, malformation syndrome and duration of extracorporeal circulation. Our major postoperative parameters were hematocrit, carbon dioxide and oxygen partial pressure, presence of coagulation disorders, inotropic support, reoperation, cardiopulmonary resuscitation and infection rate as an indicators of complicated course and death.
Results: Sixty-eight neonates, 41 males and 27 females, from 2 days to 9 months old (mean age 31 day) at the time of MBTS procedure, and 2,1 kg to 7,3 kg (mean 3,4 kg), were further divided into 2 groups, those entirely dependent on MBTS (58,8%) versus those with additional pulmonary blood flow (41,2%). Most patients received a 3,5 mm Gore-Tex shunt (70,5%), with mean shunt size/body weight ratio of 1,05. Postoperative mechanical ventilation mean duration was 6,3 days. A total of 17 patients died, 13 (76,5%) of them in early postoperative period (<30 days) while the other 4 in the next four months. Leading cause of death was low cardiac output syndrome (58,8%), while 11,8% of deaths were caused by shunt thrombosis and the rest occurred due to sepsis or uncontrolled bleeding. Revision of surgical procedure needed 24 patients (36,3%). The most common cardiac defect was pulmonary atresia (36,8%), followed by double outlet right ventricle with severe pulmonary stenosis (16,2%) and TOF (16,2%).
Conclusions: Integrative preoperative and postoperative approach is crucial to achieve the best possible outcome of MBTS procedure.

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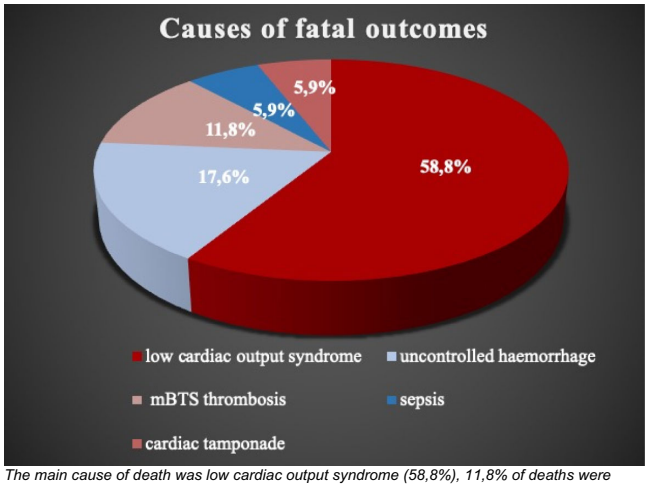
Our results suggest a constant need for precise assessment of risk factors which can be fatal such as thrombosis and to think and act always one step ahead.

Keywords: Modified Blalock-Taussig shunt, Thrombosis, Oxygen Partial Pressure, Pulmonary Atresia, Tetralogy of Fallot

Causes of fatal outcomes



The main cause of death was low cardiac output syndrome (58,8%), 11,8% of deaths were caused by shunt thrombosis, while 17,6% of them were due to uncontrolled haemorrhage, the rest of 11,8% were caused by sepsis or cardiac tamponade.

PP-368
Risk factors for perioperative pulmonary hypertension crisis after systemic-pulmonary shunt: A single-center retrospective observational study

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Background and Aim: To identify the perioperative pulmonary hypertension crisis (PHC) risk factors after systemic-pulmonary shunt (SPS) surgery for congenital heart disease.
Method: We enrolled the 155 patients who underwent SPS surgery from 2013 to 2023 in Hokkaido University. PHC was defined as sudden 20% drop in both blood pressure and SpO_2. To identify the risk factor of PHC, the ANOVA, t-, and multivariate analysis were performed for following parameters such as age, BW, chromosomal abnormalities, biventricular or Fontan candidate (B or F), preoperative pulmonary blood flow (low flow (LF)/high flow(HF)), SPS type and diameter, with or without Cardiopulmonary bypass(CPB), FiO_2, vital signs, inotrope score (IS), sedative analgesic drugs at the time of PHC or initial tracheal suction(TS).
Results: PHC occurred in 25(16%) (15 recovered within 30 min, 4 took over 30 min and not required open chest procedure, 6 required open chest or ECMO). The incidence of within 6 hours of ICU admission was 17(68%) triggered by 15 TS, 5 head cold compress, 2 echo, 2 vasoconstrictor reduction, and 1 NO discontinuation. The mean value (% or ±SD) [P] of group non-PHC vs PHC were as follow: age (year-old) 0.33 ±0.67 vs 0.10 ±0.22 [0.03], BW(kg) 4.3 ±2.3 vs 3.4 ±1.4 [0.02], chromosomal abnormalities 40(31) vs 7(28) [≥0.05], F 64(49) vs 10(40) [≥0.05], HF 17(13) vs 6(24) [0.01], SPS type (BTS/CS/RVPA) 88(68)/ 40(31)/ 2(1) vs 17(68)/ 8(32)/ 0(0) [≥0.05]/diameter (3/3.5/4.0/5.0 mm) 29/ 52/ 44/ 5 vs 9/ 10/ 6/ 0 [0.01], with CPB 47(36) vs 7(28) [≥0. 05], FiO2: 0.36 ±0.23 vs 0.53 ±0.28 [0.01], BP (mmHg) 84 ±14 vs 86 ±17 [0. 05], CVP (mmHg) 7.3 ±3.1vs 7.0 ±2.6 [≥0.05], IS 13.5 ±6.1 vs 16.9 ±9.8 [≥0.05], (dexmedetomidine/ midazolam/ chlorpromazine)/ no analgesia/ no muscle relaxant (54(42)vs 13(52)/111(85) vs 20(80)/ 87(67) vs 18(72) [all≥0.05])/ 53(41) vs 6(24) [0.01] /65(50) vs 14(64) [0.02], respectively. Multivariate analysis revealed that preoperative HF, 3.0mm SPS, $FiO_2 \geq 0.6$, no muscle relaxants were independent risk factors for PHC.
Conclusions: To prevent perioperative PHC after SPS surgery, minimal handling with muscle relaxants is recommended especially for patients with preoperative HF, 3.0mm SPS, or difficulty in reducing FiO_2 within 6 hours of ICU admission.

Keywords: pulmonary hypertension crisis, shunt surgery

PP-370
Postoperative atrioventricular block in pediatric patients: Impact of congenital cardiac malformations and medications

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Background and Aim: Postoperative atrioventricular block may occur after pediatric cardiac surgery. A small proportion of those who develop atrioventricular block will require pacemaker placement. The primary aim of this study was to determine factors associated with postoperative atrioventricular block. Secondary aims included determining factors associated with pacemaker placement in those with atrioventricular block.
Method: Data from the PHIS data was utilized to identify patients under 18 years of age who underwent cardiac surgery. Those who did and did not develop atrioventricular block were then compared. Univariable analyses and regression analyses were conducted to determine factors associated with postoperative atrioventricular block. Similar analyses were conducted to determine factors associated with pacemaker placement in those with atrioventricular block.
Results: A total of 43,716 admissions were identified. Of these, 2,093 (5%) developed atrioventricular block and 480 (1% of total admissions) underwent pacemaker placement. Approximately 70% of those with atrioventricular block received steroids but this was not associated with a decrease in pacemaker placement. Risk factors (congenital malformations of the heart, comorbidities, medications) associated with increased risk of atrioventricular block and pacemaker placement were identified.
Conclusions: Postoperative atrioventricular block occurred in 5% of pediatric admissions for cardiac surgery. Of these admissions with postoperative atrioventricular block, 23% required pacemaker placement. Isoproterenol and steroids were not associated with a reduction in the likelihood of pacemaker placement.

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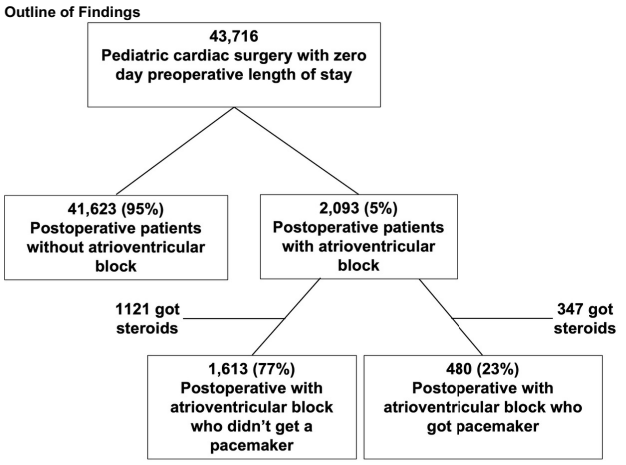
Keywords: postoperative atrioventricular block, cardiac surgery, steroids



A total of 43,716 admissions were included in the analyses. Of these 43,716 admissions, 2,093 (5%) developed postoperative atrioventricular block. Of these, 2,093 admissions who developed atrioventricular block, 480 (23%) underwent pacemaker implantation. Of the 2,093 postoperative admissions with atrioventricular block who ultimately underwent pacemaker implantation, 347 (72%) had received steroids. Of the 1,613 postoperative admissions with atrioventricular block who did not undergo pacemaker implantation, 1,121 (69%) had received steroids. The proportion of steroids in those with postoperative atrioventricular block who did or did not undergo pacemaker implantation was not statistically significantly different.

PP-371

Detecting acute kidney injury following congenital heart surgery using a complex rule-based clinical decision support system

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Background and Aim: Acute kidney injury (AKI) is a common complication after congenital heart surgery in children. To diagnose AKI, pediatricians must access and evaluate data from multiple sources, such as serum creatinine levels from laboratory information systems and urine measurements from nursing documentation. Since such data is now available in a digital machine-readable format in pediatric intensive care units, computer-based calculations can support and accelerate this process for physicians. Our aim was to develop a time-sensitive, rule-based clinical decision support system (CDSS) capable of detecting, staging and assessing the temporal progression of AKI.
Method: Clinical routine data from n=256 children who had undergone congenital heart surgery with cardiopulmonary bypass between 2015 and 2021 were randomly chosen and integrated into a standardized dataset. Based on a previous power analysis for accuracy testing of a CDSS, this cohort was enriched with several patients previously diagnosed with AKI 3 to establish a reference cohort. Criteria from the Kidney Disease Improving Global Outcome guidelines were adapted and transformed into computable logic rules to be programmed into the CDSS. To provide a reference standard for CDSS, blinded clinical experts manually evaluated and labelled the clinical course of each patient of the reference cohort for the presence, stage and duration of AKI. Using a nonparametric method that adjusts for clustered data, the sensitivity and specificity of the CDSS were determined for each stage.
Results: In the reference cohort, blinded clinical experts identified AKI incidences of 32.4% (AKI 1), 15.2% (AKI 2), and 7.8% (AKI 3). In a preliminary version, the CDSS scored sensitivities of 97.8% (IQR 95.0-99.0%) for AKI 1, 93.4% (81.6-97.8%) for AKI 2, and 99.4% (96.1-99.9%) for AKI 3. Specificities of 99.0% (97.4-99.6%), 99.4% (97.8-99.8%), and 99.7% (98.2-100%) were reached, respectively.
Conclusions: We have shown the ability of a rule-based computerized system to perform AKI detection and staging with high accuracy. An advantage of CDSS is that it can label AKI in the given data sets within a fraction of the time required by clinical experts. This benefit will be used in the future to characterize a larger data set and develop a prediction model for AKI.

Keywords: acute kidney injury, clinical decision support system, pediatric intensive care, digital medicine, congenital heart defect, cardiopulmonary bypass

PP-372

Extra corporeal membrane oxygenation (ECMO) in neonates with transposition of the great arteries (TGA): 12.5 year experience in a single-center

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Background and Aim: Extracorporeal membrane oxygenation (ECMO) can provide support for neonates with Transposition of the Great Arteries (TGA) experiencing severe cardiorespiratory failure, acting as a bridge to recovery in both pre-and postoperative TGA management. However, ECMO carries formidable challenges and the characterization of its use, outcomes and complications in TGA-patients have not been widely studied.
Method: Retrospective single-center study encompassing all TGA patients between January 2011 and September 2023 who underwent ECMO at Leiden University Medical Center, the Netherlands. Data are presented as median with interquartile range (IQR). Chi-square and Mann-Whitney U test were used to compare survivors and non-survivors. A p-value of $p<0.05$ was considered statistically significant.
Results: Throughout the study period, 149 neonates underwent an arterial switch operation (ASO) and 21 TGA patients (62% male) received venoarterial (V-A) ECMO. Median age and weight at time of ECMO cannulation were 7 (2-10) days and 3.5 (3.2-3.9) kg respectively. Morphological diagnosis consisted of TGA with intact ventricular septum in 16/21 (76%), TGA with an inlet ventricular septal defect (VSD) in 1/21 (4.8%) and Taussig-Bing anomaly (double outlet right ventricle and subpulmonary VSD)

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in 4/21 (19%). Eleven neonates (52%) were supported before ASO, because of persistent pulmonary hypertension (10/11, 90%) or hypoxia after pulmonary arterial banding (1/11, 9%) in a patient with late TGA diagnosis. Four neonates (36%) died on/after ECMO prior to the ASO, (55%) were weaned off ECMO prior to the ASO and 1 patient (9%) underwent ASO on ECMO and continued ECMO postoperatively. Postoperative ECMO support was used in 11/21 (52%) patients (1/11, 9% also received ECMO preoperatively), after failure to wean from cardiopulmonary bypass: 6/11 (55%), low cardiac output in ICU: 2/11 (18%), or during cardiopulmonary resuscitation: 3/11 (27%). Median time on ECMO was 71 (IQR: 47-178) hours. Ultimately, 12/21 (57%) patients survived to hospital discharge. There were no statistically significant differences in demographic data (Table 1) or complications between survivors and non-survivors while on ECMO, except for renal failure (p=0.010).
Conclusions: This single-center retrospective study of ECMO in TGA neonates showed a 57% survival to hospital discharge. Renal failure was the only identified risk factor for mortality.

Keywords: Congenital heart disease, Transposition of the Great Arteries, Pediatric Intensive Care, Extra Corporeal Membrane Oxygenation

PP-373
Impact of establishment of pre-operative intensive care unit on outcome of D-transposition of great arteries: an ambispective analysis

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Background and Aim: Transposition of great vessels is an anomaly leading to cyanosis in the early neonatal period, which needs surgical correction at the time of presentation. Due to less available surgical expertise in Low- and middle-income countries, these children get referred for advanced care to Apex hospitals for its definitive management. Survival and long-term outcomes of these children lies on the initial and peri-operative stabilization. Many children present in critically ill state with Cyanosis, respiratory distress, shock and need pre-operative stabilization. Data is scarce on clinical profile and outcome of these critically ill children from LMICs.
Method: Medical records were reviewed retrospectively (Nov 2018–Sept 2021) and prospectively (Oct 2021-Oct, 2023). We assessed impact of a newly commissioned pre-operative PCICU with special Govt. Aid, towards the critical care needs and Peri-operative outcome of these children. A total of 124 cases had been analyzed as three groups [ward setting n=65; intensive care without JSSK, n=14; intensive care with JSSK, n=45] of Demographic and clinical details, critical care needs, pre-operative and post-operative outcomes of children with d TGA were compared amongst these three groups and shown in below table.
Results: Delayed presentation was almost universal, so is the need for intensive care till these children got their corrective surgeries done. The median age of admission had improved from 42 days to 14, and diagnosis from 30 to 14. There was 20 % increase in survival, 14.7 % increase in transfer for surgery, 21.4% drop from being referred back from hospital (LAMA), and reduction of 6.5 hospital stay was observed
Conclusions: This establishment had improved the outcomes in various aspects from survival to reduction in hospital stay

Keywords: Pediatric Cardiac Intensive Care Unit, Critical care needs, Survival outcome

Table

Characteristics		Ward Setting (n=65)	ICU without JSSK (n=14)	ICU with JSSK(n=45)	p-value
Proportion of d TGA		12.1%(N=533)	20.8%(N=67)	15.6 %(N=287)	
Monthly average		2.8	2	2.5	
Gender	Male	43	9	28	
	Female	22`	5	16	0.764#
	Ambiguous			1	
Age (days) [Median (IQR)]		42(7;120)	26.5(9.5;51.75)	14(5.5;30)	
Age of Diagnosis (days)[Median (IQR)]		30(9;93.5)	8.50(4-46)	14(3.50-30)	
Age Groups	First Week	16	3	16	0.107#
	Post 7-days Neonates	14	6	20	
	Post Neonatal Infants	34	4	9	
	Post-infancy	1	1	---	
Pre-operative NIRS, n (%)		16 (45.7%)	5(62.5%)	27(75%)	**0.041#**
Pre-op NIRS duration among survivors (days) [Median (IQR)]		4 (2.5;11.5)	2(1;16.5)	6(3;9)	0.592*
Need of Ventilation, n (%)		13(37.1%)	4(50%)	21(58.3 %)	0.201#
Pre op Ventilation Duration among survivors (days) [Median (IQR)]		8(2;17)	12(4;32)	8(3;15.50)	0.840*
Need of Inotropes, n (%)		10(28.5%)	1(12.5%)	12(33.3%)	0.50#
Inotrope duration among survivors (days) [Median (IQR)]		2(1;5.5)	1(1;1)	6.5(2;9)	0.179*
Maximum VIS [Median (IQR)]		35(20;85)	20(20;20)	40(20;70)	0.631*
Pre-Operative ICU Stay among preop survivors(days) [Median (IQR)]		9 (4; 18)	11(9.25;19.25)	9(6;16.5)	0.70*
Pre-Op ICU Stay - who underwent surgery (days) [Median (IQR)]		14(6;24)	11(9.25;19.25)	9(6;15)	0.54*
Pre-operative Outcome	Discharge n(%)	22(33.8%)	6(42.8)	25(58.1%)	0.98#
	Deaths, n (%)	23(35.3%)	4(28.5)	14(32.5%)	
	LAMA, n (%)	20(30.7%)	4(28.5)	4(9.3%)	
	Transfer for surgery among survivors	25 (71.4%)	8(100%)	31(86.1%)	0.102#
Post-op Outcome among those went for surgery (n=64)	Discharge	12(48%)	6(75%)	20(68.%)	0.306 #
	Death	13(52%)	2(25%)	9(31.1%)	
Total Hospital Stay who got operated and survived (days) [Median (IQR)]		43(30.25;61.75)	37(26;55)	36.5(25.75;50)	**0.05***

Kruskal Wallis test had been applied * , Chi-square test had been applied #

The below mentioned table consists of the observations of the critical care needs of the enrolled

PP-374
Pheochromocytoma induced dilated cardiomyopathy: As rare as treatable

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Background and Aim: Dilated cardiomyopathy (DCM) is often considered a grim diagnosis, with clinicians scrounging for treatable causes with good prognosis. Excessive catecholamines in a case of pheochromocytoma lead to cardiac remodelling, which eventually becomes irreversible. This can manifest as myocarditis or a subtype of cardiomyopathy (Takosubo, hypertrophic or dilated). Improvement in cardiac function may be seen following resection of pheochromocytoma, til several months. We present a case of

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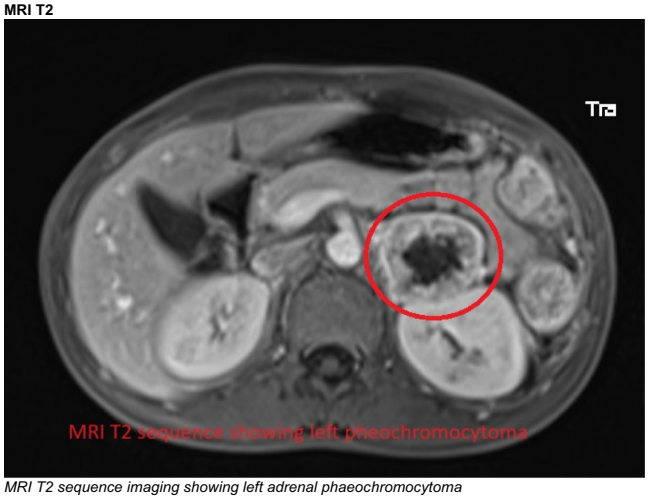
phaeochromocytoma induced DCM who is currently under our follow up. We aim to create awareness about this condition and how timely diagnosis can be beneficial.
Method: We retrospectively reviewed a single case and collected data from records. Review of literature on phaeochromocytoma induced dilated cardiomyopathy was done.
Results: Case Summary A 13-year-old male with a recent diagnosis of DCM, presented to us in cardiogenic shock and multiorgan dysfunction. He was admitted and stabilized in the intensive care unit. ECG revealed sinus tachycardia. 2D Echocardiography showed DCM with severe left ventricular (LV) dysfunction (LV ejection fraction 10-15%), all chambers dilated, grade II mitral regurgitation, moderate tricuspid regurgitation (PG 24mmHg), dilated IVC with no thrombus. An adrenal incidentaloma was detected during work up for heart transplant. Biochemical features and imaging (MRI and GA-DOTANOC PET) were suggestive of pheochromocytoma. He was started on oral alpha-blocker for 7 days, and thence underwent left adrenal excision. His post operative period was uneventful. Pre discharge echocardiography revealed global hypokinesia, with LVEF 20%, mild tricuspid regurgitation, PASP – 26mmHg and dilated LV. Child was discharged on oral diuretics and is currently on follow up.
Conclusions: Due to the atypical presentation of pheochromocytoma as cardiomyopathy, the diagnosis is often delayed. Early resection of the tumour is crucial, potentially leading to cure on follow up. Hence, thorough evaluation of reversible causes in all cases of dilated cardiomyopathy is warranted.

Keywords: Phaeochromocytoma, Dilated Cardiomyopathy



MRI T2 sequence imaging showing left adrenal phaeochromocytoma

PP-376
Severe pulmonary hypertension related to arteriovenous malformation of the vein of galen
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Background and Aim: Arteriovenous malformation of the vein of Galen (AVG) is a rare condition with therapeutic challenges that result in high morbidity and mortality. We report a successful strategic management in a neonate, known fetal diagnosis of AVG, presented with near dismal clinical condition due to persistent pulmonary hypertension of the newborn (PPHN).
Method: The neonate presented with severe hypoxic respiratory failure related to PPHN and congestive heart failure (CHF) within the few hours of life. Despite aggressive respiratory support and PPHN management, including the use of a high-frequency ventilator and inhaled nitric oxide, the newborn's condition deteriorated. Notably, the clinical sign of "reverse differential cyanosis" was observed, prompting an echocardiogram evaluation. The illustration revealed a dilated right atrium, right ventricle, and pulmonary artery, along with patent foramen ovale and large ductus arteriosus, both predominantly right -to-left shunt. Aortic reversal flow across the aortic arch suggested high flow towards the low resistance AVG. Therefore, we opted for urgency endovascular embolization despite unstable clinical condition. The goal was to control the high blood flow shunting across the AVG in order to decrease aortic reversal flow and systemic venous return from AVG to right heart and pulmonary circulation.
Results: Within 24 hours of life, the first endovascular embolization was successfully occluded 60% of the shunt flow across AVG. The clinical condition improved with more stable mild hemodynamics and sign of "differential cyanosis". Intermittent severe desaturation responding to inhaled iloprost was observed. Considering the high flow across the remaining AVG as a contributing condition to PPHN, another embolization was performed on the day 6. All clinical signs of CHF, respiratory failure, and PPHN improved. Subsequently, nitric oxide and ventilator support were weaned and taken off by day 8 and 13.
Conclusions: Understanding of complex cardiovascular hemodynamics, illustrating by echocardiogram, contributes to successful management strategy and outcome of severe CHF, PPHN and hypoxic respiratory failure related to AVG. Urgent endovascular embolization and aggressive medical management improve the prognosis and outcome of AVG and related conditions included severe PPHN.

Keywords: persistent pulmonary hypertension of the newborn, Arteriovenous malformation of the vein of Galen, congestive heart failure, respiratory failure

PP-377
Feeding disorders in pediatric heart transplant patients
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Background and Aim: Feeding is a complex multi-systemic process that is frequently affected in pediatric heart transplant (HT) patients. Feeding disorders can have a medical and developmental impact both short and long term. Little has been published on

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feeding difficulties in this population. Our goal is to describe feeding difficulties, feeding modalities, and the need for feeding therapy during the transplant admission.
Method: Retrospective cohort study of consecutive HT patients at a tertiary center between 09/01/2017 and 09/30/2022. Demographic, and pre-, intra-, and post-HT clinical and feeding therapy data were collected up to HT hospitalization. Data are presented as frequency(%) or median(Inter Quartile Range-IQR).
Results: Of the 66 HT patients, 39 (59%) were male, median age was 10(4, 15) years. Median weight at HT 15 (8,46) kg. Non-cardiac genetic anomalies were encountered in 24 (36%) patients. Median length of stay was 82(29,165) days. Of these patients, 16 (24%) required pre-HT VAD, 8(12%) required VAD and ECMO pre-HT, and 4(6%) required ECMO post-HT. Median time on vent post-HT was 3 days (2,6). Of all HT patients, 60 (91%) had some type of feeding difficulty post-HT. Decreased calorie intake being the most frequent in 53(80%), dysphagia in 24 (36%), and oral aversion in 14 (21%). Feeding modalities are described as 1 day pre-HT, 1-week post-extubation, and at discharge in Table 1. Feeding therapy was implemented in 14 (21%) patients pre-HT and in 24 (36%) post-HT. Of the 18 stroke patients (including pre and post-HT), 16 (88%) had feeding difficulties post-HT and 11(60%) required feeding therapy pre- and/or post-HT. At discharge NGT were present in 7(11%) patients and 1(2%) NJT. G/J-tubes were present in 5 (8%) pre-HT and 9 (14%) at discharge. 28 of the 66 patients required mechanical support (MS) pre/post-HT. When compared to patients that did not require MS, those with MS had enteral feeds 43 vs 29 % at discharge.
Conclusions: Feeding is frequently disrupted due to a variety of reasons both pre- and post-HT. Use of feeding support seems higher in patients who required cardiac MS or had a stroke. Further studies with a greater number of patients might allow to develop predictive models.

Keywords: Pediatric, heart transplant, feeding, feeding disorder, feeding therapy, rehabilitation

Feeding modalities pre and post transplant

Table 1. Feeding modalities used at various periprocedural time points by pediatric patients pre- and post-HT grouped by cardiac mechanical support type(s) used.

Feeding Mode	Cardiac Mechanical Support											
	No ECMO / No VAD			Pre-HT VAD			Pre-HT ECMO & VAD			Post-HT ECMO		
	1 d pre n(%N)	1 w post n(%N)	At DC n(%N)	1 d pre n(%N)	1 w post n(%N)	At DC n(%N)	1 d pre n(%N)	1 w post n(%N)	At DC n(%N)	1 d pre n(%N)	1 w post n(%N)	At DC n(%N)
PO	25 (66%)	21 (55%)	27 (71%)	12 (75%)	6 (38%)	10 (63%)	0	0	2 (25%)	2 (50%)	0	0
NG tube	0	1 (3%)	0	1 (6%)	3 (19%)	1 (6%)	2 (25%)	2 (25%)	1 (13%)	0	0	0
G/GJ tube	3 (8%)	4 (10%)	5 (13%)	0	0	1 (6%)	1 (13%)	0	2 (25%)	1 (25%)	1 (25%)	1 (25%)
NJ tube	1 (3%)	0	0	0	0	0	1 (13%)	3 (38%)	1 (13%)	0	0	0
PN	0	0	0	0	1 (6%)	1 (6%)	2 (25%)	1 (13%)	0	0	0	2 (50%)
PO & NG tube	2 (5%)	4 (10%)	1 (3%)	2 (13%)	3 (19%)	3 (19%)	2 (25%)	2 (25%)	1 (13%)	0	0	0
PO & G/GJ tube	7 (18%)	7 (18%)	5 (13%)	0	0	0	0	0	1 (13%)	1 (25%)	1 (25%)	1 (25%)
PO & NJ tube	0	1 (3%)	0	1 (6%)	2 (13%)	0	0	0	0	0	0	0
Total (N)	38			16			8			4		

Note: ECMO = Extracorporeal membrane oxygenation, VAD = ventricular assist device, 1 d pre = 1 day pre-HT, 1 w post = 1 week post extubation, DC = discharge, PO = per os or by mouth, NG = naso-gastric, G/GJ = gastric/gastric-jejunal, NJ = naso-jejunal, PN = parenteral. The n value represents the total number of patients within each cardiac mechanical support group (i.e., No ECMO / No VAD, Pre-HT VAD, Pre-HT ECMO & VAD, or Post-HT ECMO).

PP-379

The prognostic value of serial risk stratification in adult and pediatric pulmonary arterial hypertension: A systematic review

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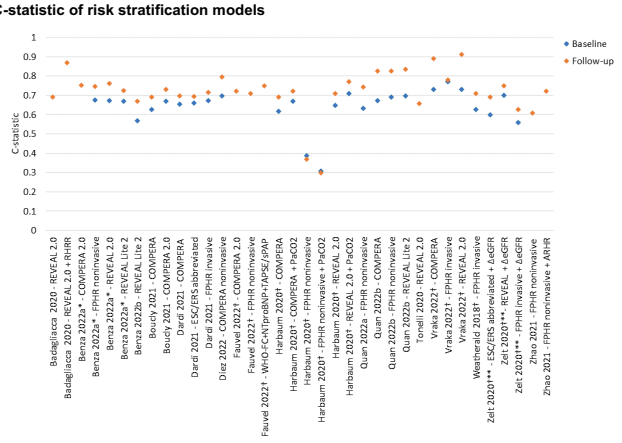
Background and Aim: Risk stratification is recommended to guide the treatment course of pulmonary arterial hypertension (PAH). With risk stratification, patients are stratified into multiple risk categories based on clinical and hemodynamic variables. However, prognostic value of baseline risk stratification has shown to be moderate to good, at best. Serial assessment of risk stratification may improve the prognostic value and can potentially contribute to evaluating treatment response. This systematic review aims to report the prognostic value of serial risk stratification in adult and pediatric PAH and to explore the possibility of serial risk stratification to be used as a potential treatment target.
Method: Electronic databases PubMed, Embase, and Web of Science were searched up to January 30, 2023, using terms associated with PAH, pediatric pulmonary hypertension, and risk stratification. Observational studies and clinical trials describing risk stratification at follow-up or changes in risk score were eligible for inclusion. Case reports, case series, guidelines and reviews were excluded. C-statistic, a measure for the discriminatory ability of a model, was extracted from the included studies, along with the (change in) risk score at baseline and follow-up.
Results: 65 studies were found eligible for inclusion, including 2 studies in a pediatric population. C-statistic range at baseline was 0.31-0.77 and improved to 0.30-0.91 during follow-up (Figure). The average low-risk rate increased from 18% at baseline to 36% during a median follow-up of 244 days (IQR 140-365, n = 40 studies), and in 53% risk status changed (42% improved, 12% worsened) over 168 days (137-327, n = 22 studies). In placebo-controlled drug studies, risk statuses of the intervention groups improved more and worsened less compared to the placebo groups. Furthermore, a low-risk status, but also an improved risk status at follow-up was associated with a better outcome, whereas a worsened risk status was associated with an inferior outcome. Similar results were found in the 2 pediatric studies.
Conclusions: Serial risk stratification has better prognostic value compared to baseline risk stratification and a change in risk status between baseline and follow-up was found to correspond to a change in survival. These data support the use of serial risk stratification as treatment target.

Keywords: Pulmonary arterial hypertension, pediatric pulmonary hypertension, risk stratification, risk assessment, outcome, prognosis

C-statistic of risk stratification models



*C-statistic of the risk stratification models at baseline and follow-up for all-cause mortality, unless stated otherwise. †transplant-free survival, *clinical worsening, **ΔeGFR of the first 6 months after diagnosis added to baseline risk stratification.*

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PP-380

15 years of paediatric cardiac transplantation in hungary

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Gottsegen National Cardiovascular Center

Background and Aim: For children suffering from terminal myocardial failure or congenital heart disease (CHD), in the absence of specific contraindications, transplantation (Tx) is the only curative solution. Aim of this study is to present the experience of the first fifteen years of paediatric cardiac transplantation and its afferent mechanical circulatory support (MCS) program.
Method: Between 2007 and 2022 62 patients underwent transplantation. Median age at transplantation was 9.4 years, with a male/female ratio of 33/29. Mean follow-up was 52 months. MCS was available as a bridging therapy in rapidly worsening cases. The immune induction therapy was based on basiliximab. All diagnostic, procedural and postoperative variables considered relevant, including the time spent on waiting list, and outcomes for both MCS and Tx were retrospectively reviewed.
Results: In 42 cases (67%) the cause was identified as myocardial, while in the remaining 20 it was CHD not amendable to conventional corrective surgery. There were 5 losses (4/5 as MCS complications) among listed patients while awaiting suitable donor. Average waiting was 97 days. From the total 62 transplantations 18 were bridged to Tx by MCS. MCS runs averaged 150 days, 8/18 presenting severe, non-fatal device related complications. Average donor ischemic time was 189 min., for an average op time of 404 min. Three early (≤30 days) graft failures were encountered in 2 patients. Further 3 deaths occurred within the first year (2 infectious, 1 tumour). From the 13 severe late rejections 8 recovered, 5 being unsavable. Overall graft survival during follow-up was 80%.
Conclusions: Paediatric cardiac transplantation offers good long-time survival. Despite shorter waiting times, the rapid course of the disease imposes MCS in a high percentage of the patients, and MCS is mainly responsible for the waiting list mortality. Tumours and rejections are the main contributors for late mortality.

Keywords: transplantation, heart failure, mechanical support

PP-381

Abdominal pain and constitutional symptoms in a recently transplanted pediatric patient: A diagnostic challenge

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Background and Aim: Abdominal pain and constitutional syndrome are concerning symptoms in transplant recipients, requiring post-transplant lymphoproliferative disorder (PTLD) to be ruled out. We present the case of a patient who developed these symptoms in the early post-transplant period making the differential diagnosis challenging.
Method: Clinical case report.
Results: a 12-year-old patient received a heart transplant due to severe hypertrophic cardiomyopathy caused by MYH7 mutation. Postoperative course was uneventful except for asymptomatic moderate cellular rejection at day +11 in relation with failure to achieve target tacrolimus levels despite increasing doses, resolving with corticosteroids boluses. Pharmacogenetic test confirmed fast metabolism of tacrolimus, so a prolonged-release formulation was prescribed. One month after discharge she presented to the ED with fever associated to asthenia and abdominal pain during the previous week. Lab tests showed severe neutropenia and CRP elevation. She was admitted with antibiotics, requiring G-CSF to normalize neutrophil count despite previous suspension of potentially causing drugs (mycophenolate mofetil [MMF] and valganciclovir). An abdominal ultrasound demonstrated thickening of the ascending colon not compatible with typhlitis, thus PET-CT and biopsy were scheduled to rule out PTLD, both showing nonspecific inflammatory changes. Within a few days she developed diarrhea with stool culture positive for C. difficile, receiving treatment with oral vancomycin, after which abdominal pain and fever disappeared, being discharged with reintroduction of MMF. Two months later she complained again of asthenia, abdominal pain and intermittent diarrhea with weight loss of 3 kilograms. EBV blood load was repeatedly negative. New abdominal ultrasound and PET-CT showed persistence of right colon thickening with moderate metabolic activity and pathological lymph nodes. A new colonoscopy demonstrated moderate ulcerative colitis and lymphoid hyperplasia, with anatomopathological diagnosis of apoptotic colopathy with immunophenotype not compatible with lymphoproliferative syndrome, suggestive of pharmacological colitis. In view of these findings, MMF was suspended, and tacrolimus dose was reduced, with progressive resolution of symptoms.
Conclusions: MMF gastrointestinal side effects are well known, being abdominal pain and diarrhea that disappear after suspension the most common presentations. However, some patients can develop severe apoptotic colopathy with a clinical course mimicking PTLD, requiring high index of suspicion and anatomopathological confirmation.

Keywords: Heart Transplant, PTLD, Mycophenolate mofetil, Apoptotic colopathy

PP-382

Digital subtraction angiography in pediatric pulmonary hypertension assessment: Safety and feasibility

Dijana Popevski, Joseph Andreola, Steven Bibevski, Thomas Forbes and Kak Chen Chan
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Background and Aim: Cardiac catheterization remains the gold standard for diagnosis of pediatric pulmonary hypertension (PPH)1. Pulmonary arterial (PA) angiography may precipitate a crisis causing hemodynamic instability and even death. Digital subtraction angiography (DSA) is an established technique using computational manipulation to enhance its imaging capability2. Its use in pediatric cardiology has been limited despite advantages of lower dose of radiographic contrast3. The aim of this study is to review our experience using DSA during evaluation of PPH at cardiac catheterization.
Method: We performed a retrospective review of patients with PPH who underwent catheterization evaluation with DSA from 2018 to 2023. All procedures were performed using general anesthesia by a single operator (KCC). All DSA were performed after baseline hemodynamic study and whilst on 100% oxygen and iNO. The DSA were power injections acquired at 15 frames per second using half strength contrast at standard volume and injection rates. Data collected were age, weight, contrast volume,

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radiation dose and adequacy of DSA for analysis. All the angiograms were reviewed by DP and KCC.
Results: The patient data are summarized in the Table 1. There were no major complications related to the cardiac catheterization. There were no pulmonary hypertensive crises precipitated by the DSA. All PA angiograms were of adequate quality for analysis (Fig. 1a). Abnormalities in the pulmonary artery were identified (Fig. 1b).
Conclusions: DSA is a safe and effective diagnostic technique that can be used in PPH patients. The advantage of DSA includes lower contrast load and potentially better image quality with delineation of the pulmonary vasculature for analysis of perfusion of the pulmonary vascular bed. The main disadvantage of this technique is the need of controlled apnea during the acquisition of the images and the need for general anesthesia.
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2. Tonkin, Ina L. M.D. Imaging of Pediatric Congenital Heart Disease. Journal of Thoracic Imaging 2000; 15:274-279.
3. Dickinson DF, Wilson N, Partridge JB Digital subtraction angiography in infants and children with congenital heart disease. Heart. 1984;51:485-491.

Keywords: pulmonary hypertension, digital subtraction angiography

Fig 1a, Fig 1b, Table 1

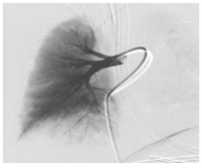

Fig 1a

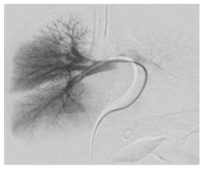

Fig 1b

Table 1.

Age range	Number of patients	Average of BSA (m2)	Average of Radiation time (minutes)	Average of Radiation dose (mGy)	Average of Radiation DAP (uGy*m2)	Average of Volume of contrast (ml)
0-1 years	16	0.25	21	117	1,684	8
1-5 years	28	0.52	22	164	3,750	16
5-10 years	4	0.78	41	191	2,492	27
10-15 years	7	1.16	8	117	2,144	24
15-18 years	3	1.75	15	334	8,378	26
>18 years	2	1.66	22	290	7,650	15
Grand Total	**60**	**0.64**	**21**	**161**	**3,289**	**16**

BSA: body surface area; DAP: dose area product

Patient data

PP-383
Digital subtraction angiography in assessment of pediatric pulmonary hypertension: Pattern of pulmonary vasculature abnormalities

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Background and Aim: Pulmonary hypertension comprises a variety of etiologies spread across the entire age spectrum from newborn to late adolescence. Cardiac catheterization remains the gold standard for diagnosis of pediatric pulmonary hypertension (PPH). Digital subtraction angiography (DSA) is increasingly being used during cardiac catheterization for PPH in our center. Potential benefits of DSA include lower dose of contrast media, decreased risk for precipitation of pulmonary hypertensive crisis whilst providing adequate delineation of the pulmonary vascular bed anatomy. The aim of this study is to review the ability of DSA in defining the patterns of abnormalities of the pulmonary vascular bed in patients with PPH.
Method: We performed a retrospective review of patients with PPH who underwent cardiac catheterization and DSA between 2018 and 2023. All the procedures were performed under general anesthesia by a single operator (KCC). All DSA were performed after baseline hemodynamic study and whilst on 100% oxygen and iNO. The DSA were power injections acquired at 15 frames per second using half strength contrast at standard volume and injection rates. All the angiograms were reviewed and categorized by DP and KCC. The pulmonary vascular abnormalities were categorized by Group 1. Gross anatomical abnormality (absent, hypoplastic, stenosis, venous anomaly – Fig. 1a), Group 2. Perfusion distribution abnormality (lobar, segmental, patchy – Fig. 1b hypoperfusion), Group 3. Arborization abnormality (decreased arborization "Tree-in-Winter", decreased longitudinal perfusion – Fig. 1c "pruning").
Results: DSA were adequate for analysis in 56 patients. DSA provided adequate imaging of the pulmonary vascular bed for delineation of abnormalities associated with PPH. There were 17 patients in Group 1, 30 patients in Group 2 and 40 patients in Group 3. (Table 1).
Conclusions: DSA provides adequate delineation of the anatomy of the pulmonary vascular bed in PPH. DSA provides good information on the state of the pulmonary perfusion and venous anomalies on recirculation. However, the morphological variations demonstrated by DSA must be correlated with the pulmonary

Abnormalities associated with PPH

Fig 1a. – Scimitar vein

Fig 1b – Patchy hypoperfusion and decreased arborization.

Fig 1c - Pruning

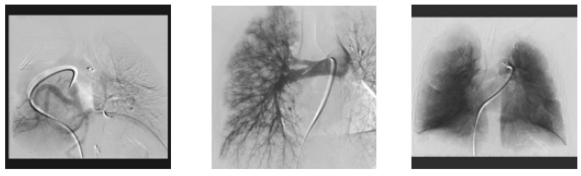

Table 1

Group 1	**17**
Unilateral PA abnormalities	7
Absent branch PA	2
Branch PA stenosis	3
Pul. Vein abnormalities	5
Group 2	**30**
Decreased vascularity	14
Pruning	16
Group 3	**40**
Lobar hypoperfusion	4
Segmental hypoperfusion	15
Patchy hypoperfusion	21

The pulmonary vascular abnormalities were categorized by Group 1. Gross anatomical abnormality (absent, hypoplastic, stenosis, venous anomaly – Fig. 1a), Group 2. Perfusion distribution abnormality (lobar, segmental, patchy – Fig. 1b hypoperfusion), Group 3. Arborization abnormality (decreased arborization "Tree-in-Winter", decreased longitudinal perfusion – Fig. 1c "pruning").

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hemodynamics. Its use in assessment of patients with PPH provides an additional avenue to further assess these high risk patients.

Keywords: pulmonary hypertension, digital subtraction angiography

PP-384

Percutaneous atrial septal defect closure in patients with pulmonary arterial hypertension

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Background and Aim: Pulmonary arterial hypertension (PAH) is a dreaded, although rare, complication of uncorrected atrial septal defect (ASD). Therapeutic strategies when PAH is established are controversial. 2022 ESC/ERS guidelines stipulate that ASD closure should be considered only when PVR is below 5 Wood units (WU). However, transcatheter shunt closure beyond this threshold seems to be feasible in selected patients. We aimed to assess outcomes after transcatheter ASD closure in patients with PAH.
Method: This study is based on a cohort of 2,047 consecutive patients (27 years [11;49] and 68,2% female) who underwent percutaneous ASD closure (ASOlong registry, IDRCB 2019-A02691-56) between May 1998 and June 2020. A sub-group of 20 patients with PAH was identified (1.0% of the ASOlong registry, 17/20 (85%) females). Right-heart catheterization (RHC) was performed in hemodynamically stable and spontaneously breathing patients, allowing direct measurement of oxygen consumption. Pulmonary output (Qp) and systemic output (Qs) were calculated using the Fick principle.
Results: Median age at PAH diagnosis was 39 years [23-48] and 41 [34-51] at the time of ASD closure. The majority (70%) of patients had recent onset of symptoms (< 24 months). 55% patients were in NYHA III-IV. The "treat-and-repair strategy" was proposed in six patients. Transcatheter ASD closure were performed with the non-fenestrated Amplatzer Septal Occluder device. No patient had immediate PAH deterioration after closure. Median follow-up time was 5.9 years [2.3- 9.9]. At last follow-up, mPAP remained significantly lower compared to baseline (Δ mPAP -16 mmHg [-10; -21)]; P= 0.02), as well as PVR (Δ PVR -2.3 WU [- 1.2; - 4.2]; P= 0.03). In patients who underwent the "treat and repair strategy", mPAP decreased 5.5 mmHg [-1 to -14] as well as PVR -1.5 WU [-0.2 to 1.9]). 95% patients were in NYHA I-II without reported PAH deterioration. Ten patients had persistent mild to moderate PAH. All of them were identified as "low-risk" according to the 2022 ESC/ERS guidelines four-strata risk stratification.
Conclusions: ASD closure may be an interesting treatment strategy with sustained clinical and haemodynamic improvements and low morbidity in selected patients with moderate to severe PAH (PVR < 10 WU) and persistent significant left-to-right shunt.

Keywords: Pulmonary hypertension, congenital heart disease, atrial septal defect, transcatheter closure

PP-385

Morphology and rheology of pulmonary arteries: a relationship with right ventricular function in patients with pulmonary arterial hypertension associa

Estibaliz Valdeolmillos[1], Hichem Sakhi[2], Paul Vignaud[1], Marion Audie[1], Marc Antoine Isorni[3], Emmanuelle Fournier[1], Lisa Guirgis[1], Olivier Sitbon[4], David Montani[4], Xavier Jais[4], Laurent Savale[4], Marc Humbert[4], Arshid Azarine[3] and Sebastien Hascoet[1]
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Background and Aim: The relationship between right ventricular (RV) function and anatomical and rheological modification of the proximal pulmonary artery (PA) remains unknown in patients with pulmonary arterial hypertension (PAH) associated with atrial septal defect (ASD).
Method: This study sought to evaluate the intrinsic properties of PA and its relationship with RV function and PA dilatation in patients with PAH associated with ASD. A multiparametric evaluation, including complete heart catheterization and a 4DFlow-CMR, was performed in 24 patients with PAH associated with ASD.
Results: The study population has highly impaired intrinsic AP properties. RV ejection fraction (RVEF) does not correlate with AP area but correlates with VD-AP coupling (correlation coefficient=-0.91, p<0.000), right PA compliance (correlation coefficient=-0.40, p=0.0470), and right PA stiffness index (correlation coefficient=0.43, p=0.0329). Intrinsic parameters were more severe in the group with RV function preserved (RVEF > 45%), characterized by a persistent left to right shunt with a decreased right PA compliance (p= 0.0164), decreased distensibility (p= 0.0164), decreased pulsatility index (p= 0.0695), increased β-stiffness index (p= 0.0139), and increased elastance (p= 0.0164).
Conclusions: In patients with PAH associated with ASD, proximal PA are highly remodeled with increased dilatation and stiffness. The remodeling seems to depend on the barometric overload but also on the volumetric overload by the left-right shunt through the ASD. ASD appears to impact the relationship between RV function and proximal PA.

Keywords: Pulmonary arterial hypertension, congenital heart disease, atrial septal defect, cardiac magnetic resonance.

PP-386

Pediatric tolvaptan use in heart failure: A retrospective analysis of hyponatremia correction and clinical outcomes in two cases

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Background and Aim: Hyponatremia is a common complication in heart failure patients undergoing diuretic therapy, primarily due to the inhibition of sodium reabsorption in the kidneys by commonly used diuretics. Tolvaptan, an oral vasopressin V2-receptor

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antagonist, has shown promise in ameliorating congestion and elevating serum sodium levels in adult patients with heart failure patients and hyponatremia. Since there have been reports on the effectiveness and safety of Tolvaptan in pediatric patients, our study aims to analyze two cases of Tolvaptan use in pediatric patients with heart failure and hyponatremia.
Method: We retrospectively identified two pediatric patients (3 and 10 years old) with heart failure and hyponatremia who were prescribed Tolvaptan at Children's Health Memorial Hospital in Poland between January and September 2023. Baseline and post-7-day therapy data, including laboratory parameters, urine output, and body weight, were recorded.
Results: Initially, both patients were on concurrent loop diuretics (furosemide), thiazide diuretics (hydrochlorothiazide), and oral sodium supplementation. The first patient (restrictive cardiomyopathy) received an initial dose of 0.375 mg/kg, resulting in increased diuresis and thirst. The dose was later adjusted to 0.25 mg/kg every other day. Seven days after the first tolvaptan dose, serum sodium levels normalized, and body weight decreased. However, after three weeks, weight gain and pericardial effusion occurred. Tolvaptan was then discontinued, and diuretic doses increased, with no subsequent decrease in serum sodium levels.
The second patient (hypoplastic left heart syndrome, post-Glenn operation) received a daily dose of 0.3 mg/kg. After seven days, serum sodium levels normalized, there was an increased diuresis and a decrease in body weight. However, due to elevated transaminase levels, the medication was stopped. As heart failure signs worsened, the patient was admitted to the Intensive Care Unit. In both cases, tolvaptan administration facilitated a reduction in diuretic doses and oral sodium supplementation.
Conclusions: Our findings suggest that Tolvaptan can effectively correct hyponatremia in pediatric patients with heart failure. The improvements in serum sodium levels and urine output indicate the potential utility of Tolvaptan in managing hyponatremia in this population. Further research is needed to assess its safety and efficacy in a larger pediatric heart failure population.

Keywords: tolvaptan, hyponatremia, heart failure, diuretics, urine output

PP-387
The assessment of novel biomarker – galectin-3 in children with congenital heart defects and heart failure in preoperative period

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Background and Aim: Congenital heart diseases (CHD) are the most common cause of heart failure (HF) in children. In clinical assessment of HF application of biomarkers and scales is useful. Galectin-3 is biomarker with proved utility in adults and little research in children. The aim was to analyze galectin-3 in children with CHD according to HF progression in preoperative period.
Method: Study included 41 children with CHD listed for cardiac surgery (aged 1 day to 15 months) and 27 controls. In both groups a physical examination was performed, medical history was taken, and galectin-3 was determined. In study group clinical HF scales (NYHA-Ross, NYHA-Ross adapted by Laer and Reithmann, NYU PHFI) were used, laboratory tests (NTproBNP and hemoglobin) and diagnostic studies (echocardiography, chest x-ray) were performed.
Results: Galectin-3 in study (11,83ng/mL) and control (11,67 ng/mL) group was not significantly different and didn't correlate with variables (age, sex and weight). There was no difference in galectin-3 between patients with HF compared to those without or those from control group. Galectin-3 was not a significant predictor of HF intensity in any scale. Statistical trend was observed in Ross-Reithmann scale($p=0,058$). Highest median galectin-3 (16,38 ng/ml) was in class IV of NYHA-Ross scale. In statistical model with NTproBNP as second predictor a trend was observed ($p=0,094$) with galectin-3 in relation to Ross-Laer scale and both galectin-3 and NTproBNP were significant predictors of Ross-Reithmann scale result ($p<0,05$). Galectin-3 was lower in children treated with propranolol or spironolactone (Figure 1) and higher in those taking captopril($p<0,05$). There was no correlation with furosemide. There was no relationship between galectin-3 and increased pulmonary blood flow or enlarged heart on chest x-ray, LV systolic function and NTproBNP or hemoglobin concentration.
Conclusions: Galectin-3 in children with CHD before surgery does not differ significantly from healthy children and is independent of sex, age and weight. Galectin-3 is lower in children with CHD treated with spironolactone or propranolol and higher in treated with captopril. Galectin-3 is not related to hemodynamic disturbances in chest x-ray or echo. Galectin-3 does not differentiate HF intensity in children with CHD assessed in HF scales.

Keywords: Galectin-3, heart failure, congenital heart diseases

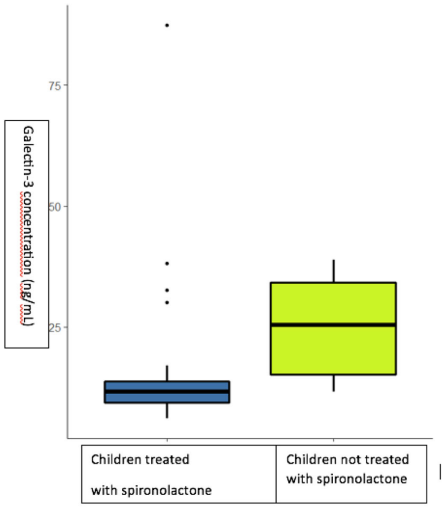

Figure 1. The analysis of galectin-3 (ng/mL) concentration in children with HF between those treated and not treated with spironolactone.

Cardiac Dysrhythmias and Electrophysiology

PP-388
Analysis of novel biomarker– galectin-3 in relation to postoperative arrhythmia and arrhythmia risk factors in children with congenital heart defects

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Background and Aim: Cardiosurgery is crucial phase of congenital heart disease (CHD) treatment in children. Arrhythmia is important death risk factor in postoperative period with most severe form

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being junctional ectopic tachycardia. However, there are known risk factors of postoperative arrhythmia (i.e Aristotle Basic Score (ABS) for congenital heart surgery, cardiopulmonary bypass time (CBT), aortic crossclamp time (ACT), deep hypothermia time (DHT), intensive care unit (ICU) stay and mechanical ventilation time(MVT)), no biomarker being related was found. Galectin-3 has proved utility as risk factor in adults with heart failure or arrhythmia. The aim was to assess galectin-3 in children with CHD undergoing cardiosurgery according to arrhythmia and arrhythmia risk factors in postoperative period.
Method: Study group was 33 children with CHD listed for cardiac surgery (aged 1 day to 15 months). Control group was 27 healthy children. In both groups galectin-3 was determined. In the study group electrocardiography and 48-hours Holter monitoring were performed. Arrhythmia risk factors were assessed: ABS for congenital heart surgery, CBT, ACT, DHT, ICU stay and MVT.
Results: Galectin-3 in the study group in the postoperative period was 15,87 ng/mL and was statistically significantly higher than in the preoperative period or in the control group ($p < 0,05$). Galectin-3 in the pre- and postoperative period, was not significant predictor of any type of arrhythmia. Galectin-3 in the pre- and postoperative period in children affected with JET was not statistically significantly different than in those without JET. The analysis revealed that preoperative galectin-3 concentration is statistically significant predictor of cardiopulmonary bypass time and aortic crossclamp time ($p<0,05$) – Figure 1 and 2. Galectin-3 in postoperative period and arrhythmia risk factors showed no statistically significant results ($p>0,05$).
Conclusions: Galectin-3 in the preoperative period is higher in children with CHD in whom cardiac surgery is related with longer cardiopulmonary bypass time and aortic crossclamp time. Galectin-3 increases in children after cardiosurgery for CHD. Galectin-3 does not differ in children with arrhythmia in the post-cardiac surgery period.

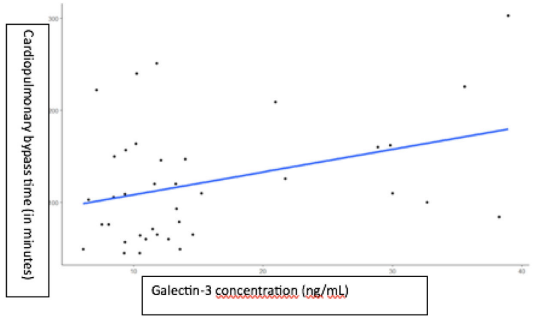

Figure 1. The relation of preoperative galectin-3 (ng/mL) serum concentration to cardiopulmonary bypass time.

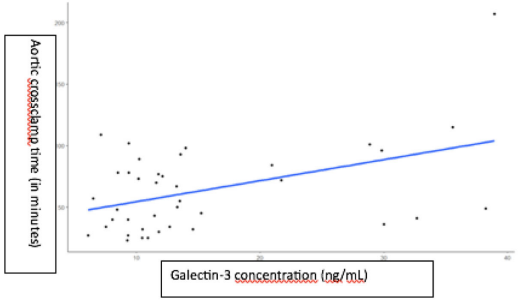

Figure 2. The relation of preoperative galectin-3 (ng/mL) serum concentration to aortic crossclamp time.

Keywords: Galectin-3, arrhythmia, congenital heart diseases

Pulmonary hypertension, heart failure and transplantation

PP-389
Bilateral lung transplantation for pediatric pulmonary arterial hypertension: Perioperative management and one-year follow-up

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Background and Aim: Bilateral lung transplantation (LuTx) remains the only established treatment for children with end-stage pulmonary arterial hypertension (PAH). Although PAH is the second most common indication for LuTx, little is known about optimal perioperative management and mid- to long-term clinical outcomes.
Method: Prospective observational study on consecutive children with PAH who underwent LuTx with scheduled postoperative VA-ECMO support at Hannover Medical School from December 2013 to June 2020.
Results: Twelve patients with PAH underwent LuTx (mean age 11.9 years; age range 1.9–17.8). Underlying diagnoses included idiopathic (n=4) or heritable PAH (n=4), PAH associated with congenital heart disease (n=2), pulmonary veno-occlusive disease (n=1), and pulmonary capillary hemangiomatosis (n=1). The mean waiting time was 58.5 days (range 1–220d). Three patients required emergency VA-ECMO cannulation pre-LuTx because of acute right heart failure/pulmonary vascular crisis, and were bridged to LuTx on awake VA-ECMO. Intraoperative VA-ECMO/cardiopulmonary bypass was applied and VA-ECMO was continued postoperatively in all patients to assist the pressure-unloaded RV and volume-loaded LV. The mean duration of post-LuTx VA-ECMO support was 185 h (range 73–363 h). All patients were extubated whilst on ECMO support (awake VA-ECMO) to avoid pressure and shear stress on the transplanted lungs. The median postoperative ventilation time was 28h (range 17–145h). Transthoracic echocardiography 12 months after LuTx showed full and sustained recovery of RV systolic function in all 12 children after bilateral LuTx by means of echocardiographic conventional and strain analysis. All PAH patients are alive two years after LuTx (median follow-up 53 months, range 26–104 months). Three-year outcome data are currently under analysis.
Conclusions: LuTx in children with end-stage PAH resulted in excellent midterm outcomes (100% survival two years

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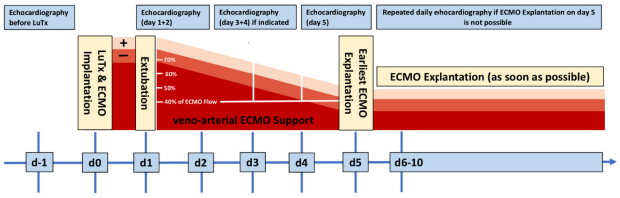
post-LuTx). Postoperative VA-ECMO facilitates early extubation with rapid gain of allograft function and sustained biventricular reverse-remodeling and systolic function after RV pressure unloading and LV volume loading.

Keywords: lung transplantation, pediatric pulmonary arterial hypertension, extracorporeal membrane oxygenation (ECMO), awake ECMO

Schematic treatment and weaning algorithm of VA-ECMO treatment after LuTx



All patients were treated according to this interdisciplinary, in-house consensus standard. Abbreviations: ECMO, extracorporeal membrane oxygenation; d, day; LuTx, lung transplantation.

PP-390

The pulmonary artery banding model of rv pressure load: hemodynamic and deep cardiovascular phenotyping reveals complex rna-signatures in rv failure

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Background and Aim: We previously reported RNA regulatory networks and expression of genes specific to RV hypertrophy (RVH) in the non-failing human heart (tetralogy of Fallot vs. VSD; (DOI: 10.1016/j.isci.2021.102232). The aim of this study was the comprehensive characterization of both hemodynamics and regulatory RNA networks in RV pressure overload heart failure in pulmonary artery-banded (PAB) rats.

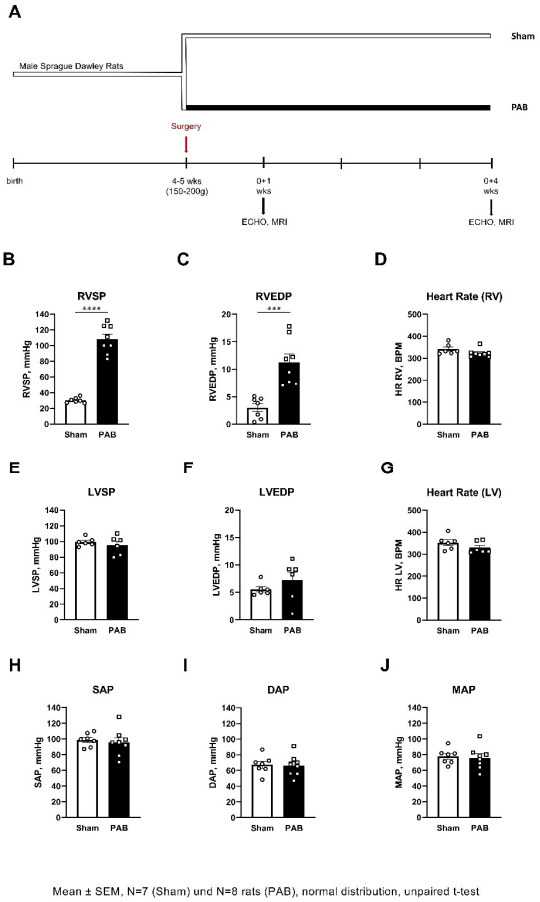
Method: 4-5 weeks old ventilated male SD rats (body weight 150-200g) underwent surgical clipping (0.6mm; DOI: doi:10.3791/58050) of the PA trunk (PAB; n=8), or Sham procedure (no clip; n=7), followed by suture and extubation. Four weeks after surgery, rats underwent echocardiography (echo), cardiac MRI and right-left heart catheterization (closed-chest, spontaneously breathing), followed by heart tissue harvest. We conducted ventricle-specific RNA-Seq of circ-, lnc-, mi- and mRNA to establish regulatory RNA-networks, as previously developed by our group (DOI: 10.1016/j.xpro.2021.100769).

Results: PA CW-Doppler velocities were 3.5-5m/s in PAB vs. <2m/s in Sham rats, indicating proper PA-banding. PAB vs. Sham rats had greatly elevated RVSP (83 vs. 27 mmHg; p<0.0001) and RVEDP (11 vs. 5 mmHg; p<0.001), while LV and aortic pressures were similar (Figure 1). Echo and MRI data are pending; qualitative imaging and cardiac catheterization indicate RV systolic and diastolic dysfunction, hypertrophy and dilation in PAB vs. Sham rats. RNA-Seq. of competing endogenous (circ, lnc) RNA, miRNA and mRNA unraveled strong cardiac induction of pathways associated with apoptosis, DNA damage, angiogenesis, reduced fatty acid oxidation, increased glycolysis, hypoxia, reactive oxygen species, and inflammation in PAB vs Sham rats. The strongest pathological regulation occurred in the septal hinge points, followed by the RV anterior and LV posterior walls. Indeed, we found upregulated pro-cardiofibrotic miRNAs (hinge points: miR-132, miR-21, miR-34a; RV: miR-132, miR-21; LV: miR-101a, miR-132) and upregulated heart-failure-associated miRNAs (hinge points: miR-18a, miR-221, miR-223, miR-652; RV: miR-18a, miR-221; LV: miR-301a).

Conclusions: This is the most comprehensive, multi-modal characterization of the PAB rat model of RV pressure load, using a new protocol (body weight, PAB-duration). Such clip-induced PA trunk-banding produces reliable results with low variance. RNA-Seq analysis demonstrates complex regulatory networks in RV pressure overload heart failure affecting cardiac glucose/lipid metabolism, inflammation, fibrosis, and hypertrophy in the RV and septal hinge points.

Keywords: Pulmonary artery banding, RNA regulatory networks, RV failure, septal hinge point fibrosis, heart failure

Figure 1



Mean ± SEM, N=7 (Sham) und N=8 rats (PAB), normal distribution, unpaired t-test

Experimental setup and catheterization hemodynamics

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PP-391

Single centre experience on the use of rivaroxaban as thromboprophylaxis in paediatric patients with dilated cardiomyopathy

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Background and Aim: Children with dilated cardiomyopathy (DCM) are at risk of thromboembolism, affecting mortality and morbidity. Paediatric guidelines recommend ongoing systemic anticoagulation in the presence of arrhythmias or in the presence of previous thromboembolism, thrombophilic conditions or ejection fraction (EF) <25%. Direct Oral Anticoagulants (DOACs) including rivaroxaban, eliminate monitoring limitations associated with conventional therapies such as heparin and warfarin and are preferred for thromboembolism prevention in adults and children. Published literature on rivaroxaban in children demonstrates a favourable reduction in venous thromboembolism (VTE) events, and embolisms in congenital heart disease. Here we report our centre's experience using rivaroxaban in children with DCM.
Method: Retrospective case note review of children with DCM commenced on rivaroxaban between June-August 2023 conducted at a tertiary paediatric cardiac centre. Children were identified from pharmacy records and relevant clinical data collated from patient records including echocardiograms, renal and liver function, full blood count, iron studies and patient reported adverse effects including bleeding events.
Results: Five children median age 9 years (range 5 months–14 years) with DCM and an initial median EF of 21% (range 12-23%) were commenced on rivaroxaban for prevention of intracardiac thrombus. Dosing for four patients followed the UNIVERSE study; one patient initiated on dosing for VTE prophylaxis as per product licensing following a resolved intracardiac thrombus. No evidence of intracardiac thrombus was seen for any patient during the 16-week study period. Rivaroxaban continued throughout for four patients as EF remained consistently <30%. Rivaroxaban was switched to heparin after 6 weeks for one child due to worsening left ventricular dysfunction who subsequently died due to this. Platelets, liver and renal function remained in range; demonstrating nil adverse effects. Three patients had iron deficiency anaemia; a common finding in heart failure and were on supplementation prior to starting rivaroxaban. No adverse effects were reported by patients.
Conclusions: In a small selection of patients with DCM and poor function at our centre, rivaroxaban was well tolerated and effective with no development of intracardiac thrombus or side effects. However, further studies in a larger study population are required before this can be recommended routinely.

Keywords: Dilated cardiomyopathy, Heart failure, Paediatric, Rivaroxaban, Anticoagulation

PP-393

Septal flattening in the absence of a shunt is a poor indicator of early pulmonary hypertension in preterm infants

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Background and Aim: Diagnosing early pulmonary hypertension (PH) in preterm infants is challenging as invasive validation with cardiac catheterization is not possible nor desirable. Especially in the absence of shunts, septal flattening is one of the echocardiographic parameters commonly used to identify PH, although it's sensitivity and specificity for diagnosing early PH in these infants is debated. We aimed to assess the association between septal flattening and tricuspid regurgitation (TR) as indicators of early PH in preterm infants in the absence of a shunt.
Method: We included preterm infants born with a gestational age<30 weeks and/or a birth weight<1000 grams born between October 2015 and June 2020. Infants either underwent echocardiographic screening for PH on day 3-10 after birth as part of a prospective cohort study or on clinical grounds. We defined PH as the presence of at least one of the following: - A bidirectional or right-to-left shunt. Phenotype:"persistent pulmonary hypertension of the newborn (PPHN)"; - A left-to-right shunt through the PDA with low velocity (Vmax<2m/s or Vmax< 3m/s with septal flattening). Phenotype:"flow-PH" - In the absence of shunts: Any degree of septal flattening and/or a TR>2.8 m/s or TR/systolic blood pressure (SBP)>0.5. Phenotype:"PH without shunt"
Results: Out of 194 included infants, 101 infants (52%) had early PH; 25 PPHN (13%), 60 flow-PH (31%) and 16 PH without shunt (8%). PH without shunt was diagnosed based on septal flattening only in all 16 infants. In these 16 infants, reliable TR measurements co-existed in 6 and none were above the pre-defined threshhold of 2.8 m/s, whereas 2/2 (100%) of reliable TR measurements in the PPHN-cohort and 7/9 (78%) of reliable TR measurements In the flow-PH cohort were > 2.8 m/s or > 0.5 of SBP.
Conclusions: In the absence of a shunt, we found a poor association between septal flattening and TR at echocardiography as indicators of early pulmonary hypertension in preterm infants in this combined prospective and retrospective study. These observations illustrate the difficulty of diagnosing early PH in preterm infants in the absence of a shunt. An integrative approach using multiple echocardiographic measurements may be needed in these patients.

Keywords: Pulmonary hypertension, persistent pulmonary hypertension of the newborn, preterm infants

PP-394

Physiological-based cord clamping and early oxygenation in infants with TGA using a new purpose-built resuscitation table: A feasibility study

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Background and Aim: Infants with Transposition of the Great Arteries (TGA), especially with intact ventricular septum (TGA-IVS), are highly susceptible for developing pulmonary hypertension (PPHN) after birth. Physiologically based cord clamping (cord

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clamping until after lung aeration has been established; PBCC), proven beneficial in term infants, leads to a more stable cardiovascular adaptation with improved oxygenation. It could therefore decrease the risk for PPHN in TGA-newborns. We assessed the feasibility of combining PBCC while administering extra oxygen immediately after birth.
Method: The stabilisation approach was performed at the Leiden University Medical Center from January – September 2023. Newborns with TGA were stabilised on a new purpose-built resuscitation table (Concord; Figure 1), provided with standard stabilisation equipment. All newborns received supplemental oxygen (initially 2L High Flow with fraction of inspired oxygen (FiO2) of 1.0). Cord clamping was performed when the infant was stable (Heart rate (HR) >100 bpm along with spontaneous breathing while receiving oxygen therapy) with minimum of 3 and maximum of 10 minutes until cord clamping. Continuous HR, oxygen saturation (SpO2), 5 and 10-minute Apgar scores and temperature at admission were determined.
Results: So far, eight infants (75% male) were included, of which 3/8 (37%) had ventricular septal defects (VSD). Vaginal delivery occurred in 6/8 (75%), with median (IQR) gestational age of 39 (37-40) weeks and birth weight of 3.3 (2.8-3.5) kg. PBCC was successful in all 8 infants with cord clamping time of 9.2 (3.8-10.5) min. There were no maternal or neonatal adverse events during stabilisation. Preductal SpO2 values were 60% (51-69%), 66% (42-82%) and 79% (76-88%) at 3, 5 and 10 minutes after birth. HR levels were 157 (117-168), 164 (137-180) and 156 (135-170) bpm, respectively. Following initial stabilisation, all vaginally born term infants were able to have skin-to-skin contact with their mother for at least 10 minutes with continuous monitoring and supplemental oxygen. Two infants (25%) required NO-therapy for pulmonary hypertension within the first 24 hours after birth.
Conclusions: Combining early oxygenation with PBCC using the Concord in TGA-newborns is feasible. No adverse events were observed during stabilisation; HR remained stable, and SpO2 increased gradually with early oxygen supply.

Keywords: Congenital heart disease, Transposition of the Great Arteries, Physiological-based cord clamping

PP-395
Does ECHO assessment correlate with invasive haemodynamics in paediatric restrictive cardiomyopathy?

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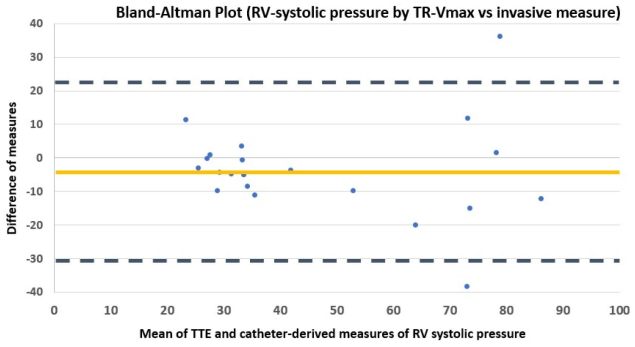
Background and Aim: Restrictive physiology in primary cardiomyopathy presents a significant challenge to monitor paediatric patients. Longitudinal assessment of left ventricular (LV) diastolic function and right heart pressures is key to offering our patients optimal treatment, prognostication, and timing transplant listing. The gold standard is use of invasive haemodynamics, performed at regular intervals. Transthoracic echocardiography (TTE) however is far more accessible, presents no anaesthetic risk and is routinely performed in the outpatient setting. Whilst TTE assessment is well validated in primary pulmonary hypertension, only a single small study evidences its use in restrictive disease.
Method: This single-centre, retrospective study aims to review data from all those undergoing cardiac catheterisation, between 2019-2023, for either primary restrictive or hypertrophic cardiomyopathy with restrictive physiology. Comparison is made between these data and concurrent TTE assessment. Right heart pressures are compared to tricuspid regurgitation maximal velocity (TR-Vmax) and pulmonary regurgitation end-diastolic velocity (PR-EDV). LV end-diastolic pressure (LV-EDP) or pulmonary capillary wedge pressures (PCWP) are compared to atrial area, inflow dopplers and tissue doppler parameters.
Results: Thirty-eight diagnostic catheter studies were undertaken in 24 patients during the study period. Eighteen utilised MRI flow data. TR-Vmax was measurable with good doppler trace on 71% of occasions, with strong correlation between ECHO-derived and invasive estimates of systolic PAP ($r=0.82$, $p<0.001$). No measurement of TR-Vmax <3.0m/s correlated with transpulmonary gradient above 10mmHg or indexed pulmonary vascular resistance <3.0WU.m^2. TR-Vmax was not measurable however in 4 patients with abnormal invasive measurements. Of these, 3 datapoints showed elevated PR-EDV (>1.6m/s), the other was also not measurable. Septal E/e' was measurable at 80% of opportunities, though only showed moderate correlation with PCWP or LV-EDP as measured ($r=0.53$, $p<0.005$). 74% of those with normal septal E/e' (<14) still had elevated LV-EDP (>10mmHg).
Conclusions: This represents the largest validation study for the use of TTE to monitor right heart pressures in paediatric cardiomyopathy with restrictive physiology. Where measurable, TR-Vmax offered useful estimates of systolic pulmonary artery pressures, and values under 3.0m/s correlated with normal pulmonary vascular resistance. Other measures, including indexed atrial area, pulmonary venous and mitral inflow dopplers, correlated poorly with corresponding invasive measures, so questioning their clinical use.

Keywords: Restrictive, cardiomyopathy, pulmonary, pressures, echo



Bland Altman Plot - RV systolic pressures, ECHO-derived vs invasive measures

PP-396
Right ventricle and pulmonary artery flow dynamics by blood speckle tracking echocardiography in neonates with pulmonary hypertension

Jose Carlos Aldana Aguirre[1], Pablo Perez Lopez[2], Wei Hui[1], Matthew Henry[1], Solveig Fadnes[3], Siri Ann Nyrnes[3], Amish Jain[4] and Luc Mertens[1]
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Background and Aim: Pulmonary hypertension of the neonate (PHTN) can lead to hypoxic respiratory failure and cardiovascular

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dysfunction. It is associated with altered cardiovascular flow dynamics. The objective of this study is to assess the right ventricle (RV) and pulmonary artery flow dynamics in neonates with PHTN and compare them to healthy controls using Blood Speckle Tracking echocardiography (BST).
Method: In total 36 controls and 16 PPHN neonates were included in the study. Echocardiography was performed within 72 hours after birth. BST data were acquired using a 12MHz probe on the apical four-chamber and high-parasternal short-axis views using E95 ultrasound scanner (GE Healthcare, USA). BST parameters analyzed off-line using custom-made software (Pyusview, NTNU, Norway). Analysis included energy loss (EL), vortex complexity (VC), vorticity (VO). Conventional functional parameters included right ventricular (RV) fractional area change (RVFAC), tricuspid annular plane excursion (TAPSE), and RV longitudinal strain. Differences between the groups were assessed using non-parametric statistical tests.
Results: The neonates with PHTN were more frequently males, had lower Apgars and all were on mechanical ventilation. The most common etiology for PHTN was meconium aspiration syndrome (MAS). Compared to normal controls, neonates with PHTN had significantly higher average RV systolic EL (0.0433 vs 0.0119 mW/m, p=0.043); and higher average and maximal systolic VO (0.3602 vs 0.1616 Hz, p=0.037and 2.06 vs 1.10 Hz, P=0.024). Compared to controls, neonates with PHTN had higher maximal PA diastolic EL (0.3940 vs 0.0666 mW/m, p=0.047). VC in the RV and PA was significantly higher in PHTN. Conventional RV functional parameters were lower in neonates with PHTN (Tables 1 and 2).
Conclusions: PHTN is associated with altered flow dynamics in the RV and in the PA. In both structures there is a more turbulent flow pattern represented by the higher vortex complexity. In the RV this mainly occurs in systole while in the PA this is mainly observed in diastole. The abnormal diastolic PA blood flow is likely related to increased pulmonary vascular resistance or ductal flow. This will need further evaluation in future studies.

Keywords: pulmonary hypertension, echocardiography, blood speckle tracking, neonates

Table 1 and 2. Echocardiographic data

Table 1. Echocardiographic analysis from apical 4 chamber view.

	Controls (n=36)	Pulmonary Hypertension (n=16)
Hours post birth at the time of echocardiography	22 (16-27)	21 (19-42)
Heart rate (bpm)	124 (110-131)	114 (97-145)
Conventional echocardiographic analysis		
TAPSE (cm)	0.84 (0.68-0.90)	0.63 (0.59-0.82) *
RVFAC (%)	30.7 (25.3-33.8)	16.8 (10.1-28.0) *
RV strain (%)	-20.9 (-18.8, -24.3)	-14.0 (-6.8, -20.7) *
BST echocardiographic analysis		
Avg EL systole (mW/m)	0.0119 (0.0070-0.0199)	.0433 (.0141-0.1237) *
Avg EL diastole (mW/m)	0.6328 (0.3038-1.13)	0.5554 (0.3055-1.61)
Max EL systole (mW/m)	0.1188 (0.364-0.3353)	0.2270 (0.0848-0.8336)
Max EL diastole (mW/m)	1.28 (0.8925-3.099)	1.09 (0.5024-3.60)
Avg VC systole	0.0106 (0.0038-0.0525)	0.0562 (0.0254-0.0990)*
Avg VC diastole	0.0421 (0.0184-0.1052)	0.1289 (0.0602-0.2321)*
Max VC systole	0.0632 (0.0039-0.2287)	0.3134 (0.1006-0.3885)*
Max VC diastole	0.1641 (0.0893-0.3311)	0.4445 (0.2766-0.6939)*
Avg VO systole (Hz)	0.1616 (0.0665-0.2506)	0.3602 (0.1280-0.7461) *
Avg VO diastole (Hz)	5.12 (3.11-7.82)	5.41 (1.55-10.20)
Max VO systole (Hz)	1.10 (0.45-1.66)	2.06 (1.26-6.31) *
Max VO diastole (Hz)	11.91 (8.69-17.6)	12.86 (7.46-22.21)

Avg, average; BST, blood speckle tracking; EL, Energy loss; Max, maximal; RVFAC, right ventricular fractional area change; TAPSE, tricuspid annular plane systolic excursion; VC, vortex complexity; VO, Vorticity.
Values expressed as median, quartiles (Q1-Q3).
*When *p* value is <0.05.

Table 2. Echocardiographic analysis from main pulmonary artery (parasternal short axis).

BST echocardiography analysis	Controls (n=36)	Pulmonary Hypertension (n=16)
Avg EL systole (mW/m)	0.7567 (0.5002-1.1657)	0.6101 (0.3958-1.0218)
Avg EL diastole (mW/m)	0.0066 (0.0015-0.0423)	0.0353 (0.0055-0.253)
Max EL systole (mW/m)	1.5282 (1.0058-2.1365)	1.3241 (0.6849-1.9508)
Max EL diastole (mW/m)	0.0666 (0.0264-0.3763)	0.3940 (0.0949-0.6327)*
Avg VC systole	0.0274 (0.0166-0.0422)	0.0627 (0.0390-0.1665)*
Avg VC diastole	0.0091 (0.0002-0.0521)	0.0515 (0.0069-0.1642)*
Max VC systole	0.0876 (0.0320-0.1882)	0.3312 (0.0844-0.4134)*
Max VC diastole	0.0461 (0.0004-0.2244)	0.2626 (0.0231-0.4214)*
Avg VO systole (Hz)	16.7392 (12.5076-18.6648)	13.5239 (9.0795-20.0076)
Avg VO diastole (Hz)	0.1999 (0.0354-1.4165)	0.9315 (0.314-5.1652)
Max VO systole (Hz)	27.3315 (20.8726-32.0219)	25.6486 (18.7071-28.066)
Max VO diastole (Hz)	2.947 (0.5795-9.8449)	7.2081 (2.4023-16.6491)

Avg, average; BST, blood speckle tracking; EL, Energy loss; VC, vortex complexity; VO, Vorticity.
Values expressed as median, quartiles (Q1-Q3).
*When *p* value is <0.05.

PP-397
Preventing permanent pacing in paediatric heart transplantation with theophylline

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Background and Aim: Bradyarrythmias are a common early complication after heart transplantation (HT), mostly sinus node dysfunction (SND). Biatrial anastomosis, older donors, prolonged ischaemic time, donor-recipient somatometric discordance and antiarrhythmic drugs are risk factors. Theophylline, an adenosine antagonist, has been used successfully in adults to facilitate chronotropic support withdrawal and prevent permanent pacemaker implantation (PPMI). We aim to present a paediatric case of prevention from PPMI after HT using theophylline.
Method: We reviewed the data and reported the case.
Results: We present a 16-year-old female with history of cytomegalovirus infection with pericarditis and severe allograft vasculopathy 4 years after her first HT, received for genetic dilated cardiomyopathy. Treatment with tacrolimus, steroids and mycophenolate, switched to sirolimus after vasculopathy diagnosis. Negative HLA antibodies. No history of arrhythmias or antiarrhythmic drugs use. She underwent second HT from a young donor, without donor-recipient weight discordance. Shumway-Lower technique was used, with 170 minutes of ischaemic time. Induction therapy was thymoglobulin, mycophenolate, steroids and tacrolimus. The allograft showed atrioventricular desynchrony and heart rate of 100 bpm from nodal origin since surgery, suspecting SND. She needed transient pacemaker until fifth day after surgery and isoprenaline until tenth day, presenting 60bpm after withdrawal. Despite, she was discharged from paediatric intensive care the eleventh day without other mayor complications. On the cardiology ward she maintained 35-50bpm, without pauses or syncope but showing dyspnoea during rehabilitation. Theophylline was started on twenty-third day (200mg bid, increased to 300mg bid three days after, reaching adequate levels of 14μg/ml), maintaining 50-80bpm. She suffered minor side effects such as mild tremor and sleep disturbance, but she could do rehabilitation without symptoms. Heart biopsies on eighth and twenty-fifth days without rejection. She was discharged on thirty-third day. She recovered sinus rhythm the fourth month with good response to exercise, proved by 24h ECG Holter monitoring. Then, theophylline was gradually withdrawn and suspended three months after. Followed up to eighteenth month, she maintains 65bpm sinus rhythm at rest, without symptoms and functional class I.
Conclusions: SND is a complication after HT in children with risk factors. Theophylline could be an alternative to PPMI to treat early post HT SND in children.

Keywords: Paediatric heart transplant, bradyarrhythmias, sinus node disfunction, permanent pacemaker, theophylline

PP-398
Diagnostic yield of routine biopsies in a pediatric heart transplant collective

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Background and Aim: Routine endomyocardial biopsies (EMB) are part of the standard of care in pediatric heart transplantation, although the set intervals vary between transplant centers. In patients beyond infancy, routine biopsies are usually carried out four weeks, six months to one year and every five years after transplantation at our center. As possible less invasive alternatives begin to arise, we set out to evaluate the results of routine versus clinically indicated biopsies.

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Method: Biopsies carried out over the course of 6.5 years at our pediatric heart transplant center were retrospectively analyzed with regard to indication and therapeutic consequence.
Results: 122 biopsies were done in 48 organ recipients. Of 17/122 (14%) biopsies indicated by clinical suspicion of acute rejection, 47% showed histopathological signs of rejection and 30% had therapeutic consequences (number needed to treat = 3.4), 13% had incipient histopathological transplant vasculopathy. 105/122 were surveillance biopsies, 46 of which were carried out after recent transplantation (<6 weeks) or as follow-up after recent rejection and had a diagnostic yield of 54%, with therapeutic consequences in 17% and vasculopathy in 2%. Of the remaining routine biopsies (n = 59), 87% showed no rejection and in the residual 13% only mild cellular or humoral rejection was detected (1R, AMR1); signs of vasculopathy were found in 3%.
Conclusions: In our pediatric heart transplantation center, indication biopsies as well as biopsies after recent transplantation or rejection had a high diagnostic yield during the observation period, corresponding to a number needed to treat of 3.4 and 5.8, respectively. Routine biopsies detected mild rejection or incipient histopathological transplant vasculopathy. Intensified risk management was necessary in a fraction of cases but did not result in profound changes of immunosuppression. Despite EMB being the gold standard to detect rejection after HTX, the number of routine biopsies in post-transplant care can possibly be reduced further by non-invasive measures, if there is evidence of adequate sensitivity and specificity in the pediatric age group.

Keywords: heart transplantation, rejection, biopsy, surveillance

PP-399

Torque-teno viral load - a new tool to predict donor-specific antibodies in pediatric heart transplantation recipients

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Background and Aim: In pediatric solid organ transplantation, individual clinical immunosuppression varies despite standardized and trough level controlled immunosuppressants. Assessment of Torque-Teno virus (TTV) has recently been proposed as a surrogate parameter for this purpose. We report the first worldwide experience with TT viral load in pediatric heart transplant recipients.
Method: We retrospectively evaluated TT viral load in pediatric heart transplant recipients over the course of 6.5 years at our pediatric heart transplant center regarding the development of donor-specific antibodies (DSA) and biopsy-proven rejection.
Results: 360 TTV measurements from 43 pediatric heart transplant recipients were analyzed. Median viral load was 107 copies/mL ±1.8 (IQR 5-8, range 0-10). Patients who developed donor-specific antibodies had significantly lower TTV values. This was also true for those with biopsy-proven humoral or cellular rejection. Multivariate regression uncovered TTV levels as the leading predictor for DSA development. Patients with TTV log10 levels of 5 or less had a threefold increased relative risk for DSA development compared to those with TTV of log10 of 7 or more (Odds Ratio 7.15).
Conclusions: We evaluated TTV as a possible surrogate parameter for individualized guidance of immunosuppression over a period of 6.5 years in our pediatric heart center in an observational study. Data suggest that patients with TTV log10 levels of less than 6 are at a significantly increased risk for DSA development and allograft rejection independent of tacrolimus trough levels.

Keywords: transplantation, rejection, donor-specific antibodies, immunosuppression, Torque-Teno, surveillance

PP-400

Impaired pulmonary circulation and right ventricular function in trisomy 18 with ventricular septal defect after pulmonary artery banding

Kanta Kishi[1], Shintarou Nemoto[2], Noriyasu Ozaki[1], Yutaka Odanaka[1], Atsuko Ashida[1], Hayato Konishi[2], Akiyo Suzuki[2] and Akira Ashida[3]
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Background and Aim: Surgical cardiac intervention improves the outcomes of Trisomy 18 (T18) with ventricular septal defect (VSD). The outcome after palliative surgery, such as pulmonary artery banding (PAB), remains poorer than that after corrective surgery. One of the causes of death in T18 is cardiac issues, including pulmonary hypertension and heart failure. The hemodynamic impairments could remain even after PAB in T18. We aimed to clarify the hemodynamic issues after PAB in T18.
Method: This retrospective study was conducted at a single institute. Infants with VSD who underwent PAB before corrective surgery between 2009 and 2022 were included. Infants with genetic disorders other than T18 were excluded. We divided the infants into two groups, normal karyotypes (NK) and T18, and compared the parameters of cardiac catheterization before corrective surgery and clinical backgrounds between the two groups.
Results: Seventeen NK infants and 21 T18 infants were included. The age at PAB and circumference of the PAB did not differ between the two groups. The age of the T18 group at corrective surgery was higher than that of the NK group, but the body weight was similar between the two groups. Regarding the parameters of cardiac catheterization, mean pulmonary arterial pressure (mPAP), pulmonary vascular resistance index (PVRi), and right ventricular end-diastolic pressure (RVEDP) were significantly higher in the T18 group compared to the NK group. (mPAP: 23.7 +/- 1.5 vs. 18.5 +/- 6.3 mmHg, p=0.0265, PVRi: 3.49 +/- 0.34 vs 2.19 +/- 0.38 WU*m2, p=0.0152, RVEDP: 10.6 +/- 0.6 vs 7.6 +/- 0.6 mmHg, p=0.0008, representing mean +/- SD) The Qp/Qs ratio and the parameters related to left ventricle and systemic circulation were not different between two groups. The NT-proBNP levels were higher in the T18 group than in the NK group. (3284.2 +/- 2817.1 vs 528.7 +/- 555.8 pg/ml, p=0.0007) The relation between mPAP and RVEDP (r=0.571, p=0.0001), RVEDP and NT-proBNP (r=0.550, p=0.0006) indicated the significant positive correlations.
Conclusions: In T18, mPAP and PVRi were still elevated even after PAB, accompanied by RVEDP and NT proBNP elevation.

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Persistent pulmonary circulation impairments with right ventricular dysfunction remain in T18 after PAB.

Keywords: Trisomy 18, pulmonary artery banding, right ventricular dysfunction, pulmonary hypertension, cardiac surgery

PP-401

Initial experience with the molecular microscope diagnostic system (MMDX-HEART) in paediatric heart transplant recipients – a case series

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Background and Aim: The Molecular Microscope Diagnostic System for heart transplantation (MMDx-Heart) uses microarrays to measure biopsy-based gene expression and ensembles of machine learning algorithms to interpret the results and compare each new biopsy to a large reference set of earlier biopsies. MMDx assesses T cell–mediated rejection (TCMR), antibody-mediated rejection (AMR), parenchymal injury, and atrophy-fibrosis, continually "learning" from new biopsies (Halloran PF, Madill-Thomsen KS. The Molecular Microscope Diagnostic System: Assessment of Rejection and Injury in Heart Transplant Biopsies. Transplantation. 2023;107(1):27-44.). The aim was to summarize initial experience with MMDx-Heart in paediatric patients and to evaluate the impact of MMDx results on their clinical management.
Method: A single-centre retrospective analysis of MMDx results (November 2022 – November 2023). An additional sample for MMDx was taken during each endomyocardial biopsy in patients after heart transplantation. After obtaining standard results (histology, donor specific antibodies), MMDx was performed only in patients with discrepancies between clinical (echocardiography, haemodynamic parameters) and histological findings. A comparison was made between the classification of rejection using standard methods and MMDx, taking into account the impact on further patient management.
Results: MMDx tests were performed in 3 patients (multiple biopsies in each). In patients 1 and 2, rejection was suspected from histology in the context of favourable clinical findings. Despite a grade 2R TCMR in one patient and a grade pAMR1 (H+) in the other one, MMDx showed no definite rejection in both, resulting in less anti-rejection treatment than histology would have directed. No deterioration occurred in both. Patient 3 had a history of persistent AMR and low-yield histology showed no significant rejection. MMDx demonstrated severe AMR and resulted in a more aggressive anti-rejection treatment.
Conclusions: MMDx results influenced the treatment substantially in all 3 patients in whom the method was performed. MMDx-Heart can improve the diagnosis of rejection from endomyocardial biopsies after heart transplantation. It may be particularly advantageous in children, where the number of biopsies is more limited than in adults. MMDx-Heart is now available in Europe.

Keywords: heart transplantation, rejection, microarray, mRNA expression

Sports Cardiology, Physical Activity and Prevention

PP-403

Marfanpower: results of a home-based cardiorespiratory and muscle rehabilitation program for children and young adults with marfan syndrome

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Background and Aim: Marfan syndrome (MFS) is a rare genetic disease leading to a multisystem damage related to connective tissue fragility. Chronic fatigue and decreased physical endurance are almost constant complaints of patients with MFS, whose muscular mass is reduced from childhood. Muscle mass worsens through adolescence, which could explain the bone mass deficit observed in this population. We hypothesize that a personalized exercise rehabilitation program will improve general fitness and quality of life of these patients.
Method: Self-controlled study with a 6-month home-based cycloergometer and muscular strengthening personalised rehabilitation program based on first ventilatory-threshold. Baseline evaluation was performed 3 months prior to the start of the rehabilitation program (control-phase), then at the beginning of the rehabilitation program and at 6 months, with a mid-term evaluation at 3 months.
Results: We included 28 Marfan or associated syndrome patients between 7 and 20 years (mean 12.8±3.69 years), of which 11 were females, with a mild aortic dilatation for 50% (mean z-score +2,4), no major valvopathy, no cardiac impairment and a history of pneumothorax for 3 of them; most where under preventive beta-blocker treatment (93%). After a 6-month rehabilitation program no progression in aortic diameters was found. Significant improvement in first ventilatory-threshold was achieved (mean 17.83 to 22.37 mL.min-1.kg-1; $p=0.035$), alongside an increase in maximal sustained workload with a mean +24.5 Watts (CI95%= [17.37; 31.63]; $p<0.001$) associated to a small but significant increase in VO2 (+131.58 ml.min-1; CI95%= [30.17; 232.99]; $p=0.016$), whereas maximal heart rate at effort was reduced by 29.95 bpm (CI95%= [16.17; 43.73]; $p<0.001$). Muscular strength gain was shown both by dominant hand-grip (mean +4.3 kg; CI95%= [2.45; 5.2]; $p<0.001$) and dominant leg strength (+67.33 Nm; CI95%= [25.08; 109.59]; $p=0.005$). Overall, QoL of participants improved.
Conclusions: Young patients with Marfan or associated syndromes can safely benefit for a home-based rehabilitation program personalised according to their baseline capacities, improving both their overall fitness, muscular strength and cardiovascular health; which may have a favourable impact in their QoL.

Keywords: Rehabilitation, Marfan Syndrome, Sport, CPET, Muscular strength, Quality of Life

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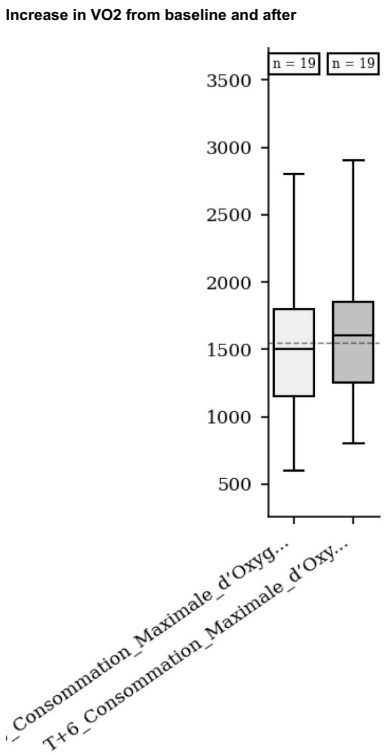

The difference between maximal oxygen uptake (mL.min-1) during cardiopulmonary exercise test using a cyclo-ergometer with incremental load, increased from baseline to the end of a 6-month rehabilitation program. Assessed with the Student's paired t-test. Alpha risk was set to 5% (α = 0.05). The normality was verified with the Shapiro-Wilk test..

PP-404
Does severity of congenital heart defects has an impact on growth?

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Background and Aim: Treatment options and therefore, general health in children with congenital heart disease (CHD) got increasingly better over the last decades. Growth is an important marker to evaluate healthy development in childhood and adolescence, especially in patients with chronic diseases. This study aimed to evaluate the somatic development in children with CHD compared to their healthy peers in Germany.
Method: Retrospective, longitudinal analysis of standardized measurements of height, weight, and head circumference of children with CHD aged 0 to 18 years (n=18027) were conducted by using the national computerized CrescNet database. These data were compared to the data of healthy children and adolescents from the same geographic region. Inclusion was based on the ICD-10 codes for CHD, patients with other growth altering diseases or medications were excluded. Cardiac defects were grouped into mild, moderate, or severe according to severity of the disease.
Results: Preliminary results show an impact of the severity of the heart defect on the height, final height, weight, and head circumference of the patients. Comparison with predicted height according to parental height was made where available. Specific analyses were made for different heart defects including single ventricles and transposition of the great arteries. Furthermore, somatic development of patients with atrioventricular septal defects (AVSD) with and without Down´s Syndrome in comparison with children with Down´s syndrome and without heart defects was studied. Final results will be presented at the meeting.
Conclusions: Despite good overall outcomes and better survival rates, growth as a marker of healthy development in children with CHD can still be altered and should be closely monitored to offer possible treatments.

Keywords: growth, somatic development

Cardiac Dysrhythmias and Electrophysiology

PP-405
Sudden cardiac arrest reveals predictable double "Double hit"

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Background and Aim: Clinical CASE: We report on a 4-year-old boy who survived sudden cardiac arrest due to ventricular tachycardia. The previously healthy and normally developed child had a witnessed arrest related to activity/excitement. Immediately performed bystander cardiopulmonary resuscitation over 8 minutes was continued by paramedics. The initial rhythm revealed ventricular tachycardia. After defibrillation return of spontaneous circulation was established. ECG monitoring showed severely prolonged QTc intervals above 600 ms, which did not normalize after correction of an initially seen hypokalemia. Monitor and 12-lead ECG also showed T alternans and PVCs with R-on-T phenomenon. The diagnosis of Long QT Syndrome was made clinically according to the Schwartz-Score and the child was started on propranolol. Under beta blockade no further cardiac events occurred. In a shared-decision making process with the family the indication for a secondary-prophylactic ICD-implantation was confirmed.
Method: Family counseling and genetics:
Results: Next-Generation-Sequencing (NGS)-based panel diagnostics revealed a "double hit" situation in the index patient with compound heterozygosity in the KCNQ1 gene: a rare missense mutation (c.613G>A, p.(Val205Met)) and a rare nonsense

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mutation (1344dup, p.(Glu449Argfs*14)). Genetic testing of first-degree relatives was performed and showed one of the KCNQ1 mutations in each parent, who clinically both presented with borderline QTc and mild repolarization abnormalities. Of two siblings, one showed the same "double hit" situation with both mutations and phenotypically a QTc >500ms, the other one was genotypically and phenotypically unaffected. The affected sibling was started on beta blocker therapy prior to the genetic test result due to the severely abnormal ECG. Additionally, in retrospect both affected children had shown fetal bradycardia, which had not been followed up postnatally.
Conclusions: Take home messages: "Double hit" mutations in LQTS can cause severe phenotypes with early manifestation. Timely clinical and genetic testing in family members of confirmed LQTS patients is mandatory. Early warning signs like fetal bradycardia should be followed up consequently.

Keywords: LQTS, compound heterozygosity

Sports Cardiology, Physical Activity and Prevention

PP-406
The use of an apple watch for the oxygen saturation measurement in cyanotic heart disease

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Background and Aim: Accurate measurement of transcutaneous oxygen saturation (tSO2) is of upmost importance for the assessment of cyanosis in congenital heart disease. Aim of this study was the evaluation of a supplementary tSO2 measurement with an Apple watch® in children with cyanotic heart disease.
Method: During a six minute walk test (6MWT), measurement of tSO2 was performed simultaneously with an oximeter (Nellcor, Medtronic, USA) and an Apple watch® Series 7 (Apple inc, USA) in 36 children with cyanotic heart disease.
Results: Median age was 9.2 (IQR 5.7-13.8) years. tSO2 measurement with the Apple watch® was possible in 35/36 and 34/36 subjects before and after 6MWT, respectively. Children, in whom Apple watch® measurement was not possible, had a tSO2 < 85% on oximeter. Before 6MWT, median tSO2 was 93 (IQR 91-97) % measured by oximeter and 95 (IQR 93-96) % by the Apple watch®. After a median walking distance of 437 (IQR 360-487) m, tSO2 dropped to 92 (IQR 88-95, ($p< 0.001$)) % by oximeter and to 94 (IQR 90-96, $p=0.013$) % measured with the Apple watch®.
Conclusions: In children with mild cyanosis measurement of tSO2 with an Apple watch® showed valid results if tSO2 was > 85%. In this setting, assessment of tSO2 at home with the apple watch® could be helpful for patients and their families. In children with severe cyanosis tSO2 measurements with the Apple watch® were not reliable and thus not helpful. Overall, there was a small but significant tendency for the Apple watch to display higher oxygen saturations compared to the medical oximeter.

Keywords: cyanotic heart disease, apple watch, oxygen measurement, children with cyanotic heart disease

PP-407
Cardiopulmonary function in pediatric post-covid-19: a controlled clinical trial

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Background and Aim: The importance of post-COVID-19 in adults has been recognized in many publications. However, in children, all studies are limited to surveys and chart analyses. Especially objective data in the form of CPET are still scarce. This study therefore aims to investigate the cardiopulmonary effects of post-COVID-19 on children and adolescents in order to objectify the symptoms of post-COVID-19 in children. This is the first study to evaluate cardiopulmonary exercise data in children after infection with SARS-CoV-2.
Method: In the FASCINATE study (a cross-sectional study) children fulfilling the criteria of post-COVID-19 and children who showed no signs of post-COVID-19 after infection with SARS-CoV-2 underwent cardiopulmonary exercise testing on a treadmill. A questionnaire with regards to physical activity before, during and after the infection with SARS-CoV-2 was completed.
Results: We were able to recruit 20 children suffering from post-COVID-19 (mean age: 12.8 ± 2.4 years, 60% females) and 28 children after SARS-CoV-2 infection without symptoms of post-COVID-19 (mean age: 11.7 ± 3.5 years, 50% females). All participants completed a maximal treadmill test with a significantly lower in the post-COVID-19 group (37.4 ± 8.8 ml/kg/min vs. 43.0 ± 6.7 ml/kg/min). This significant difference was limited to the female group. This significance did not persist when comparing the achieved percent of predicted norms. No other parameters (O2pulse, peak VE, VE/VCO2) showed any significant differences. The only significant correlation proved to be between and the rest period from physical activity after the infection with SARS-CoV-2.
Conclusions: This is the first study to investigate post-COVID-19 in children using objectifiable measurements such as CPET. Although there was a significantly reduced in the group suffering from post-COVID-19, this was not true when analyzing the percent of predicted values and was limited to the girls. Furthermore, no pathological findings with respect to cardiac or pulmonary functions could be discerned. In light of the significant correlation between and rest period after infection, deconditioning seems to be the main cause for the symptoms experienced by children suffering from post-COVID-19.

Keywords: VO2 peak, cardiopulmonary exercise testing, post-COVID, fitness, exercise capacity, physical activity

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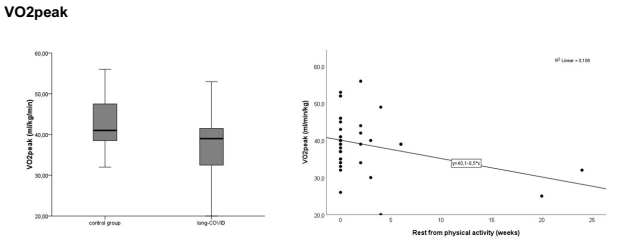

a: Median, as well as interquartile range, Minimum and Maximum of the VO2 peak between the children suffering from post-COVID-19 and the control group. The difference was significant. b: Correlation between the rest period after infection with SARS-CoV-2 in weeks and the VO2 peak.

PP-408

Aerobic physical capacity and health-related quality of life in children with sickle cell disease

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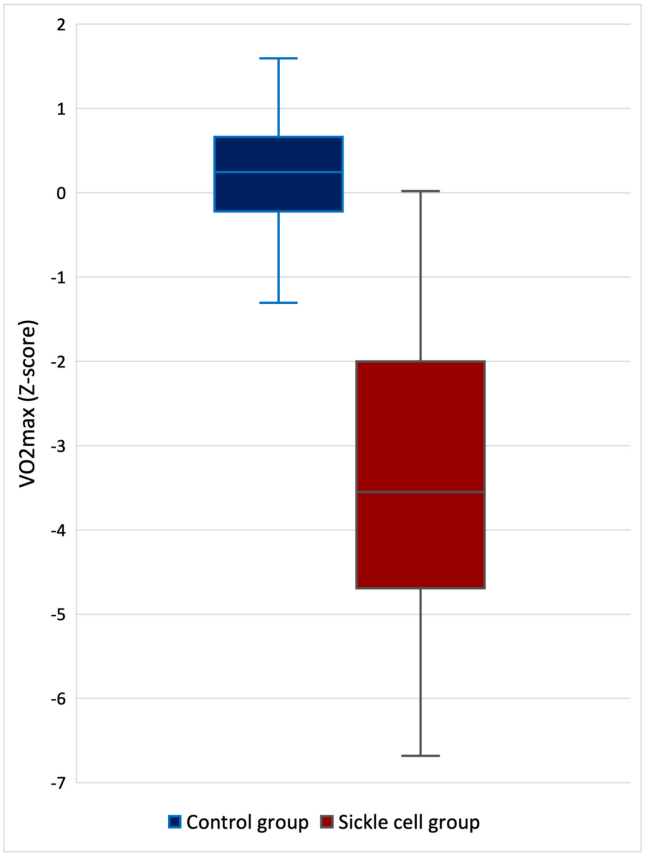
Background and Aim: Patients with sickle cell disease are dramatically affected by cardiovascular morbidity. Aerobic fitness is a predictor of cardiovascular health which correlates and health-related quality of life. We purpose to evaluate the aerobic capacity by cardiopulmonary exercise test (CPET) in children and adolescents with sickle cell disease in comparison with healthy matched controls and to determine the factors (considering HRQoL, level of physical activity, knowledge of disease) associated with impaired maximum oxygen uptake (VO2max) in this population.

Method: This prospective cross-sectional controlled study was carried out from 2021 to 2022 in a tertiary care academic institution. Children and adolescents aged 6 to 17 years with a confirmed diagnosis of sickle cell disease were consecutively screened during their routine follow-up and healthy age and sex-matched controls were recruited during the same study period. The CPET was performed for these 2 groups and study questionnaires included the PedsQL™ 4.0. questionnaire, the physical activity questionnaire by Ricci and Gagnon and a sickle cell disease knowledge questionnaire.

Results: A total of 72 children (24 with sickle cell disease and 48 healthy controls), aged 6 to 17 years old underwent a complete CPET. Children with sickle cell disease had a poor aerobic capacity, with median VO2max Z-score values significantly lower than matched controls (-3.55 [-4.68; -2.02] vs. 0.25 [-0.22; 0.66], P<0.01, respectively), and a high proportion of 92% children affected by an impaired aerobic capacity (VO2max Z-score<-1.64). The ventilatory anaerobic threshold was impaired in 71% of children with sickle cell disease. The VO2max decrease was associated with the level of anemia, the existence of a homozygote HbS/S mutation, restrictive lung disease (low forced vital capacity associated to low total lung capacity) and health-related quality of life. In multivariate analysis, VO2max Z-score was associated with hemoglobin level (ß=0.37; 95%CI= [0.01; 0.72]; P=0.04).

Conclusions: Aerobic capacity is poor in children with sickle cell disease. VO2max decrease is associated with the level of anemia, the existence of a homozygote HbS/S mutation, lung function,

VO2max Z-score in the sickle cell disease group and in matched controls



Box plot of VO2max Z-score. The top and bottom of the box represent the first and third quartiles. Inside each box, the band represents the second quartile (median). The whiskers represent the minimum and maximum values.

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and health-related quality of life. These results represent a signal in favor of early initiation of cardiac rehabilitation in patients with sickle cell disease.

Keywords: VO2max, aerobic exercise, heath-related quality of life, cardiac rehabilitation

PP-409
Sport participation in children with congenital heart disease

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Background and Aim: Participation in sport benefits the growth and development of children and can affect activity habits over the lifespan. Children with congenital heart disease (CHD) may be limited in their ability to participate in sport due to delayed gross motor development, physiological, psychological, or medical limitations. We sought to understand how many of our CHD patients participate in sport.
Method: A single-centre retrospective review (May 2016-October 2023) was conducted in CHD patients with a cardiopulmonary exercise test (CPET) and an assessment of sport participation. Criteria for a maximal CPET test included: a respiratory exchange ratio>1.0, and/or a peak heart rate >200 bpm. VO2peak and HR z-scores were calculated. Sport participation was recorded based on frequency, intensity, time, and type and was categorized into 3 groups: Participation (at least 2 days/week); No sport participation but regular activity; No participation. Frequency tables were generated. A one-way ANOVA with a Tukey post-hoc test was used to determine differences between groups. $P<0.05$ was considered statistically significant.
Results: Seventy-four CHD patients were included: 8 with aortic valve disease (no intervention); 12 with coarctation; 6 following the Ross procedure for aortic valve stenosis; 25 with tetralogy of Fallot, 11 with transposition of the great arteries; and 12 with Fontan palliation. Forty-six percent of our study cohort were involved in sport, 19% regularly attended the gym or achieved >10,000 steps/day and 35% did not participate in any sport. There was no difference in HR z-score (-0.77 vs -1.03 vs -0.73; $p\geq0.05$) or peak HR (190 vs 186 vs 183 bpm; $p\geq0.05$) for those who participate, do not participate but are active, or those who do not participate in sport, respectively. VO2peak z-score was different between groups ($p=0.002$) with the lowest z-score in those who do not participate in sport compared to those who do participate in sport (-1.63 vs -0.77; $p<0.001$).
Conclusions: Thirty-five percent of our patient group did not participate in sport and had a lower VO2peak z-score compared to those who did participate in sport. Understanding the barriers to sport participation in this population may provide important insight and direct intervention strategies.

Keywords: Exercise, adolescents, cardiology

PP-411
Cardiological assessment results prior to sports participation in school-aged children of sports-enthusiastic cities

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Background and Aim: Sports enhance physical health and fitness in children, yet also bring potential health risks, including the risk of sudden cardiac death (SCD) during sports activities, deeply impacting society. The scope and effectiveness of pre-participation evaluations in sports remain a globally debated issue. This study, conducted in three centers in Giresun, Ordu, and Rize in the Black Sea region of Turkey, aims to assess the health status of children before participating in sports, identify risk groups, and and raise awareness regarding this critical concern.
Method: The study encompassed 2500 children who visited pediatric cardiology clinics for pre-participation evaluation from May 2022 to November 2023. Participants were assessed according to the American Heart Association (AHA)'s 12-question guideline. Electrocardiogram (ECG) and echocardiography assessments were conducted. Exercise tests and rhythm holter were also requested for some patients. Data included personal and family medical history, anthropometric features, physical examination, sports history, and electrocardiographic and echocardiographic information. Patients with incomplete data were excluded from the study.
Results: Of the participants, 1687 were male (67.5%) and 813 were female (32.5%), with a median age of 12.2 years (range 5-18 years). The most common sports disciplines were football, athletics, taekwondo, and swimming. Electrocardiographic abnormalities were found in 146 cases (5.84%). As a result of the examinations, 32 participants (1.28%) were not approved for competitive sports. Three cases were denied participation due to the diagnosis of long QT syndrome in basal ECG. Echocardiographic findings of varying degrees were observed in 220 participants (8.8%), with the most common findings being mitral valve insufficiency and patent foramen ovale. 29 participants with echocardiographic findings (6 hypertrophic cardiomyopathy, 19 bicuspid aorta and aortic root dilation, 2 aortic valve insufficiency, 2 mitral valve insufficiency) were not approved for sports participation following the recommendations of the American Heart Association.
Conclusions: While pre-sport cardiac assessment is important in many aspects, how it should be conducted remains controversial in the global and Turkish context. The study emphasizes the importance of pre-sport cardiac assessment aimed at reducing the risk of SCD. The outcomes of this multi-center study could guide the development of future guidelines.

Keywords: Sports participation, pediatric, sudden death, cardiac assessment

PP-412
Cardiopulmonary parameters in patients with tetralogy of fallot: The reference values for treadmill and cycle ergometer

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Background and Aim: Despite successful repair of Tetralogy of Fallot (rToF) in childhood, the residual pulmonary regurgitation in the majority of cases leads to right ventricular (RV) dilatation and/or dysfunction, left ventricular dysfunction and progressive impaired exercise capacity. Cardiopulmonary exercise test (CPET) has been

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able to assess really the cardiopulmonary function in these patients. However, although both cycle ergometer and treadmill protocols are often used interchangeably, there are still uncertainties regarding the "normal" values range for each approach in this population. Therefore, our aim was to assess the most used parameters in CPET, divided by sex and method.
Method: All CPET data of asymptomatic rToF patients who had a cardiac magnetic resonance and perform the test on a cycle ergometer or a treadmill between 2020-2023 were collected.
Results: Four hundred thirty-eight CPET exams (163 treadmill and 275 cycloergometer) were analyzed. The patients performing the test using the cycloergometer were slighter older (mean age 22.6±7.7 vs 21.5±8.5 years old). The CMR and CPET parameters subdivided according to gender and method are shown in Table 1 and 2.
Conclusions: The importance of our study is to provide useful range values of CPET parameters stratified by method and sex in a wide population of asymptomatic rToF patients

Keywords: cardiopulmonary exercise stress test, Tetralogy of Fallot, peak V02

Table 1 and Table 2

TREADMILL

	F					M				
	Mean	Median	Dev Std	25th	75th	Mean	Median	Dev Std	25th	75th
RVEDVi	117,7	116,8	23,6	102,6	133,7	126,0	124,6	24,0	111,8	141,2
RVESVi	51,4	51,1	15,2	40,5	58,5	58,0	59,6	12,8	48,8	66,5
RV EF	56,7	57,0	6,0	52,0	61,0	53,9	54,0	5,2	50,8	56,4
RV SV	101,9	101,0	21,3	89,0	113,5	115,3	115,5	31,3	92,8	136,4
LVEDVi	76,6	72,9	13,0	68,2	84,1	78,6	78,4	13,5	71,1	88,6
LVESVi	31,1	30,9	6,0	26,6	34,9	33,6	34,1	7,4	28,8	38,3
LV EF [%]	59,3	59,0	6,8	56,0	63,0	57,3	57,0	4,4	54,9	60,0
Peak VO2	26,9	26,4	5,6	23,4	30,0	31,5	31,7	6,6	25,7	35,9
Percent-predicted peak	83,7	82,0	16,0	74,3	93,2	74,0	73,0	12,1	65,8	81,3
Peak O2 pulse [mL/beat]	9,0	9,1	1,9	7,5	10,2	11,7	11,6	2,7	10,0	13,3
Percent-predicted	86,0	86,0	14,5	77,3	97,9	75,1	72,2	13,4	66,4	83,9
VE/VCO2 peak	32,7	32,8	5,4	30,2	35,2	30,7	31,0	4,8	27,0	33,7
VE/VO2 peak	35,9	36,4	4,8	32,2	39,5	33,2	33,0	6,0	28,9	37,3
VE/VCO2 slope	30,3	29,8	4,3	27,0	33,7	29,2	28,9	4,8	25,2	33,0

BYCICLE

	F					M				
	Mean	Median	Dev Std	25th	75th	Mean	Median	Dev Std	25th	75th
RVEDVi	116,4	114,7	23,1	104,5	128,7	122,8	121,4	25,0	108,9	136,5
RVESVi	52,5	52,6	14,9	42,7	61,1	58,6	57,6	15,8	48,1	67,0
RV EF	55,0	55,1	6,2	50,4	59,9	52,5	51,4	8,0	48,6	56,0
RV SV	102,0	99,8	23,3	89,0	119,0	113,9	112,1	29,3	94,1	125,6
LVEDVi	80,1	77,7	15,7	70,6	87,5	86,8	87,8	15,3	75,4	96,4
LVESVi	33,2	31,7	8,7	27,5	37,4	38,1	38,6	9,1	31,7	43,0
LV EF [%]	57,9	58,0	5,3	55,3	61,0	56,5	56,2	6,8	53,8	59,0
Peak VO2	24,4	23,9	5,5	20,3	27,4	26,9	26,5	6,3	22,8	30,8
Percent-predicted peak	78,7	78,3	15,6	68,0	90,6	64,3	64,0	14,0	54,7	73,4
Peak O2 pulse [mL/beat]	8,8	8,5	2,4	7,2	9,9	11,2	10,9	2,8	9,3	12,8
Percent-predicted	82,8	79,5	18,4	74,1	89,9	80,8	76,9	27,4	63,8	97,1
VE/VCO2 peak	34,2	33,5	6,8	30,4	37,6	31,3	30,9	4,7	28,0	33,4
VE/VO2 peak	38,7	38,0	6,7	33,5	42,6	35,2	34,7	5,5	31,0	38,8
VE/VCO2 slope	30,2	29,3	5,2	27,0	32,5	27,8	27,4	4,2	25,2	30,4

The range values of CPET parameters stratified by method (Treadmill vs Bicycle)and gender

PP-413
Mental health in children with congenital heart disease – an anxiety and depression assessment

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Background and Aim: Nowadays, up to 95% of children with congenital heart disease (CHD) reach adulthood. This progress is largely due to medical advances and extensive research on predictors of physical health in children with CHD. However, little is known about alterations in mental health in this population. Therefore, the aim of this study was to examine differences in anxiety and depression between children with CHD and healthy children. Additionally, risk factors that may have an impact on the mental health of children with CHD are examined.
Method: Between August 2022 and October 2023, 123 children and adolescents (15.1 ± 2.8 years, 47.2% girls) with CHD and 66 healthy controls (12.9 ± 2.9 years, 57.6% girls) were recruited during routine medical examinations. The Beck-Anxiety-Inventory and Beck-Depression-Inventory-II were used to assess the extent of anxiety and depression. Single-factor ANCOVA was used to investigate differences in anxiety and depression scores between the CHD and the control group.
Results: According to published cut-off values, at least a mild form of anxiety was present in 42.4% of the CHD patients and 29.8% showed signs of depression. Comparing patients with CHD to healthy controls adjusted for sex and age, no difference in anxiety score (CHD: 9.4 ± 9.7 vs. healthy: 7.7 ± 8.5, p=0.200) and depression score (CHD: 7.9 ± 8.4 vs. healthy: 6.4 ± 7.9, p=0.214) was found. Multiple linear regressions revealed no significant associations between anxiety and depression scores and severity of disease (simple, moderate, complex), number of surgeries, as well as intake of medication.
Conclusions: Anxiety and depression are common among children with CHD. Larger sample size studies are needed to establish a clear comparison with healthy counterparts and to find parameters that are associated with anxiety and depression to tackle them in intervention studies.

Keywords: congenital heart disease, congenital heart defect, mental health, anxiety, depression

PP-415
Physiology of long distance diving data in pediatric fin swimmers

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Background and Aim: Swimming and diving are popular recreational activities. Although, to date, little is known about the cardiovascular adaption to submersion in children, a current study reported the first data on the diving reflex as cardiovascular adaption in children in a static protocol. Therefore, the current study intends to evaluate the effects of dynamic exercise during diving on the physiology in children.
Method: We used a stepwise apnea protocol with repetitive dives of 25m, 50m and 75m in a cohort of children training in the regional fin swimming club. Continuous measurement of heart rate, oxygen saturation and peripheral resistance index were done.
Results: Physiologic data and analysis of influencing factors on heart rate, oxygen saturation and peripheral vascular tone response are reported.
Conclusions: Preliminary data showed moderate redcution in oxygen saturation in distances more than 25m of diving and only a mild reduction in heart rate. Furthermore no adverse events or fatalities were recorded.

Keywords: diving, swimming, children, cardiovascular physiology, children

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PP-416

Motor skills in children with and without chronic diseases

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Background and Aim: Sports motor skills include the essential physical attributes and competencies necessary for engaging in athletic activities. These skills encompass a range of aspects, including strength, endurance, speed, agility, coordination, and stability. For children with chronic diseases, motor skills hold significant importance due to their influence on fitness levels, overall well-being, cognitive advancement, and involvement in everyday tasks. Additionally, these skills play a role in lowering the risk of additional chronic conditions and encouraging a lifelong commitment to physical activity.
Method: At the KidsTUMove summercamp 2023 in Bavaria Germany, the motor performance of children and adolescents with chronic diseases, like congenital heart defects and cancer, was measured using a strength, coordination, agility and a endurance test (Parts from the German Motoric Test and Munich Fitness Test).
Results: A total of 34 children and adolescents, aged between 6 and 17 years (10.94 ± 2.54) participated. Among them,15 had chronic diseases.The chronically ill children achieved a statistically significant higher number of jumps when jumping back and forth sideways (35.67±9.42 vs 26.68±7, 86, p=0.005). Girls with chronic diseases performed significantly better in sideways jumps (38.37 ±6.98 vs 24.44±7.53, p=0.001) and standing long jumps (146.25±17 vs 118.22±13,71, p=0.002). No statistically significant differences were observed when comparing the group of boys, as well as comparing children with heart disease to those with oncological conditions. In comparison to established norm values, 22 of the children demonstrated average to above-average performance, while 12 exhibited below-average performance.
Conclusions: The importance of training motor skills in children with chronic dieseases lies in improving their physical and mental health, enhancing their quality of life, and reducing the risk of secondary illnesses. An active lifestyle based on the development of motor skills can contribute to safeguarding their health in the long term. Therefore, the training of motor skills should be considered an integral part of the treatment and support for these children. While this study demonstrates that children with chronic illnesses do not perform worse than healthy children, it is nevertheless important to pursue further research in the area in chronically ill children and adolescents in order to be able to establish better rehabilitation measures.

Keywords: motor skills, chronic diseases,children, physical activity, health

PP-417

Benefits of sports in children with chronic diseases. A systematic review within the project kidstumove goes europe-cordially fit

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Background and Aim: The aim of this study was to identify sports-based interventions for children and adolescents with chronic diseases (CaAcd) and evaluate their influence on physical, psychological, and social well-being. The outcomes of this research will contribute to the comprehension of the potential advantages of sports interventions for CaAcd and offer insights for future initiatives to enhance their overall health and well-being.
Method: Within the EU-project KidsTUMove goes Europe-cordially fit, a total of 5 partner countries are involved, a systematic review across eight databases was conducted, adhering to PRISMA guidelines and employing an extensive search strategy to locate studies related to sport-based interventions for CaAcd. The review encompassed randomized controlled trials and observational studies that emphasized physical and psychosocial consequences.
Results: A total of 10,123 titles and abstracts were screened, 622 full-text records were reviewed, and 53 primary studies were included. These studies encompassed 2,384 participants, with an average of 45 ± 37 participants per study. Among the studies focused on CaAcd, aged 3 to 18 years, 19% (10 studies) concerned the attention for deficit hyperactivity disorder, 21% (11) to cerebral palsy, 30% (16) to autism spectrum disorders, and 17% (9) to obesity. Other conditions studied included cancer (5), asthma (1), and cystic fibrosis (1). The interventions involved diverse sports and physical activities tailored to each specific chronic disease. The duration and frequency of these interventions varied across studies. Most studies assessed physical outcomes, such as motor performance and physical fitness measures. Psychosocial outcomes were also evaluated, focusing on behavioral issues, social competencies, and health-related quality of life.
Conclusions: In summary, sport-based interventions demonstrated effective improvements in physical and psychosocial outcomes for CaAcd. These interventions were generally considered safe, and participants adhered to the prescribed protocols willingly. Nevertheless, there remains an insufficient amount of evidence concerning the implementation of these interventions. Future research should include interventions tailored to address the common challenges faced by CaAcd, offering a comprehensive understanding of the impact of sports interventions on those affected.

Keywords: children, chronic diseases, physical fitness, quality of life,

PP-418

Single coronary artery in a 14-year old basketball player – can he be cleared for competitive sports?

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Background and Aim: A single coronary artery (SCA) is a seldom anomaly. A right coronary artery arising from the left coronary artery system is an extremely rare and usually benign variant as long as an interarterial course can be excluded. The clinical significance

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of this condition in active children though remains unclear as there has been published an association of sudden cardiac death with SCA in young competitive athletes.
Method: CASE:
Results: A 14-year-old competitive basketball player presented with frequent premature ventricular contractions (PVC) in ECG and Holter-ECG on a sport's medical examination. Retrospectively he describes rare episodes of chest tightness as well as palpitations that usually terminate during exercise. ECG showed left axis deviation with sinus bradycardia, the PVCs had an inferior axis and left bundle branch block. Holter ECG in our institution showed extreme bradycardia of 27 bpm at nighttime with a mean frequency of 50 bpm as well as 7% monomorphic single PVC`s with no couplets or salves. Stress test showed 6 single PVCs at the beginning of exertion and none thereafter. Echocardiographic findings included a large left ventricle with a LVEDD of 68mm with a normal shortening fraction of 37%. Subsequently, cardiac MRI suggested a dilative Cardiomyopathy (LVEDVI 137 ml/m^2, RVEDVI 159 ml/m^2) with low but normal biventricular function. Therefore, cardiac catheterization was performed showing normal hemodynamics but a single coronary artery arising from the left coronary cusp and a wide circumflex branch peripherally giving origin to a right coronary artery with retrograde perfusion. There was no evidence for ischemia in the subsequent stress echocardiography also. Histopathologic evaluation gave evidence to a morphologically normal myocardial structure with no sign for cardiomyopathy or infection. During invasive electrophysiological study, he showed no PVCs, therefore no ablation therapy could be performed. The patient was then treated with Propranolol and cleared for sports.
Conclusions: Considering the reports that the risk for sudden cardiac death in young athletes is the highest in male basketball players combined with the single coronary artery possibly associated with SCD we nevertheless cleared our patient for competitive sports after a thorough workup for ischemia and under beta-blocker protection.

Keywords: Single Coronary Artery, Premature ventricular contractions

PP-419
Systemic vascular changes in response to normobaric hypoxia in healthy children

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Background and Aim: Hypoxia is well known to trigger an increase in pulmonary vascular resistance and right ventricular pressure. There has been relatively little focus on its effect on the systemic vasculature. In adults, it has been shown that the augmentation index (a marker of pulse wave reflection) decreases while carotid-femoral pulse wave velocity increases. This could indicate differential effects of normobaric hypoxia on the small-medium sized vs large arteries. It is the aim of this study to evaluate systemic arterial characteristics in healthy children under normoxia and normobaric hypoxia.
Method: This is an ongoing pilot study. 20 healthy children and young adults will be included. Testing is performed under normoxia and then repeated and under normobaric hypoxia, corresponding to an altitude of 3500m above sea level. Central blood pressure, heart rate, augmentation index corrected to a heart rate of 75/minute and pulse wave velocity as well as aging index are measured (SphygmoCor XCEL, DPA Meridian). Data are presented as median (interquartile range). Statistical analyses included the Wilcoxon signed rank test for paired samples.
Results: To date, 9 probands have undergone testing under both normoxia and hypoxia. Median age was 12 (range 9-18) years. There were no significant changes in heart rate, central blood pressure or pulse wave velocity. We did observe decreases in both parameters assessing arterial wave reflection, i.e. augmentation index corrected to a heart rate of 75/minute (mean -13.5 vs -23, p 0.025) and aging index (mean -0.8 vs -1.1, p 0.043).
Conclusions: Even though the number of children included at the present time is small, we already see a significant decrease of arterial wave reflection parameters, indicating peripheral arterial dilation. Our findings lay the groundwork for future studies in children with cardiovascular diseases. In addition, hypoxia testing under controlled conditions may help risk stratify children with selected conditions prior to "natural" high altitude exposure (like long distance flights or trips to higher mountain areas like the alps).

Keywords: hypoxia, arterial stiffness, vascular physiology, augmentation index

PP-420
Physical activity is the most important predictor of health related quality of life in adults with congenital heart disease - a swedish registry study

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Background and Aim: Traditional cardiovascular risk factors put patients with congenital heart disease (CHD) at increased risk for acquired cardiovascular disease and mortality – more so than patients without CHD. In the general population, health related quality of life (HRQoL) is associated with regular physical activity. It was the aim of this study to evaluate the most important predictors of HRQoL in adults with CHD (ACHD).
Method: This is a registry study using single center data collected between 2004-2022. Data include demographic data such as age and sex, body mass index (BMI) type of CHD, prior surgeries, physical activity and HRQoL using the EQ-5D-3L questionnaire. CHD severity was classified based on European Society of Cardiology (ESC) criteria. The cohort was divided based on self-reported levels of physical activity.
Results: A total of 2469 patients were included in this study. 1661 individuals had at least two visits. 878 (25.6%) patients had mild, 1151 (46.9%) moderate and 329 (13.3%) severe CHD. Patients who were not doing regular exercise were significantly older, were more likely to be female, had a higher BMI, and had a lower

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HRQoL than their physically active peers. ACHD category was not signifiantly different between the groups following Bonferroni correction. In a logistic regression model including covariates, physical activity was the most important predictor of a perfect HRQoL score in all five domains (mobility, self-care, usual activities, pain & discomfort, and anxiety & depression), especially if performed for >3h/week (Odds ratios (OR) 2.1 - 7.5, all p<0.001). Other important predictors were younger age (OR 0.99, p<0.001), male sex (OR 1.58, p<0.001), mild-moderate CHD (OR 1.59, p<0.001) and being underweight or obese (OR 1.44, p<0.001). Interestingly, underweight patients had similar HRQoL as obese patients.
Conclusions: Regular physical activity in ACHD patients is associated with better HRQoL. Patients with underweight and obesity alike are also at risk for impaired HRQoL. We suggest that ACHD follow-up visits should include counselling on life-style issues in order to enhance HRQoL and minimize modifiable risk factors for acquired cardiovascular disease.

Keywords: Physical activity, health related quality of life, adults with congenital heart disease, underweight, obesity, body mass index

Table 1

	No sports (n=1107)	<3h/week (n=765)	>=3h/week (n=448)	p(all)
DEMOGRAPHIC VARIABLES				
Age	38 (28-52)	34 (26-45)	29 (23-37)	<0.001
Female sex (%)	583 (52.6%)	364 (47.6%)	171 (38.2%)	<0.001
BMI	25.4 (22.2-29.1)	24.3 (21.8-27.4)	24.0 (21.9-26.2)	<0.001
BMI Category				
- Underweight	48 (4.3%)	35 (4.6%)	15 (3.3%)	<0.001
- Normal weight	475 (42.9%)	392 (51.2%)	265 (59.2%)	
- Overweight	342 (30.9%)	240 (31.4%)	140 (31.3%)	
- Obese	242 (21.9%)	98 (12.8%)	28 (6.3%)	
ACHD Category				0.026
- Mild	378 (34.1%)	292 (38.2%)	181 (40.4%)	
- Moderate	555 (50.1%)	351 (45.9%)	217 (48.4%)	
- Severe	174 (15.7%)	122 (15.9%)	50 (11.2%)	
EQ5D: No problems with				
- Mobility	857 (77.8%)	694 (90.8%)	435 (97.5%)	<0.001
- Self-care	1012 (92.2%)	730 (95.5%)	443 (99.1%)	<0.001
- Usual activities	863 (78.5%)	659 (86.3%)	420 (94.0%)	<0.001
- Pain & discomfort	649 (59.3%)	533 (70.1%)	363 (81.6%)	<0.001
- Anxiety & depression	661 (60.3%)	514 (67.6%)	348 (79.1%)	<0.001
TTO-score	.914 (.848-.969)	.969 (.880-.969)	.969 (.914-.969)	<0.001

Comparison of patient characteristics based on reported physical activity levels

PP-421
Validity of smartwatch derived ekg intervals in children compared to standard 12 lead EKGS

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Background and Aim: EKGs obtained by smartwatches have gained entrance into the clinical arena e.g. for detection of atrial fibrillation, but may also be useful for detecting arrhythmias such as supraventricular tachycardias. Smartwatches also provide measurements of EKG intervals. This could be useful when monitoring patients taking QTc prolonging medications. It was the aim of this study to determine the validity of smartwatch derived EKG intervals in children.
Method: Prospective cohort study at a pediatric cardiology outpatient clinic at a national tertiary care center. Smartwatch EKG (Withings ScanWatch) and 12-lead EKG were obtained simultaneously in the supine position. The automated measurements were documented and compared to manual measurements for both EKG modalities. Intraclass correlation coefficients (ICC) were determined. An ICC >0.9 was defined as excellent, 0.75-0.89 as good and 0.5-0.74 as moderate reliability. The study was approved by the national ethics committee. Written informed consent was obtained in all cases.
Results: 100 consecutive children (54 males, 46 females) with and without congenital heart disease were included. Median age 13 years (range 5-18 years). For automated smartwatch compared to automated 12-lead EKG measurements ICC was excellent for heart rate (ICC 0.97, p<0.001), good for PR-intervals (ICC 0.86, p<0.001), and moderate for QRS duration (ICC 0.7, p<0.001) and QTc (ICC 0.53, p<0.001). When using manual measurements of smartwatch EKGs, reliability was improved for PR interval (ICC 0.93, p<0.001), QRS duration (ICC 0.92, p<0.001), and QTc duration (ICC 0.84, p<0.001). For 12-lead EKGs, reliability of automated compared to manual measurements was excellent for all parameters (ICC >=0.9, p<0.001).
Conclusions: Automated smartwatch measurements are most reliable for determination of heart rate, which may be useful for detecting supraventricular tachycardias. Validity of automated smartwatch measurements is also good from PR-intervals, but is clinically less relevant. Automated determination of QTc by smartwatch appears least reliable, but validity can be improved when measuring QTc manually.

Keywords: smartwatch, EKG, QTc, validity

PP-422
Digital-platform guided home-based exercise training programme in patients with congenital heart disease (CHD): Impact randomized controlled trial

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Background and Aim: Home-based exercise training is a promising alternative to conventional supervised training for patients with congenital heart disease (CHD), but knowledge concerning variety and utility of home-based programmes is still lacking.
Main objective of this study was to ascertain if a structured digital-platform guided cardiac rehabilitation (CR) program can improve the fitness of young children with CHD. Main end point was an increase in peak oxygen uptake (VO2p) following the intervention.
Method: IMPACT-trial is a multicentre, randomised, controlled study, aiming to assess the impact of a digital-platform guided home-based cardiac rehabilitation program on the exercise capacity of children and adolescents (7 to 18 years old) with CHD.
Patients with a peak oxygen uptake (VO2max) <80% and a history of CHD sufficiently relevant to imply a restriction of recreational or school sport have been elected. Patients were randomised into 2 groups: intervention group (A) followed a 12-week

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digital-platform guided cardiac rehabilitation program and control group (B) carried out usual physical activity. Following randomisation, all patients underwent standardised cardiopulmonary exercise test (CPET), as recommended by the European Society of Cardiology (ESC). CPETs were repeated after 12 weeks and main variables before and after the intervention period were compared.
Results: Between june and october 2023, 26 patients were recruited, 16 in group A (12 boys, 75%) and 11 in group B (6 boys, 60%). Both groups were homogeneous in terms of age (group A median 13,9±2,5 years vs B, 13,6±2,6; p=0,792) and body mass index (group A median 20 kg±5,9 kg/m2 vs 22,8±5,2 group B; p=0,4). 2 patients dropped out the study (7%). At baseline, both groups had homogeneous pVO2 (group A median 1550±629ml vs group B 1605±632ml; p=0,792), but after the intervention period, group A improved significantly pVO2 from 1550± ml to 1690±715ml; p=0,011). There were no significant differences between groups at baseline or after intervention in oxygen pulse rate (OP), ventilatory anaerobic threshold (VAT) oxygen uptake, ventilatory efficiency (VE/CO2p) or oxygen uptake efficiency slope.
Conclusions: Digital-platform guided home-based exercise interventions is a useful alternative to supervised cardiac rehabilitation for all age groups of patients with CHD. A structured intervention programme can significantly increase VO2p.

Keywords: Cardiac Rehabilitation, Digital-platform guided exercise programme

PP-423

Impact of a digital-platform guided home-based exercise training programme on quality of life of patients with congenital heart disease (CHD)

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Background and Aim: Home-based exercise training is a promising alternative to conventional supervised training for patients with congenital heart disease (CHD), but knowledge concerning the impact on quality of life is still lacking.
We aim to assess changes in quality of life after completing a digital-platform guided home-based exercise training programme in children with CHD.
Method: IMPACT (Initiative for Multicentric Pediatric Cardiopulmonary Telerrehabilitation) trial is a multicentre, randomised, controlled study, aiming to assess the impact of a digital-platform guided home-based cardiac rehabilitation program on the quality of life of children with CHD. Elected patients were randomised into 2 groups: intervention group (A) followed a 12-week digital-platform guided cardiac rehabilitation program and control group (B) carried out usual physical activity. Following randomisation, all patients completed a self-reported quality of life score (PedsQL generic pediatric quality of life questionnaire) before and after the 12 weeks of follow-up. Results in the evaluated four multidimentional scales with a total of 23 items (in physical, emotional, social and school functionning) were compared. Each item used a 5-point scale from 0 (never) to 4 (almost always), higher scores indicating worse quality of life (maximum score: 92 points).
Results: Between June and October 2023, 32 patients were recruited, 16 in group A (12 boys, 75%; median age 13,9±4 years) and 16 in group B (8 boys, 50%; median age 13,4±3,4 years). 1 (3%) patient did not complete any test and 6 (19%) patients did not complete final test. At baseline, both groups had global homogeneous results (group A median 19±20 points vs B, 19,5±24 points; p=0,59), with a slight improvement at the end of the follow-up (group A median 9,5±21 points vs B, 13±3,1 points; p=0,926). But regarding physical scale, there was a significant improvement in group A in experienced running difficulties (baseline median 2±2 points vs end median 0±1 point, p=0,042) and sports participation impediments (baseline median 1±2 points vs end median 0±0 points, p=0,036).
Conclusions: Digital-platform guided home-based exercise interventions is a useful alternative to supervised cardiac rehabilitation for young patients with CHD. A structured intervention programme can improve quality of life.

Keywords: Congenital Heart Disease, exercise programme, fitness, quality of life

PP-424

Children with repaired tetralogy of fallot have lower overall daily physical activity than healthy peers

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Background and Aim: Children with repaired Tetralogy of Fallot (rToF) show decreased exercise capacity already early in childhood. Current treatment for associate pulmonary regurgitation consists of pulmonary valve replacement (PVR), but timing of PVR is subject of debate. Cardiopulmonary exercise testing (CPET) plays an important role in clinical decision making, but may not be an optimal measure for exercise tolerance in childhood. A measure of interest for clinical decision making is daily physical activity (DPA), but DPA patterns of children with rToF and its implications remain unknown. Therefore, we aimed to determine DPA in children with rToF and its main determinants.
Method: Children with rToF wore a tri-axial accelerometer (Actigraph) for one week. Counts (accelerations at waist level) per minute (CPM) were measured and expressed as CPM per axis. Y-axis CPM were used to classify physical activity intensity, including sedentary, light, moderate, vigorous and moderate to vigorous physical activity (MVPA) using cutpoints of Evenson et al. Z-scores of y-axis CPM and percentage in MVPA were calculated. DPA was compared to healthy references and correlated with VO2peak, grip strength, right ventricular (RV) function measured with MRI, and quality of life (QoL; PedsQL 4.0 Generic Core).
Results: 42 children with rToF and median age 13.1 (11.1, 16.5) years followed between 2016 and 2019 at the UMCG participated. CPM (y-axis) z-score of rToF patients was significantly

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lower than reference z-score 0 (p=0.003), whereas MVPA z-score was not (p=0.192). Participants spend a median time of 34 minutes per day in MVPA. Only 8 subjects (19%) met the WHO DPA advice of ≥ 60 minutes MVPA per day. In this study we could not demonstrate any correlation between DPA and VO2peak, grip strength, RV-function or QoL that was independent of age and sex.
Conclusions: Children with rToF from a contemporary cohort appear to have significantly lower overall daily physical activity than healthy peers, and only few meet WHO advice for daily physical activity. This could not be explained by VO2peak, grip strength or RV-function and was not related to QoL. Reasons for this reduced physical activity (somatic, psychosocial or others) should be further explored.

Keywords: repaired Tetralogy of Fallot, Accelerometry, Daily Physical Activity

PP-425

Use of cardiopulmonary exercise test for exercise prescription in young patients after fontan operation

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Background and Aim: Children and adolescents with congenital heart disease (CHD) should be encouraged to adopt a physically active lifestyle, but in many cases they need specific and individualized exercise programs. Cardiopulmonary exercise testing (CPET) could be a very useful tool in this context. The aim of the study is to investigate the use of CPET in a selected population of young people with univentricular heart after Fontan surgery.
Method: All Fontan pediatric patients (<18 years old) followed at the cardiology department of the Bambino Gesù Children's Hospital were evaluated for enrollment in the study. After clinical examination and echocardiogram, all stable and compliant patients underwent CPET on a cycloergometer (ramp protocol) or treadmill (Bruce ramp protocol) depending on cycling ability. Demographic, surgical, clinical, ECG and CPET data were collected.
Results: 91 Fontan patients aged 14.6 ± 2.8 years were enrolled (52 males), 44 (48.4%) with systemic right ventricle (26 males) and 47 (51.6%) with systemic left ventricle (26 males). 54/91 (59.3%) cases underwent a cycle ergometer test (32 males) and 37/91 (40.7%) cases a treadmill test (20 males). Peak oxygen uptake (pVO2) and VO2 at anaerobic threshold (VO2-AT) were significantly lower at the cycle ergometer in female (21.3±3.1 vs 24.8±7.1, p=0.001 and 15.8±2.1 vs 19.5±2.7 ml/min/kg, p<0.01, respectively) patients, whilst the significance was reached only for VO2-AT in male subjects (pVO2 25.4±4.6 vs 27.8±6.7 ml/min/kg, p=0.144; VO2-AT 17.4±3.8 vs 23.1±6.1 ml/min/kg, p<0.01). In contrast, only 27% (10/37) tested at the treadmill were able to achieve a maximal test (RQ> 1.05) compared with 96% (51/54) tested at the cycle ergometer (p<0.0001).
Conclusions: Many parameters of CPET may differ depending on how the test is performed. In particular, the anaerobic threshold appears significantly earlier at the cycle ergometer than at the treadmill. These should be taken into account when exercise is to be prescribed.

Keywords: Fontan operation, cardiopulmonary exercise testing (CPET), exercise prescription

PP-426

In fontan patients improvement of physical performance during 6-month exercise program depends on systemic ventricle morphology

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Background and Aim: In surgically palliated single ventricle Fontan patients (FP) transpulmonary flow and ventricular preload are dependent on systemic venous pressure and pulmonary vascular resistance. Reduced cardiopulmonary function (CPF) is a common outcome of single ventricular hemodynamics even in Fontan patients with "ideal cardiovascular conditions". Enhancing physical performance with an individualized exercise program improves FP quality of life and prognosis. We studied whether a 6-month exercise program improves CPF in Fontan patients and if the response would be dependent on ventricular morphology.
Method: 20 pediatric stable Fontan patients with right (FP-RV all with HLHS, n12) or left dominant ventricle morphology (FP-LV, n8), and normal peripheral oxygen saturation were included. A questionnaire on daily physical activity was followed by tests for CPF, body composition and muscle strength. Each patient received an exercise program with daily step goal and home workout focusing on lower limb strength. Following the 6-month period of self-guided training, patients repeated tests. In final analysis patients with FP-RV were divided according to coronary flow being antegrade (FP-RVa, n=6) and retrograde (FP-RVr, n=6). Statistical differences between groups were determined using one-way ANOVA and 6 month's responses within groups were tested with Mann-Whitney U test.
Results: (Table1.) All groups met their exercise goals. FP-LV were superior in CPF-testing and improved anaerobic threshold. All FPs had restrictive lungs, most severely in FP-RV. FP-LV and FP-RVa demonstrated significant improvements in their squat performance, with FP-L patients being the only group to enhance their standing broad jump and anaerobic threshold.
Conclusions: FP with systemic LV demonstrated superior response for tailored exercise program. FP with left-sided atresias (HLHS) reported the least weekly hours of voluntary physical activity and demonstrated poorer response to exercise. In addition, these Fontan patients demonstrated most severe lung restriction.

Keywords: Fontan circulation, Preventive, Exercise prescription, Ventricular morphology

Table 1

Baseline					Post			
	FP-RVa (n6)	FP-RVr (n6)	FP-LV (n8)		FP-RVa (n6)	FP-RVr (n6)	FP-LV (n8)	
CPX				**ANOVA**				**ANOVA U-test**
VO2max	29.1 ± 0.3	23.9 ± 6	32.5 ± 6	**0.043**	30 ± 6	23.9 ± 7	34.7 ± 7	**0.031**
AT	19.9 ± 4	17.1 ± 5	21.5 ± 4*	0.196	20.3 ± 5	16.5 ± 4	24.8 ± 6*	**0.029 *0.016**
MaxW	117.8 ± 23	96.3 ± 30	129 ± 45	0.249	139 ± 30	100 ± 40	144 ± 43	0.109
SPIROMETRY								
FVC(Z)	-3.55 ± 1.3	-3 ± 1	-1.52 ± 1	**0.007**	-3.3 ± 1.1	-2,5 ± 0,3	-1,6 ± 1.1	**0.033**
FEV1(Z)	-3.92 ± 2.8	-3.2 ± 1.2	-1.2 ± 1.1	**0.033**	-3.6 ± 2	-2.6 ± 0.8	-1.7 ± 1.6	0.172
EUROFIT								
Squats (30s)	17.8 ± 3.8*	21.2 ± 4.3	20.6 ± 5**	0.381	22.7 ± 2.5*	23,1 ± 4	25,5 ± 4**	0.319 ***0.0002** ****0.0279**
Standing broad jump	139 ± 30	137 ± 21	166 ± 34	0.157	153 ± 31	138.3 ± 29	173.9 ± 30	**0.121**
PHYSICAL ACTIVITY								
Strenuous excercise/week (h)	2.7 ± 1	1.5 ± 2	3.6 ± 1.5	**0.043**				
Daily steps					11443 ± 1839	9092 ± 3365	11729 ± 4304	**0.348**

*VO2max=Maximal oxygen uptake, AT=Anaerobic threshold, MaxW= Maximum workload, FVC= Forced vital capacity, FEV1=Forced expiratory volume in one second

Results from physical activity questionnaire, spiroergometria test, EUROFIT test and self guided exercise program.

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PP-427

Sports practice in severe congenital heart disease – why bother?

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Background and Aim: Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart disease. Pulmonary atresia (PA) with ventricular septal defect (VSD) is considered the extreme end of its anatomic spectrum.
The increasing long-term survival in both diseases justifies a detailed assessment of these patients' quality of life (QoL), and its promotion. We assessed parental perception of QoL in these children, and possible determining factors.
Method: Cross-sectional observational study, using an online survey for parents of children with TOF and PA with VSD (5-18 years old), as well as echocardiographic assessment of right ventricular function and dilatation. The survey consisted of an original questionnaire including the Pediatric Quality of Life Inventory 4.0 (parental version), sports practice, sleep pattern and social assessment, applied from March to July 2023. Variables associated with QoL were determined using linear regression, controlling for the severity of disease, using the diagnosis of TOF vs PA with VSD. Stata v17 was used for statistical analysis, and significance defined as $p<0.05$.
Results: 51 questionnaires were obtained, out of 74, on children with 10±3 years. 35 of these children were submitted to surgical correction of TOF, 12 to surgical correction of PA with VSD, and 4 had a palliation procedure for PA with VSD. The regular practise of sports was related to higher physical QoL (78.6% vs 64.8%, $p=0.023$) and higher psychosocial QoL (78.2% vs 67%, $p=0.016$). When controlling for the diagnosis, the practise of sports was still significant to the psychosocial component of QoL ($p=0.028$). When applied to the physical component of QoL, the practise of sports was only significant for patients with PA with VSD ($p=0.008$ vs $p=0.496$ in those with classic TOF). Factors such as age, body mass index, number of surgeries, sleep pattern, social class, and echocardiographic assessment of TAPSE and tricuspid/mitral valve ratio were not correlated with QoL.
Conclusions: Sports activity increases psychological and physical QoL in children with TOF. This work justifies the promotion of physical activity in these patients and more importantly, the prescription of safe exercise plans in those with more severe diseases.

Keywords: tetrallogy of fallot, pulmonary atresia with ventricular septal defect, exercise, sports cardiology, quality of life

Cardiac Imaging

PP-428

Accelerated cardiac magnetic resonance imaging using deep learning for volumetric assessment in children

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Background and Aim: To assess the diagnostic performance of accelerated 3RR cine MRI sequences using deep learning (DL) reconstruction compared with standard two-dimensional (2D) cine balanced steady-state free precession (bSSFP) sequences for cardiac volumetry.
Method: Twenty-nine consecutive patients (11±5 years) with biventricular physiology undergoing clinical cardiac MRI were scanned with a standard breath-holding short axis cine bSSFP and a free-breathing accelerated DL cine 3RR acquisition on a 1.5 T scanner. Biventricular volumetry was performed (semi-) automatically in both datasets retrospectively by an experienced radiologist and a medical student using a cloud-based postprocessing software. Two experienced readers visually assessed image quality of both sequences including a mesh surface rendering from semi-automatic segmentation using a four-point grading scale. Scan times and image quality were compared using a Wilcoxon rank sum test. Volumetrics were assessed with linear regression and Bland-Altman analysis. Measurement agreement was assessed with intraclass correlation coefficient (ICC).
Results: Mean acquisition time for a short axis stack was significantly shorter for the DL cine sequence 45.51±13.75s compared with 218.32±44.84s for the 2D cine sequence ($p<0.001$). No significant differences in biventricular volumetric analysis were found except for increased left ventricular (LV) mass in the DL cine compared with the 2D cine sequence (71.35±33.07g vs. 69.88±32.47g; $p<0.05$). All volumetric measurements had excellent agreement with ICC>0.9 except for ejection fraction (LVEF=0.81, RVEF=0.73). Image quality did not differ for mesh surface evaluation ($p=0.77$), but was decreased for end-diastolic and end-systolic contours, papillary muscles, and valve depiction in the DL cine images ($p<0.05$).
Conclusions: Volumetric results obtained from the DL cine sequence did not differ significantly from standard 2D cine sequences except for an overestimation of LV mass with cine DL. Overall image quality rating of DL cine images was inferior to conventional cine images. DL cine may enable more patient comfort and decrease the need for anaesthesia by the significant reduction of scan time.

Keywords: Cardiac magnetic resonance, volumetry, deep learning

Cardiac Dysrhythmias and Electrophysiology

PP-429

Wireless pacemaker in pediatric heart transplant

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Background and Aim: Paroxysmal AV block in cardiac transplantation not associated with rejection is a rare arrhythmia but requires pacemaker implantation. The main aim of this case is to show the

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utility of wireless pacemaker implantation in these patients who require permeable vascular access and biopsies during follow-up.
Method: We have searched and analyzed the patient's clinical history after receiving her consent.
Results: We present a 13-year-old adolescent girl who underwent cardiac retransplantation due to severe graft vascular disease secondary to graft rejection. She was receiving immunosuppressive treatment with tacrolimus, mycophenolate and urbason. Eight months after retransplantation, she reported episodes of dizziness and instability. A cardiac biopsy was performed with no evidence of rejection and normal coronary angiography. During electrocardiographic monitoring using a Holter T-shirt (Lanubo) for a week, a predominance of sinus rhythm at 80 bpm was observed, but she presented episodes of paroxysmal complete AV block with an escape rate of less than 40 bpm associated with the aforementioned symptoms of dizziness, which led to the decision to implant a pacemaker. Due to the patient's medical conditions (third transplant, thrombosis of multiple venous accesses and the need for cardiac biopsies during follow-up) and the paroxysmal block, it was decided to implant a Medtronic Micra model wireless pacemaker in the RV apex and programmed in VVI with a minimum pacing rate of 60 bpm. The implantation technique was by right femoral vein access through a 27F sheath (patient weight 26 kg). In the subsequent follow-up, for more than 3 years, she has not presented any episodes of dizziness, remains asymptomatic with isolated monomorphic ventricular extrasystoles and maintains her own sinus rhythm with sporadic pacemaker stimulation (2.5% of the time in the revisions).
Conclusions: Paroxysmal AV block in cardiac transplantation not associated with rejection is a rare arrhythmia but requires pacemaker implantation. We believe that wireless pacemaker is a good option in these patients who require permeable vascular access and biopsies during follow-up.

Keywords: wireless pacemaker, heart transplant, paroxysmal AV block, rejection

Adult Congenital Heart Disease

PP-430
Is survival of repaired congenital systemic-to-pulmonary shunt in adults with borderline pulmonary vascular resistant better than eisenmenger syndrome

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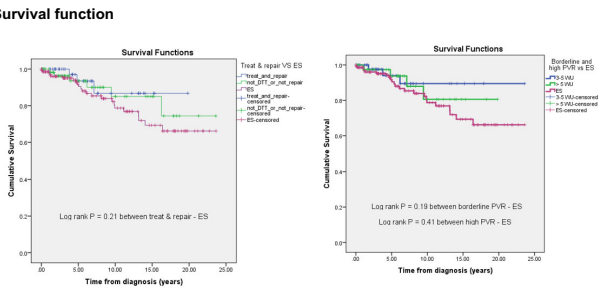
Background and Aim: Pulmonary arterial hypertension (PAH) associated with congenital heart disease (CHD) (PAH-CHD) consequent morbidity in children and adults. Studies of patients with borderline pulmonary vascular resistant (PVR) in particular who received treat-and-repair strategy compared to Eisenmenger syndrome (ES) is limited. We aimed to assess survival of repaired congenital systemic-to-pulmonary shunt in adults with borderline-high PVR compared to ES.
Method: All 495 prevalent cases of PAH-CHD with hemodynamic confirmation by cardiac catheterization in 1995-2021 were retrospectively reviewed. Patients with clinical classification 1 (ES) and 2 (predominantly left-to-right shunt) were selected. Patients in group 2 who were younger than 18 years old, or with single ventricle were excluded. We classified these patients into low PVR (< 3 WU), borderline PVR (3-5 WU) and high PVR (> 5WU). Baseline characteristics were retrieved. The survival analysis was performed at the end of 2022.
Results: A total 140 patients with ES [median age (IQR) of 26.2 (22.8) years] and 117 adults with prevalent systemic-to-pulmonary shunt [median age (IQR) of 36.4 (23.6) years] were analysed. In ES group, baseline mean pulmonary arterial pressure (PAP) and PVR were 74.7 + 16.7 mmHg and 26.6 + 14.5 WU. Of 117 adults with prevalent systemic-to-pulmonary shunt, mean PAP and PVR were 56.6 + 16.5 mmHg and 7.6 + 4.6 WU. There were 48 patients with borderline PVR and 46 patients with PVR high > 5 WU. Treat-and-repair was performed in 49 patients (34 total repair, 15 fenestrated closure). In the ES group, survival rates at 10, and 15 years were 78.7%, and 69.3%, respectively. In adults with borderline PVR, the survival rates at 10, and 15 years were 89.5%, and 89.5%, respectively and in high PVR were 80.6%, and 80.6%, respectively. Comparing to ES, survival of borderline PVR and high PVR were marginally superior to ES ($p = 0.19, 0.41$). Survival of patients with treat-and-repair strategy was slightly superior to ES ($p = 0.21$).
Conclusions: Survival of repaired systemic-to-pulmonary shunt in adults with borderline PVR tends to superior to ES, even no statistically significance. Treat-and-repair strategy and fenestrated closure possibly improve survival in selected cases. Further aggregated studies are required.

Keywords: Pulmonary arterial hypertension (PAH); congenital heart disease (CHD), targeted therapy pulmonary hypertension, Eisenmenger, prevalent systemic-to-pulmonary shunt



PP-431
Concurrent finding of severe mitral regurgitation of rheumatic aetiology and anomalous left coronary artery arising from the pulmonary artery (ALCAPA)

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Background and Aim: ALCAPA syndrome (anomalous left coronary artery arising from the pulmonary artery) is a rare coronary anomaly, with high mortality in the first year of life and risk of sudden cardiac death in adulthood. After birth, it causes left ventricular

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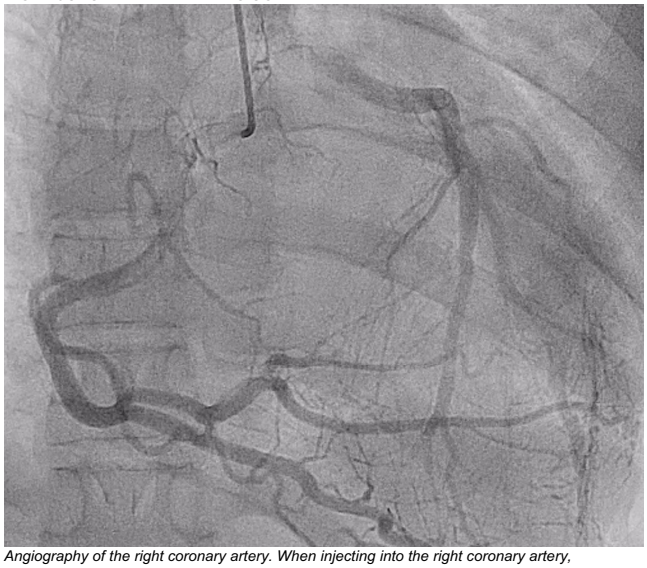
poor perfusion and can lead to silent ischaemia, ventricular dysfunction and malignant arrhythmias.
Method: We present the case of a 23-year-old woman, of Moroccan origin and with no medical history of interest, admitted to the emergency department for ventricular tachycardia (VT) at 220 bpm. Given the patient's haemodynamic instability, synchronised electrical cardioversion was performed and after restoring sinus rhythm, the patient was admitted to the intensive care unit for close monitoring and study.
Results: On admission, a transthoracic echocardiogram showed severe organic mitral insufficiency with significant restriction of the posterior mitral leaflet, thickened leaflets and fusion of the commissures, suggestive of rheumatic mitral valve disease, together with a dilated and moderately dysfunctional left ventricle, moderate functional tricuspid regurgitation and moderate pulmonary hypertension. A transesophageal echocardiogram was performed, which confirmed the diagnosis of rheumatic mitral valve disease. Before deciding on the optimal treatment for mitral valve disease, both right and left cardiac catheterisation is performed. The right catheterisation revealed moderate postcapillary pulmonary hypertension, with no elevated pulmonary arteriolar resistance. The left catheterisation revealed a healthy right coronary artery, with no left coronary artery arising from the aorta. Therefore, a cardiac CT scan was performed which showed the origin of the left coronary artery in the coronary trunk, confirming the diagnosis of ALCAPA (anomalous left coronary artery arising from the pulmonary artery). The study was completed with an MRI that showed a dilated left ventricle, slightly dysfunctional, with no evidence of late gadolinium enhancement.
Conclusions: With a diagnosis of ALCAPA syndrome and severe organic rheumatic mitral valve disease, the case was presented to the Heart Team. The case was accepted for mitral valve replacement and translocation of the left coronary artery to the aorta. Due to the patient's preferences and her gestational desire, a biological mitral prosthesis was chosen. The surgery was uneventful and the patient was discharged early, 10 days after surgery.

Keywords: ALCAPA, ventricular taquicardia, Rheumatic valvular disease

RIGHT CORONARY ARTERY ANGIOGRAPHY



Angiography of the right coronary artery. When injecting into the right coronary artery, retrograde filling of the left coronary artery with origin in the pulmonary trunk is observed.

PP-432

Clinical outcome of adult congenitally corrected transposition of the great arteries without surgical intervention during childhood

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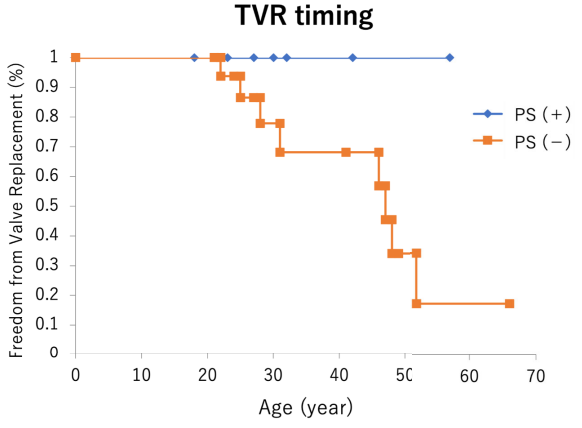
Background and Aim: Congenitally corrected transposition of the great arteries (ccTGA) is rare, accounting for less than 1% of all the congenital heart disease. The long-term prognosis is variable, though tricuspid regurgitation (TR), right systemic ventricular failure, and complete atrioventricular block (CAVB) may progress with age. The incidence of heart failure is reported around 25% by age 45 even in uncomplicated cases. We sought to assess the clinical course of adult patients without surgical intervention during childhood.
Method: We retrospectively reviewed 26 adult ccTGA patients without childhood surgical intervention. Symptoms, associated anomalies and surgical outcomes were collected from the medical charts.
Results: The current age included patients ranged from 18 to 70 years. Nine were diagnosed at birth, 6 in infancy, 6 in school age, and 5 in adulthood. Associated anomalies with ccTGA were ventricular septal defect (VSD) in 5, atrial septal defect (ASD) in 3, and pulmonary arterial stenosis (PS) in 8 cases. Tricuspid valve replacement (TVR) was performed in 9 patients without PS (22-61 years). The patients less than 30s (4/9) were performed TVR for newly recognized arrhythmia and worsened TR while morphological right ventricular function preserved. The main indication for patients elder than 40s (5/9) was their heart failure symptoms. In cases with PS, with or without VSD, TVR was not required, the severity of TR was less than moderate, and ventricular function was all preserved. Pacemaker implantation for CAVB was performed in 6 patients (15-48 years), and supraventricular tachycardia (SVT) was present in 8 patients (2-65 years), which was common with severe TR. The heart failure symptoms were observed in 8 patients (24-66 years), developed around age 40. Eighteen patients received medications (18/26), and patients over 30 were added beta-blockers with vasodilators.



Tricuspid valve replacement (TVR) was performed in 9 patients without PS (22-61 years).

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Conclusions: TR worsened less than 30s, and SVT developed mostly in severe TR cases. Although multiple medications were administered after 30s, heart failure occurred around age 40. In PS patients, even with VSD, the severity of TR was less severe compared to the patients without PS.

Keywords: congenital heart disease, congenitally corrected transposition of the great arteries, tricuspid regurgitation, pulmonary arterial stenosis.

PP-433

Lung function in single ventricle patients post fontan circulation potential role of diaphragm ultrasound imaging

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Background and Aim: Patients with Fontan circulation have reduced functional capacity due to either reduced cardiac output or abnormal lung function and respiratory muscle weakness. Ultrasound imaging (US) is a non-invasive tool to investigate the diaphragm muscle function.
Aims of the study were: 1) to evaluate lung function and functional capacity in a cohort of Fontan patients; 2) to evaluate static and dynamic diaphragmatic function in a subgroup of patients. ≤≤
Method: Consecutive patients with Fontan palliation who underwent lung spirometry and cardiopulmonary test (CPET) between January 2014 and October 2023 were included in the study. Surgical history, clinical and imaging data were collected from hospital records. A subgroup of patient underwent an US for diaphragm static and dynamic evaluation.
Results: 53 consecutive Fontan patients (age 25±12 years) were included in the study. The functionally single ventricle was morphologically left in almost half of our population (43.3%). Lung spirometry identified 52% of patients with a restrictive lung disease. Those patients, in comparison with the remaining population, presented a lower VO2 (L/min) and VO2/kg/min at anaerobic threshold (AT) (0.77±0.31 vs 1.1±0.34, p=0.009 and 12.4±4.4 vs 18.1±6.3, p=0.009, respectively); a reduced peak O2pulse (ml/min): 6.7 (IQ 5.9;8.7) vs 9.2 (IQ 8.1;10.6), p=0.03; and a reduced breathing reserve percentage (BR) (54±32% vs 87 ±29%, p=0.001). Moreover, patients with restrictive lung pattern were more often on diuretic therapy (Log OR:2,7 p≤0.001) and interestingly reported less physical activity (X2:10, p≤0.001). Diaphragm US performed in 10 patients (age 25±11 years) without documented diaphragm paralysis showed abnormal static or dynamic parameters in 80% of the population, for instance reduced right and left excursion at quiet breathing, respectively 50 and 60% and 20% presented reduced left and right max thickness. Moreover, right max thickness correlated with predicted TLC (r: 0.7, p=0.04).
Conclusions: In our post Fontan Palliation population study restrictive lung disease was associated with reduced function capacity, more frequent need of diuretic therapy and less practice of reported physical activity. Diaphragm US showed abnormal static and or dynamic parameters in this population. Further studies are needed to confirm our results and evaluate the impact of respiratory training on diaphragm function and therefore functional capacity.

Keywords: Fontan, lung function, exercise capacity, diaphragm

PP-434

Assessment of myocardial fibrosis in marfan syndrome using cardiac magnetic resonance imaging

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Background and Aim: Impaired myocardial function and arrhythmia are important manifestations in Marfan syndrome (MFS). Studies assessing myocardial fibrosis in relation to these manifestations are lacking.
Method: This cross-sectional, single-center study assessed ventricular volumes, ventricular function and myocardial fibrosis by cardiac magnetic resonance imaging (CMR) in patients with MFS harboring a (likely) pathogenic FBN1 variant. Presence and extent of fibrosis were assessed by late gadolinium enhancement (LGE) and extracellular volume measurement (ECV). Data on 24-hour Holter monitoring and clinical data were extracted from electronic patient records.
Results: The study included 32 unselected patients with MFS (median age 38 years [range 10-69], 41% women). No focal myocardial fibrosis was detected. Six patients (21%) had diffuse fibrosis (ECV>29%). No association was found between presence of diffuse fibrosis and myocardial disease. Five patients (16%) had reduced left ventricular ejection fraction (LVEF<55%). While all of these exhibited mitral annular disjunction (MAD), only two had ECV>29%. Patients with MAD had increased indexed LV volumes (median end-diastolic volume, 92 ml/m2 [IQR, 78-100] vs 78 ml/m2 [IQR, 71-87]; median end-systolic volume, 31 ml/m2 [IQR, 23-46] vs 22 ml/m2 [IQR, 21-28]), also after adjusting for presence of mitral and aortic valve regurgitation. No differences in ECV were seen between patients with and without MAD.
Conclusions: In this cohort of patients with MFS, focal myocardial fibrosis was not detected using CMR. Although diffuse fibrosis was observed in 21% of patients, no evident connection to myocardial disease was found. Future studies should evaluate the role of diffuse fibrosis in predicting outcomes.

Keywords: Marfan Syndrome, Myocardial Fibrosis, Cardiac Magnetic Resonance Imaging, Ventricular Function, Mitral Annular Disjunction

PP-435

Long-term outcome and risk factors for morbidity and mortality in ebstein´s anomaly: A comprehensive study of 398 cases over half a century

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Background and Aim: Ebstein´s Anomaly (EA) is a rare congenital heart disease with an incidence of 1 in 20,000. Throughout the last five decades, multiple surgical techniques for tricuspid valve (TV) surgery have been reported. Therefore, our study aims to provide a demographic overview and review the long-term outcomes, as well as risk factors for morbidity and mortality of patients with EA treated in Germany using data from a large registry.
Method: All patients with EA, listed in the National Register for Congenital Heart Defects (NRCHD) from 1973 until initial query (03/2021) were included for retrospective analysis. All available patient records were reviewed and relevant demographic, diagnostic, and clinical data transferred to a newly created database.
Results: 398 patients (230/58%female) met the inclusion criteria. At least one TV surgery was performed in 204 patients (51.3%) at a median age at initial surgery of 13.2 years (IQR:6-25Y.) The first surgery was a reconstruction in 83.3%, a replacement in 11.3%, and a surgical closure of TV in 5.4% of patients. Of those undergoing biventricular corrective surgery (TV reconstruction or replacement, n=193), 17 (8.8%) developed a postoperative persistent complete atrioventricular block (CAVB) complicating their first surgery. Valve replacement was associated with a 3.7-fold risk of CAVB compared to valve reconstruction (p=0.027). Each 10-year increase in age resulted in a 1.4-fold increase in risk of CAVB (p=0.021). 32-days mortality postoperatively was 1.6%. Late survival at 10, 20, and 30 years postoperatively was 98.1%,96.6% and 87.7% following TV reconstruction and 100%, 90.9% and 90.9% following TV replacement. In a multivariate analysis, patient age at initial surgery was predictive of an increased reoperation rate, with patients younger than 10 years requiring significantly more reoperations. Postoperative CAVB and cyanosis were associated with increased mortality. However, CAVB was not predictive of increased risk for reoperation.
Conclusions: The study confirms that TV surgery in EA can be performed with low perioperative mortality and excellent long-term survival. It revealed an unexpectedly high proportion of CAVB, which was identified as a significant risk factor for all-cause death. Risk factors for worse outcomes in terms of reoperation rate and mortality include young patient age, postoperative CAVB and cyanosis.

Keywords: Ebstein's anomaly, long-term outcome, morbidity, mortality, postoperative persistent complete atrioventricular block, registry-based study

PP-437

Endovascular management of aortic coarctation: About 3 cases

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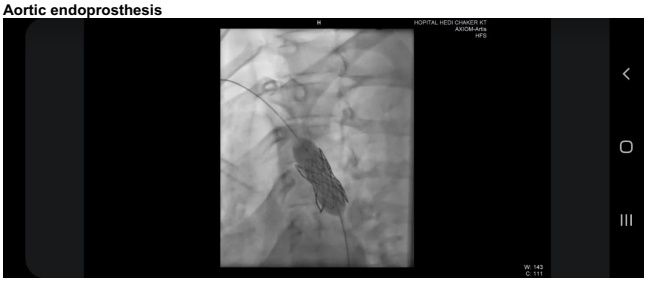
Cardiology Departement, University of Medecine, Sfax, Tunisia

Background and Aim: Coarctation of the aorta (CoA) is a relatively common congenital heart disease that can be diagnosed as a primary cause of hypertension in adolescents and adults. Historically, surgery was used to treat native CoA. However, transcatheter approaches have recently become more refined, in part due to increased operator experience, but also to improved balloon and stent technology, which has improved the safety and success of the CoA transcatheter approach.
Method: We present three cases of of severe aortic coarctation in males aged 26, 28, and 18. All of the patients had high blood pressure. One patient suffered from a hemorrhagic stroke.
Results: Two patients with recurrent CoA requiring endovascular aortic repair had undergone open repair when children. We used a transfemoral access after general anesthesia. Before treatment, we performed angiography and measured translesional pressure gradients and aortic diameters. In both cases of recoarctation, we proceeded by inflating a balloon to a low enough pressure to overcome the waist. In the third case, an endoprosthesis is placed just after the origin of the left subclavian artery, and angiography confirms its proper placement. A final angiography is performed to evaluate the outcome and rule out any complications. The residual gradient was under 20 mm Hg, which was deemed successful.
Conclusions: The main steps for a successful transcatheter CoA treatment are careful preprocedural planning, proper operator training, consolidated experience in endovascular procedures, and appropriate patient and prosthesis selection.

Keywords: coarctation, endovascular, hypertension

Aortic endoprosthesis



PP-440

Patient perspective on disease severity and palliative care: An outdated taboo in patient-centered medical care - interim results of an ongoing survey

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Background and Aim: Palliative care is still largely unknown to many CHD patients; it is associated with fears and taboos and is still far too rarely a natural part of a long-term strategic treatment concept.
Method: An online survey was conducted among adult patients with CHD from all over Germany. Patients were recruited via the National Register for Congenital Heart Defects and invited to participate by e-mail/letter.
Results: A total of 1115 CHD-patients have taken part in the survey so far (58.2% female), 41.8% were between 18 and 30 years old (38.5% 31-50 years, 19.7% >50 years). 18.4% had a simple, 49.7% a moderate and 31.9% a complex CHD. 55.2% stated that they were religious and 32.8% had a university degree as highest educational qualification. 30.8% stated that they considered their CHD to be

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mild and did not think that they would have any limitations in the course of their lives due to their condition. 50% rated their CHD as moderate and assume that they could have problems with their heart again at some point. Only 19.3% thought that their CHD was severe and that their heart would cause them problems and shorten their life expectancy. 63% had already considered what would happen if their health got worse, while 9.6% reported that they did not want to think about it. 31.7% are worried about having to rely on the help of others at an early stage, 39.3% have not yet thought about what medical care they would like to receive if they were to need assistance, and 64.7% said they were aware of the difference between a living will and a health care proxy. 28.8% have a living will and 27.1% have a health care proxy.
Conclusions: Patients assess the severity of their CHD much differently than is the case from a medical perspective. Palliative medicine, medical care in the case of deterioration, information about life expectancy, living wills and health care proxies should be a natural part of holistic medical care. This should be started at an early stage and not only when necessary, in order to ensure patient-centered and needs-oriented care at all times.

Keywords: CHD, online survey, palliative care, living wills, health care proxies

PP-441

Assessment of dyspnoea on effort in adult atrial septal defect patients by cardiopulmonary exercise testing in the six month follow up

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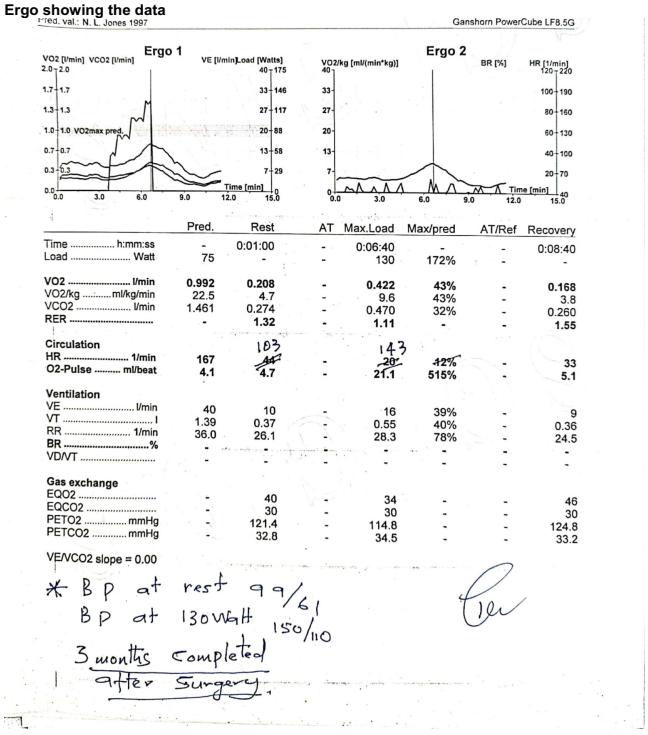
Background and Aim: The Atrial Septal Defects (ASD) are congenital cardiac disease that may also present in adults. The large defects can develop pulmonary hypertension increasing effort intolerance affecting the quality of life and may need surgical closure. Therefore, we aimed to assess the effort intolerance by subjecting them prospectively to cardiopulmonary exercise testing on a temporal scale for six months.
Method: Twenty adults patients who came for surgical ASD closure with the support of cardiopulmonary bypass at our institute consented to undergo cardiopulmonary exercise testing at their follow up at one, three and six months. The Bruce Protocol on a bicycle ergometer was used. The ECG and blood pressure were monitored noninvasively. The breath analysis was done by powercube using Ganshorn ergospirometry. They were requested for maximum effort and whenever they could not continue further the test was stopped. The data was presented as appropriate and was analysed using the SPSS software.
Results: Thirteen male and nine female patients completed follow up for six months after surgery. Median age was 30 yrs. with maximum 59 and minimum 18 years. Average height was 154±9.3 cms and weight was 47±8.9 kilograms. The maximum load on Bruce protocol attained was 92.5±4.6 before surgery and 79.75, 84.35, and 89.68 in the follow up and was significantly less than predicted for that age. The VO2 at rest before surgery was 0.23 L/min, and 0.14, 0.146, 0.168 and 0.187L/min in the follow up similarly VO2 attained at maximum load was 0.58 L/min whereas it was never near baseline level in post operative follow up with 0.28, 0.289, 0.36 l/min as significantly reduceed. There other static lung functions were also significantly reduced the respiratory quotient was also significantly reduced. All of them had sinus rhythm but have exertional dyspnea. The echocardiographic assessment was within normal limits.
Conclusions: This study demonstrates that after surgical ASD closure, the adult patients still show cardiovascular limitation to exercise even after six months in the follow up. The oxygen uptake, as well as the carbon dioxide elimination needs high ventilatory effort induced by exercise. This is likely the main factor stimulating dyspnoea and limiting exercise tolerance.

Keywords: Atrial Septal Defect, Adult Congenital heart disease, Surgery for ASD closure, Surgical followup in ASD, Effort intolerance in ASD, CPET in Adult ASD



PP-442

Maternal congenital and acquired heart disease in Guyana

Riasoya Jodah[1], Debra Isaac[2], Rana Hassan[2], Tegan Pierce[3], Michael Chin[4] and Elif Seda Selamet Tierney[5]
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Background and Aim: Guyana is a developing country where maternal mortality and morbidity (MMM) rates are significantly higher than the global average. Cardiac disease complicating pregnancy is a common cause of MMM in developing countries but remains poorly studied in Guyana. The Georgetown Public Hospital Corporation (GPHC) is Guyana's only public tertiary care center that manages most high-risk pregnancies. Since the establishment of the Guyana Echocardiography Education Program and Laboratory at GPHC, echocardiography has been available and utilized by obstetrical patients since 2012. The goal of this study is to determine the prevalence and distribution of maternal cardiac disease in Guyana.

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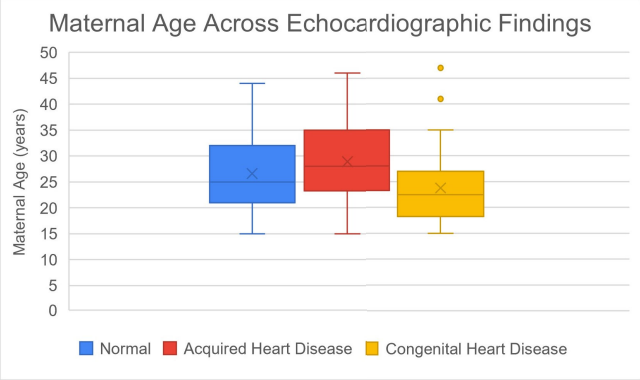
Method: This is an observational study featuring a retrospective cohort. All echocardiogram reports since the GPHC Echo Lab establishment in 2012 were screened to identify patients who were pregnant or six months postpartum. Using a standardized data extraction form, patient demographics, gestational and post-partum ages, echocardiogram indication and results were collected when available. Amongst abnormal reports, the condition was classified into acquired versus congenital heart disease and further subclassified into rheumatic heart disease, ischemic heart disease, cardiomyopathy, endocarditis, and other, and cyanotic versus acyanotic heart disease, respectively. Descriptive statistics were performed using ANOVA and Turkey HSD post-hoc tests when appropriate.
Results: A total of 3,436 echocardiogram reports were screened, and 341 echocardiograms from unique gestational or postpartum periods were identified across 334 patients. 127 (37.2%) of the 341 echocardiograms were abnormal. Amongst these abnormal echocardiograms, 92 (72.4%) showed acquired, and 36 (27.6%) showed congenital heart disease. Cardiomyopathy (n=42) was the largest cause of acquired heart disease, while ventricular septal defect (n=13) was the most common congenital heart disease. Patients with acquired cardiac conditions were significantly older than those with congenital heart disease (28.9 years vs 23.75 years; p=0.0005, Figure) and those with normal echocardiograms (28.9 years vs 26.6 years; p=0.02, Figure).
Conclusions: Our study results show that there is a high burden of cardiac disease amongst pregnant and postpartum women in Guyana. These findings highlight the need to examine the association of these findings with maternal and neonatal outcomes, as well as identify further risk factors, in order to improve the MMM rates in Guyana.

Keywords: maternal, echocardiogram, congenital, acquired

Distribution of Maternal Age Across Echocardiographic Findings



PP-443

sinus venosus asd and partial anomalous pulmonary vein return – rarely diagnosed in adulthood?

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Background and Aim: Sinus venosus type of atrial septal defect with partial anomalous pulmonary vein return (PAPVR) is a congenital heart defect that often leads to symptoms in the first years of life. The occurrence of symptoms depends on the extent of the left-right shunting. If the right ventricle is dilated, surgery is indicated. However, sometimes the diagnosis is not made until adulthood when physical performance declines.
Method: We reviewed medical records of our GUCH patients (grown-ups with congenital heart defects) with newly diagnosed sinus venosus defect and PAPVR in 2023. Baseline characteristics as well as operative data were analyzed retrospectively.
Results: Within 9 months, 5 adult patients (female: n=4) with newly diagnosed sinus venosus defect and PAPVR presented to our clinic. Mean age at presentation was 33.6 ± 10.7 years. All patients reported more than one hospitalization prior to diagnosis. 4 out of 5 patients were in NYHA class 2 or 3. Mean Qp:Qs was 1.9±0.2. Corrective surgery via partial upper sternotomy was performed in 3 cases. Full sternotomy was performed in one patient with persistent left superior vena cava and in one re-do procedure. Mean cardiopulmonary bypass time and duration of aortic cross clamping were 164 ± 61 minutes and 84 ± 20 minutes, respectively. The intra-atrial tunnels were created with bovine pericardium. A patch-plasty of the vena cava was also performed. Mean length of stay in the intensive care unit was 3 ± 2.3 days. One patient underwent thoracic surgery due to bleeding, one needed repetitive cardioversion procedures. All patients were discharged home after 12.4 ± 4.5 days in good clinical condition with sinus rhythm.
Conclusions: Sinus venosus defect and PAPVR is not that rare and should be considered as a differential diagnosis if there is no other explanation for a decline of the physical performance otherwise healthy young adults. Corrective surgery can be performed with good clinical results.

Keywords: Sinus venöses defect, partial anomalous pulmonary vein return

PP-444

A very rare case of bilateral partial anomalous pulmonary venous connection in an adult

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Background and Aim: Partial anomalous pulmonary venous connection is a condition in which some, but not all, of pulmonary veins connect to the right atrium or its tributaries, rather than to the left atrium. This rare condition more frequently occurs unilaterally, in the right or left pulmonary veins. The occurrence of bilateral partial anomalous connections is a very rare anatomic finding. The patient was diagnosed only for large atrial septal defect but in theatre was noticed that only the right superior pulmonary vein drains into the left atrium, while the right inferior pulmonary vein and all left pulmonary veins drain into a venous collector, localized extra pericardial in the left side, which drained through a large vertical vein to the innominate vein. To our knowledge, there is only one reported case that is similar to our case. We corrected the anomaly successfully and the patient did a good postoperative course.
Method: Case presentation
Results: Case presentation
Conclusions: The team work remains the key success in unpredicted scenarios

Keywords: Partial anomalous pulmonary venous drainage, congenital heart disease, adult atrial septal defect

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PP-445

Renal perfusion changes in aortic coarctation patients: Repaco trial

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Background and Aim: Successful surgical or percutaneous aortic coarctation (CoA) treatment at early age do no guarantees lifelong effectiveness. Up to 50% of patients with repaired CoA have persistent hypertension. Eventually CoA may lead to long standing renal perfusion disorder and may impact residual hypertension. In this pilot study we try to evaluate renal perfusion changes and relation to the severity of CoA.
Method: 78 patients with CoA underwent CT scan and were distributed in 2 groups. Patients with significant reCoA or native CoA had isthmus/AoD index <0.7 and Group 2 had isthmus/AoD index >0.7. The 99mTC-MAG3 captopril scintigraphy was performed for both patients groups. Time to peak, time to half-peak, peak to half-peak, 30 min/peak and 20 min/3 min count ratios were determined for whole-kidney ROIs. Five patients with significant native aortic coarctation or re-coarctation were treated with percutaneous stent implantation with repeated tests 3-6 moths after procedure.
Results: Time to peak after captopril test for Group 1 was shorter compared with Group 2 in both kidneys (p>0,05). The median time to ½ peak was significantly lower in Group 2 versus Group 1, which shows faster parenchymal perfusion in Group 2.
Renal scintigraphy in 2/5 stented patients showed that time to peak after captopril test was elevated in both kidneys after stenting compared to basic measurements and 1/5 patients time to ½ peak did not shorten, which suggests renal parenchymal dysfunction. Conversely, ACE inhibitor scintigram show decrease in 30 min/peak uptake ratio in all patients after stenting of coarctation.
Conclusions: For patients with significant CoA faster parenchymal perfusion and prolongation of time to ½ peak after captopril test aware about possibility to have renal hypoperfusion. Furthermore, renal perfusion scan shows promising data regarding effectiveness of percutaneous CoA treatment with improved renal perfusion and function. However, in some patients renal parenchyma may not recover after gradient reduction at isthmus level due to long-standing renal hypoperfusion.

Keywords: aortic coarctation, renal perfusion, residual hypertension, stenting

PP-446

Pulse wave velocity predicts future aortic dissection in patients with marfan syndrome

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Background and Aim: Aortic root dilatation is an established cardiovascular manifestation in Marfan syndrome (MFS). The ensuing aortic dissection (AoD) is the most important cause of mortality in these patients. Prophylactic aortic root replacement (AoRR) has been the mainstay in preventing AoD. To date, aortic root diameters, growth rate and family history of AoD are the main criteria used for determining the timing of prophylactic AoRR. However, some patients present with AoD before reaching surgical threshold.
Method: We conducted a prospective study including 45 patients with MFS (55,6% women, 34,5±11,6yr). Patients underwent a magnetic resonance angiography (MRA) including measurements of the aortic pulse wave velocity (PWV). To evaluate whether PWV could accurately predict AoD, patients were followed longitudinally (mean follow-up 6,96±1,46yr). For analysis, patients were divided in two groups based on whether they suffered AoD (N=6) or not. An ROC-curve analysis was performed to find an accurate PWV cut-off.
Results: As previously described, higher PWV was significantly associated with increasing age (p<0,001) and aortic root diameters (p=0,015), as well as hypertension (p=0,009). During follow-up six patients (13,3%) presented with AoD (1 with type A and 5 with type B). AoD occurred after 3,59±3,12 years follow-up. Because AoRR is known to impact PWV, patients who underwent previous surgery (N=8) were excluded from further analyses. Compared to patients without AoD during follow-up, patients with AoD had a significantly higher PWV (4,78 (IQR 4,24-5,36) versus 7,68 (IQR 6,35-8,73); p=0,001). This finding persisted independently of age and aortic root diameter. Using ROC-analysis, a PWV above 5,84m/s could predict follow-up AoD with a sensitivity of 100% and specificity of 90,3% (AUC of 0,952).
Conclusions: Increased PWV has been described in patients with MFS and has been shown to correlate with aortic growth in these patients. To our knowledge, PWV has never been shown to predict AoD in patients with MFS. Our study shows that increased PWV may be considered as a risk factor for AoD and could be a useful addition to the existing thresholds for prophylactic AoRR. Further studies with larger study populations are needed to confirm these results and specify PWV cut-offs more precisely.

Keywords: Marfan syndrome, FBN1, Aortic dissection, Pulse wave velocity

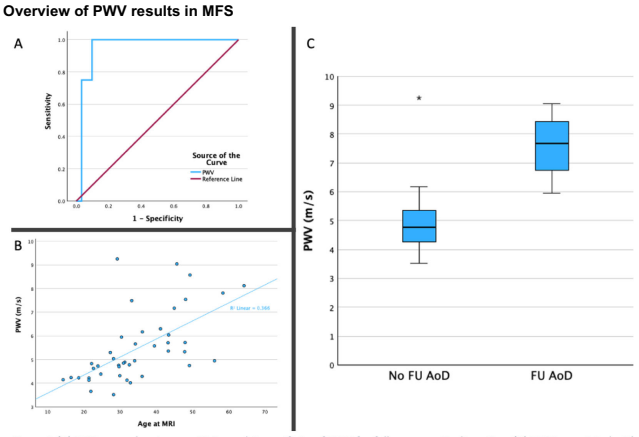

Figure 1 (A) ROC-curve showing sensitivity and 1-specificity of PWV for follow-up aortic dissection. (B) PWV associated with the age of patients at the time of MRI. (C) PWV compared between patients with (FU AoD) and without follow-up aortic dissection (No FU AoD) - excluding patients with an AoRR at baseline.

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PP-447

E-selectin and asymmetric dimethylarginine plasma levels in adult cyanotic congenital heart disease patients: clinical status and vascular function

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Background and Aim: Patients with cyanosis secondary to congenital heart disease (CHD) are characterized by erythrocytosis and increased blood viscosity, which contributes to endothelial dysfunction, increased arterial stiffness, and impaired vascular function, which may affect the final clinical presentation. Asymmetric dimethylarginine (ADMA) and e-selectin (e-sel) are valuable biomarkers for prognosis of vascular damage. Measuring biomarkers of endothelial dysfunction has the potential to be a rapid and easily accessible tool that reflects the severity of the disease. We aimed to assess e-sel and ADMA levels, their relationship with clinical status, as well as endothelial and vascular function.
Method: A cross-sectional study including 36 adult CHD cyanotic patients [(17 males) (42.3 +/- 16.3 years)] with arterial blood oxygen saturation less than 92% and 20 healthy controls [(10 males) (38.8 +/- 10.7 years)] was performed. All patients underwent clinical examination, blood testing, and cardiopulmonary tests. Endothelial function was assessed using intima-media thickness and flow-mediated dilatation (FMD), as well as vascular function using applanation tonometry methods was completed while considering aortic systolic pressure, aortic pulse pressure (AoPP), augmentation pressure (AP), augmentation index (AI), pulse pressure amplification (PPampl), and pulse wave velocity (PWV).
Results: Concentrations of e-sel were significantly lower in patients with CHD than in controls (56.3 ± 13.6 ng/mL vs. 152.6 ± 84.7 ng/mL, $p<0.001$), while ADMA levels were higher (1.48 ± 0.48 ng/mL vs. 0.46 ± 0.1 ng/mL, $p<0.001$, respectively). E-sel levels correlated with gender, blood oxygen saturation, red blood cell, hemoglobin concentration, hematocrit, and augmentation pressure forced expiratory volume in one second, forced vital capacity, and oxygen uptake. ADMA levels were found to correlate only with age.
Conclusions: E-sel level, unlike ADMA concentration, reflects the degree of severity of erythrocytosis and hypoxia and, thus, the physical status of patients with cyanotic CHD. None of the mentioned biomarkers seems to reflect the endothelial and vascular function.

Keywords: congenital heart disease, e-selectin, adma

PP-448

External validation of two emerging perioperative mortality scores for adults undergoing cardiac surgery for congenital heart disease

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Background and Aim: There have been multiple attempts to develop a suitable predictive score for perioperative mortality in adults with congenital heart disease (ACHD) undergoing cardiac surgery. The PErioperative Adult Congenital Heart disease (PEACH) score, based on seven procedural and non-procedural factors, was published in 2021. The Adults with Congenital heart disease – Anatomical Physiological (ACAP) score, combining anatomical complexity (from I to III) and physiological clinical (from A to D) classes of the patient according to the American Heart Association/American College of Cardiology classification, was published in the same year. Both reported a high discriminative ability (area under the receiver-operating characteristics curve or c-index 0.88). We aimed to further validate these new scores on a German ACHD cohort.
Method: Data of all patients with congenital heart disease who underwent cardiac surgery at the age of 18 years or more between January 2004 and December 2013 were reviewed. For each patient, the PEACH and ACAP scores were calculated. Perioperative mortality was defined as 30-day mortality. The discriminative ability to predict perioperative mortality was assessed for each score using the c-index.
Results: A total of 830 patients (51% females, 45% with a history of cardiac operation during childhood) were included. The median age at surgery was 30 years (interquartile range 23 – 40). The most frequent operations were closure of atrial septal defect (n = 119, 14%), aortic valve replacement (n = 77, 9%), and replacement of right ventricle-to-pulmonary artery conduit (n = 75, 9%). A total of 27 patients (3%) died during the perioperative period. The values of c-index for predicting perioperative mortality for the PEACH (n = 786) and ACAP (n = 739) scores were 0.703 and 0.75, respectively. Within the ACAP score, the IID and IIID classes demonstrated the best discriminative ability (observed-to-expected ratio 1.02 and 1.05, respectively).
Conclusions: In our German cohort, the recently published PEACH score performs poorly at predicting early mortality in ACHD undergoing cardiac surgery. The ACAP score demonstrated better performance, especially the IID and IIID classes. Further refinement of these scores with more robust statistical models and novel patient-related factors is mandatory.

Keywords: adult with congenital heart disease, perioperative mortality scores

PP-449

Dynamic hyperinflation during exercise in patients with congenital heart disease – preliminary results

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Background and Aim: In patients with congenital heart disease (CHD), not only the cardiac function can be a performance-limiting factor, but also dynamic lung hyperinflation (DH) is considered a limitation to exercise. However, the study of DH and its clinical utility in CHD is limited and little is known regarding its

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prevalence, risk factors and clinical associations. The study aim was to describe the prevalence of DH during cardiopulmonary exercise testing (CPET) in CHD patients and its relationship with demographic parameters, lung function and exercise tolerance to better understand if and which patients are at greater risk for DH.
Method: This observational study investigated data from 139 children and adults with CHD (33.6 ± 16.1 years, 45.3% male) who performed spirometry and CPET including inspiratory capacity (IC) manoeuvres. Indication of DH was marked as a negative change in IC from rest to peak exercise (Delta IC) as well as a volume of the tidal breath that is flow limited on expiration/tidal volume (VFL/VT) > 50% during exercise. T-test and chi-square were used to estimate p-values.
Results: In total, 10 subjects (7%) showed evidence of DH during exercise. Patient groups did not differ in sex, age and body-mass-index (p>0.05). However, subjects with DH had a significant lower Tiffeneau-index (forced expiratory volume in first second/forced vital capacity, FEV1/FVC) compared to non-DH patients (DH: 70.8% ± 7.0 vs. non-DH: 76.9% ± 6.8, p=0.002). There was no group difference in peak oxygen uptake (DH: 25.2 ml/min/kg ± 7.1 vs. non-DH: 25.9 ml/min/kg ± 8.4, p=0.785), FEV1 (DH: 2.9 L ± 0.7 vs. non-DH: 2.7 L ± 0.8, p=0.935) or FVC (DH: 3.8 L ± 1.1 vs. non-DH: 3.5 L ± 1.0, p= 0.683).
Conclusions: Prevalence of DH in patients with CHD is low, and only the Tiffeneau-index (FEV1/FVC) could be associated with DH in patients with CHD in our preliminary analysis so far. Nevertheless, the identification of DH in CHD patients might be a subtle sign of ventilatory obstruction, thereby helping to provide pneumological and adapted treatment.

Keywords: dynamic lung hyperinflation, inspiratory capacity, lung function, cardiopulmonary exercise testing, congenital heart disease, congenital heart defect

PP-450

Unveiling women's experiences with cardiovascular health during pregnancy and care provided by a pregnancy heart team: a qualitative phenomenological

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Background and Aim: Cardiovascular diseases (CVD) are currently the leading cause of mortality in pregnant women. To date, the majority of empirical studies and guidelines include an assessment of clinical status, prognosis and outcomes of these women. However, we currently lack an in-depth understanding of the needs, expectations and experiences of women regarding the follow-up they receive during their pregnancy journey.
The present study aimed to (i) gain an in-depth understanding of the experiences of women with cardiovascular disease, and (ii) identify barriers and facilitators to the follow-up they receive during pregnancy.
Method: This qualitative phenomenological study involved data collection using semi-structured interviews in a large tertiary centre with a Pregnancy Heart outpatient clinic. Purposive sampling was used and patients were contacted by email. Data collection took place between December 2022 and September 2023. The transcribed interviews were subjected to thematic analysis.
Results: Thirteen patients were interviewed. Within the overarching theme of emotional distress, we distilled the following subthemes: awareness of the impact of CVD on the pregnancy journey, loneliness resulting from unheard concerns, and the need for increased psychological support and well-being. Within the overarching theme of the organisation of care, we distilled the following sub-themes: continuity of care and the skills required of healthcare providers.
Conclusions: Pregnant women with CVD warrant increased attention during and after pregnancy in terms of continuity of care and psychological support. These patient perspectives could significantly contribute to a more patient-centred approach to care. These findings provide an opportunity to incorporate the perspective of cardio-obstetric patients into guidelines and the formulation of an integrated approach to care.

Keywords: Cardiovascular obstetrics, cardiovascular diseases, pregnancy, patient experience, advanced practice nursing and midwifery, qualitative research.

Main figure

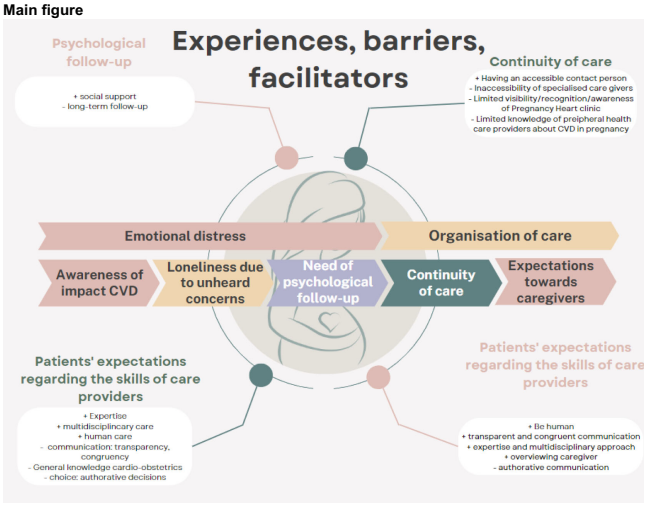

Figure 1: Main themes and sub-themes distilled from the interviews.

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PP-451

Pulmonary annular systolic excursion. towards a integral evaluation to right ventricular in patients with tetralogy of fallot and pulmonary stenosis

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Background and Aim: Surgery has improved the survival of patients with Tetralogy of Fallot and Pulmonary Stenosis. Pulmonary regurgitation is the most common complication, requiring cardiac resonance to define reintervention. The objective was to evaluate the function of the right ventricular outflow tract through excursion of the pulmonary annulus by echocardiogram and its relationship with the ejection fraction by resonance.
Method: Patients with repaired Tetralogy of Fallot and pulmonary stenosis, with recent cardiac resonance (within the last year), who were measured, by Doppler echocardiogram: the excursion of the pulmonary annulus in systole, the excursion of the tricuspid annulus in systole, right ventricular S wave tissue velocity, and fractional change in right ventricular area. They were compared with the right ventricular ejection fraction.
Results: 70 patients were included, tetralogy of Fallot in 65.7%. The excursion of the pulmonary annulus in systole was greater in the group with preserved right ventricular ejection fraction: 10.8 vs 9.1 mm (p=0.021). The excursion of the pulmonary annulus in systole showed a weak but statistically significant correlation coefficient with the right ventricular ejection fraction (r 0.27 $p < 0.05$) and with tissue velocity S (r 0.31 $p < 0.01$) but not with the excursion of the tricuspid annulus.
Conclusions: Pulmonary ring excursion is related to right ejection fraction and most indices of right ventricular systolic function.

Keywords: echocardiography, congenital heart disease, systolic function, Tetralogy of Fallot, Pulmonay stenosis.

PP-452

Propensity-matched comparison of fluoroscopic and echocardiographic guided transcatheter ventricular septal defect occlusion in chinese population

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Department of Structural Heart Disease, National Center for Cardiovascular Disease, China & Fuwai Hospital, Chinese Academy of Medical Sciences & Peking Union Medical College, Key Laboratory of Innovative Cardiovascular Devices, National Health Commission Key Laboratory of Cardiovascular Regeneration Medicine, National Clinical Research Center for Cardiovascular Diseases, Beijing, China

Background and Aim: Transcatheter therapy under fluoroscopic guidance has emerged as a minimally invasive option for treating perimembranous ventricular septal defect (pmVSD). However, patients receiving transcatheter occlusion may confront unavoidable radiation and unacceptable postprocedural or late-onset conduction abnormalities. We simplified the transcatheter process by employing echocardiographic guidance instead of fluoroscopy. The purpose of this study was to comparatively evaluate our long-term follow-up results of modified pmVSD occlusion and identify risk predictors of conduction abnormalities via robust machine learning methods.
Method: Nine hundred and fifty-four patients with pmVSD who had undergone transfemoral occlusion using fluoroscopic guidance (n=858) or transesophageal echocardiography (TEE) guidance (n=96) from January 2016 to December 2019 were retrospectively enrolled, and 54 pairs were propensity-score matched for 8 baseline variables. The primary study outcomes were the success rate of occlusion and postoperative conduction block at 3-year follow-up. Logistic regression with a stepwise selection of variables and gradient descent boosting were used to identify potential risk factors of persistent postoperative conduction abnormalities over the entire study period.
Results: Transcatheter pmVSD occlusion was successful in all patients enrolled in this study. Compared with fluoroscopic guidance group, the TEE guidance group had similarly distributed baseline characteristics, cardiac function parameters, and paravalvular regurgitation grade. We also observed that the TEE guidance group had lower rate of moderate to severe tricuspid regurgitant (18.9% vs. 40.6%, $p = 0.007$), left ventricular end diastolic diameter (42.5 vs. 44.0 mm, $p = 0.032$) at discharge. However, the proportion of patients with trivial to mild residual shunt in TEE guidance group was higher before discharge (5.7% vs. 2.1%, $p = 0.021$) but disappeared during follow-up. The occurrence of right bundle-branch block was higher in fluoroscopic guidance group without reaching statistical significance (0 vs. 1, $p = 0.609$). Both groups had no atrioventricular block occurrence. The occurrence of mitral or tricuspid regurgitation during follow-up served as a risk factor for predicting cardiac conduction abnormalities.
Conclusions: Fluoroscopic and echocardiographic guidance for transcatheter pmVSD occlusion appear comparably safe and effective, with high implant success and favorable 3-year clinical outcomes. However, the causality between valvular regurgitation and conduction abnormalities warranted larger studies with longer-term follow-up.

Keywords: ventricular septal defect, transcatheter occlusion, propensity-matched analysis, machine learning, echocardiographic guided therapy

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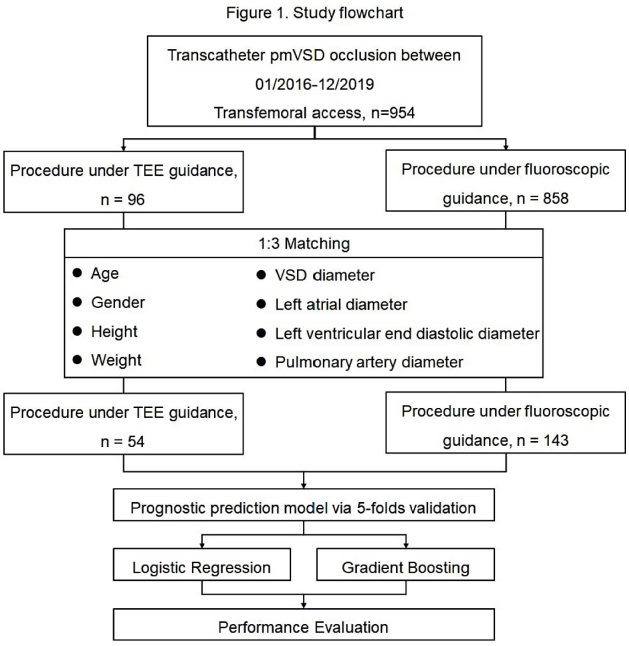


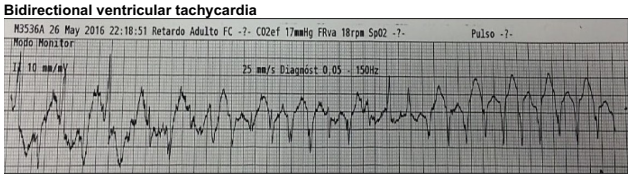
Study flowchart

changes after the return of spontaneous circulation. The diagnosis was later confirmed with genetic testing (mutation of the RYR2 gen). The patient was started on a beta-blocker and a subcutaneous implantable cardioverter-defibrillator was placed as it met the level A of evidence for defibrillator indication. Due to poor control with beta-blockers, flecainide was added. Sympathectomy is currently being considered.

Conclusions: Given the rarity of this condition, this diagnosis is often missed, which contributes to increased mortality rates. In children and young adults presenting with syncope without clear etiology in the presence of high-risk features, further evaluation should be performed including referral to cardiology to rule out chronic cardiac arrhythmias.

Keywords: catecholaminergic polymorphic ventricular tachycardia, pediatric electrophysiology, cardiac death, β-blockers.

Bidirectional ventricular tachycardia



Cardiac Dysrhythmias and Electrophysiology

PP-453
Two-way panic on the roller coaster

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Background and Aim: Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a rare congenital arrhythmogenic disorder induced by physical or emotional stress. It mainly affects children and younger adults and is characterized by rapid polymorphic and bidirectional ventricular tachycardia. Patients with CPVT may experience life-threatening arrhythmic events (LTAEs). The aim of this study is to report on a case of CPVT that went into cardiac arrest.
Method: We report a case of a patient diagnosed of catecholaminergic polymorphic ventricular tachycardia in the context of cardiac arrest.
Results: We present a case report of a 10-year-old male who collapsed during an emotional event (on a roller coaster) and went into cardiac arrest. Emergency department evaluation including imaging, laboratory studies, and EKG indicated the cause of cardiac arrest was likely a primary cardiac arrhythmia. An initial clinical diagnosis of catecholaminergic polymorphic ventricular tachycardia (CPVT) was made based on symptom onset during an emotional event, patient age, past episodes of chest pain and palpitations, absence of structural heart defect, and lack of EKG

PP-454
The efficacy and safety of adenosine triphosphate administration test in the management of Wolff–Parkinson–white syndrome in children with delta waves

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Background and Aim: Wolff–Parkinson–White (WPW) syndrome is a delta-wave syndrome frequently detected using school-based cardiovascular screening programs in Japan. Most patients are asymptomatic; however, initial symptoms may include syncope or sudden death, requiring appropriate diagnosis and management. Delta waves are also observed in the fasciculoventricular pathway (FVP), which poses no risk and requires no management. This study aimed to evaluate the efficacy and safety of the adenosine triphosphate (ATP) administration test for differentiating WPW syndrome from FVP.
Method: We retrospectively reviewed the medical records of 127 patients who were referred to our hospital between April 2008 and March 2022 due to signs of WPW syndrome on electrocardiography. WPW syndrome and FVP were determined based on atrioventricular blocks and QRS waveform changes after ATP infusion during sinus rhythm. The procedures were performed on an outpatient basis.
Results: Overall, 127 patients were included (65 males; median age: 12.8 [6.8–22.1] years, resting heart rate: 75 [40–103] bpm, PR interval: 109 [72–160] ms, and QRS duration: 101 [69–157] ms). The outcomes of the rapid intravenous ATP infusion revealed FVP, WPW syndrome, and indeterminate in 64 (50.4%), 54 (42.5%), and 9 (7.1%) patients, respectively. More than 60% of patients with QRS duration ≤120 ms had FVP. Multivariate analysis showed independent risk factors for WPW syndrome were age ≦12 years (OR: 3.32; 95% CI: 1.37 to 8.05; p = 0.008), QRS duration >120 ms (OR: 5.34; 95% CI: 1.79 to 15.93; p = 0.003),

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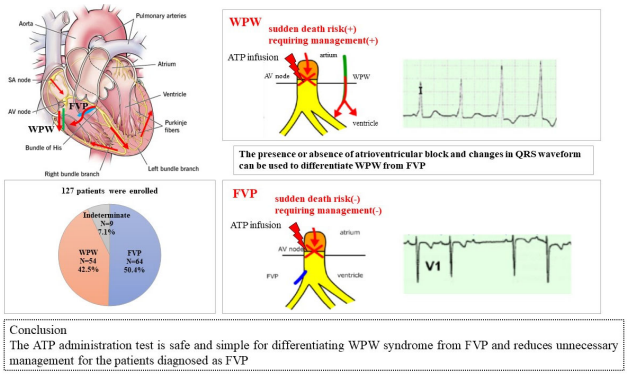
and Roseumbaum classification type A (OR: 12.51; 95% CI: 4.02 to 38.95; p = 0.000). The optimal ATP dose was 0.2–0.3 mg/kg in approximately 90% of patients; however, no patient experienced adverse events.
Conclusions: The ATP administration test is safe and simple for differentiating WPW syndrome from FVP. If the result leads to a diagnosis of FVP, the incidence of sudden death and tachycardia is unlikely, and unnecessary management can be reduced.

Keywords: Preexcitation syndrome, Wolff–Parkinson–White syndrome, fasciculoventricular pathway, adenosine triphosphate administration test, electrocardiogram, delta waves

Graphical Abstract



PP-455

Ablation of an incessant left ventricular tachycardia supported by extracorporeal membrane oxygenation in a 2-year-old patient post COVID-19 infection

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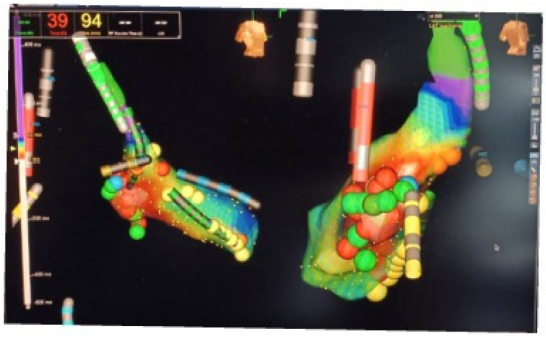
Background and Aim: This case shows the successful treatment of an incessant ventricular tachycardia (VT) in a 2-year-old boy under veno-arterial extracorporeal membrane oxygenation (VA-ECMO). Four weeks earlier he suffered from COVID-19 infection followed by general deterioration associated with tachycardia.
Method: On presentation at ER, the patient was generally unwell with a heart rate of 220 bpm, but hemodynamically stable. The ECG was interpreted as supraventricular tachycardia. Treatment with adenosine was inefficient. After suspected VT, amiodarone was administered, leading to bradycardia and pulseless electrical activity. After 13 minutes of CPR, he stabilized and was transferred to our center.
Results: The patient presented a broad drug resistant VT, treated with amiodarone, magnesium, several rounds of electric cardioversion, and later verapamil, as the suspected diagnosis was idiopathic left ventricular (LV) tachycardia. He developed a bradycardic rhythm with significant hypotension, echocardiography showed a highly impaired ejection fraction (EF). VA-ECMO was implanted that stabilized the circulation. However, the ECG developed long QT and showed an increase in polymorphic VES, resulting in Torsade-de-Point tachycardia. Therapy with verapamil was restarted and additionally transesophageal pacing.
He underwent electrophysiological examination, intubated and under ECMO-support. As initially fascicular VT was suspected, mapping was performed in the LV, with a near perfect pacemap basal inferior. During the examination the clinical VT started. Blood pressure stayed stable under EMCO-therapy. Ablation was performed in the suspected area, with subsequent termination of the micro-reentrant VT in the inferior basal LV (figure 1).
The patient was monitored for 32 hours without antiarrhythmic therapy, with no further episodes of VT. His EF improved and ECMO could be explanted without residues. In the follow up period of two years he is under betablocker therapy in good clinical condition without any VT relapse.
Conclusions: The case presented shows the possibility of VT ablation under ECMO therapy in a pediatric patient. Cardiac sequelae (such as myocarditis) following COVID infection should always be considered, and specifically investigated early on. In refractory drug resistant VT and hemodynamic instability VA-ECMO can also provide circulatory support during pediatric ablation procedures.

Keywords: ventricular tachycardia, ecmo, covid

Figure 1



Electrophysiological examination with ablation map in LV

PP-456

The challenge of transseptal access for catheter ablation of left-sided arrhythmia substrate in infants and small children under 6 years of age

Zsofia Nagy, Andrea Szabo, Levente Csakany, Tamas Major, Zoltan Som, Csaba Foldesi and Laszlo Kornyei
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Background and Aim: Young age and small cardiac dimensions are increased risk factors for catheter ablation (CA). Transseptal puncture (TSP) may be an additional risk factor. Patent foramen ovale (PFO) in infants can mitigate this risk and decrease the complication rate in the very young. This study aimed to compare the results of left-sided CA in children under 1 year and between 1-6 years of age.
Method: Between January 2010 and September 2023, 1703 pediatric CAs were performed, of which 17 left-sided ablations in children under 6 years without structural heart disease were retrospectively evaluated. CA results below 1 year and between 1-6 years of age were compared in terms of body size, procedural parameters, left atrial (LA) access, complication, and success rate.
Results: The patient group below 1 year (Group 1, n=5) has a mean age=0.13±0.03 years, mean body weight=3.6±1.1 kg, mean height=52.3±3.6 cm. Patients between 1-6 years (Group 2, n=12) have mean age=5.0±1.1 years, mean body weight=17.9

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±2.2 kg, mean height=111.2±7.4 cm. The arrhythmia substrate was an overt pathway in 3 pts (17.6%), concealed pathway in 7 pts (41.2%), and focal LA tachycardia in 7 pts (41.2%). Foramen ovale was persistent in all 5 children in Group 1 and TSP was not required. In Group 2 only 2 pts (16.7%) had PFO, and all other pts required TSP. TSP was successful in 9/10 cases (90%). No complications related to LA access or other adverse events occurred in Group 1. In Group 2, there was one case of pericardial effusion that spontaneously regressed and one case of mechanical-induced advanced AV block that resolved after six days. There were no significant differences in procedure time, fluoroscopic time, and radiation dose between the two groups (Group 1: 126.4±47.0 min, 6.4±4.5 min, 4.0±5.3 cGycm^2 vs. Group 2: 162.6±49.2 min, 7.9±9.4 min, 9.0±12.4 cGycm^2, p≥0.05). All CAs were performed with RF energy. The acute success rate of the procedures was 100%.
Conclusions: CA of left-sided arrhythmias in small children may offer a safe and effective option in selected cases. The high prevalence of PFO may explain the paradoxically less complication rate under 1 year of age.

Keywords: catheter ablation, transseptal puncture, patent foramen ovale, infants and small children

PP-457
Catecholaminergic polymorphic ventricular tachycardia: Long term experience of single pediatric centre
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Background and Aim: CPVT is a genetic disease characterized by emotion- or exercise-induced supraventricular and ventricular arrhythmias.The disease has an estimated prevalence of 1 in 10000.The present study aims to examine the long-term outcome of a single-centre pediatric cohort of CPVT patients.
Method: All data about patients with CPVT followed up in our Centre from 2005 were revised
Results: 23 patients (13 females), diagnosed as affected by CPVT at median age of 12.29 ± 4.84 years, were enrolled.
The median age at the first manifestation of the disease was 10.27 ± 4.71 years and always during exercise or emotion:syncope in 12 (52%) patients, resuscitated cardiac arrest in 5 (22%), ventricular arrhythmias detected during sport participation screening in 2 (9%). Two patients had neurological sequelae of cardiac arrest. Four patients (17%) were diagnosed due to familiar screening. Genetic testing was performed in 20 patients (87%), RYR2 variants were documented in 17 of 20 patients (85%). The median follow-up is 7.2 ± 7.4 years. All survived patients are on Nadolol (mean dose of 1,65 mg/kg) that is in combination with Flecainide (average dose of 2,85 mg/kg) in 1 (56%) patients. There were no side effects. An implantable loop recorder (ILR) was implanted in 7 patients (30%) without any complications. Seven patients (30%) underwent ICD implantation and complications occurred in 3: lead-associated thrombosis in 2 cases, electrode lead fracture in the other one. Two patients were lost to follow up. Among the remaining 21 (91,3%) patients, one (4.8%) patient died at the age of 16 due to electrical storm and arrhythmia induced cardiomyopathy in 2011, 2 (9.5%) patients had recurrent SVT exercise-induced. All patients are currently asymptomatic; where possible (20 patients, 87%), the exercise test showed the presence of complex supraventricular premature beats in 2 (10%) patients, non-sustained VT in 1 (5%), isolated monomorphic premature ventricular beats in 3 (15 %), polymorphic ventricular premature beats in 5 (25%), no arrhythmia in the remaining 9 (39%).
Conclusions: CPVT is mostly diagnosed secondary to an adrenergic impact event. Family screening after genetic testing allows for preventive diagnosis. Beta-blockers, and particularly Nadolol, still remain the cornerstone of treatment. ILR may be useful to optimize medical treatment or indicate ICD implantation despite apparently effective medical treatment. The ICD can be implanted but serious complications can occur in about 40% of cases.

Keywords: Children, Catecholaminergic polymorphic ventricular tachycardia, Arrhythmic event, Syncope, nadolol

PP-459
The importance of home-monitoring as a method of cardiac events diagnosis in a child with intellectual disability– a case report
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Background and Aim: Cardiomyopathies are rare cardiac disorders primarily affecting myocardium. The most common type is hypertrophic cardiomyopathy (HCM) which is also the most frequent genetically inherited cardiac disorder, affecting nearly 0,2% of population. HCM is frequently associated with arrhythmias leading to sudden cardiac (SCD) death and thus patients with HCM may require ICD implantation as a SCD preventive method.
Method: See below.
Results: We present a 17-year-old male patient diagnosed with HCM, nephrotic syndrome, intellectual disability and a long history of syncopies. He was diagnosed with HCM at the age of 8 and remained under regular cardiological care with a mild interventricular septum hypertrophy (IVSd 16mm, Z-score 3.2) without left ventricular outflow tract obstruction. Due to significant impairment of cognitive functions objective evaluation of patient's symptoms was impossible. Over the years, the boy presented with several episodes of syncopies, however no rhythm disorders were registered until the age of 17 when the first episode of atrial fibrillation (AF) was recorded. The patient's family history revealed dilated cardiomyopathy in his mother and SCD in a brother with intellectual disability and epilepsy at the age of 10. We performed genetic testing with next generation sequencing method which revealed presence of a variant of unknown significance in MYPB3 gene (c.2560A>T; p.Met854Leu).
Due to high risk of sudden cardiac death (12% according to ESC HCM Risk-SCD calculator) the boy had ICD implanted. He had two episodes of appropriate ICD therapy for ventricular fibrillation preceded by AF with subsequent sinus pause. The child did not report any symptoms during both episodes and we were informed about them by home-monitoring. He underwent pulmonary veins isolation for AF, but due to recurrence of arrhythmia double pharmacological treatment was given.
Conclusions: The presented case shows dangerous manifestation of mild HCM with complex medical history and severe clinical presentation. With the presented patient we want to highlight the importance of SCD risk evaluation with available calculators, as well as the necessity of ICD implantation as a SCD preventive method. Home-monitoring can serve as a method of detecting

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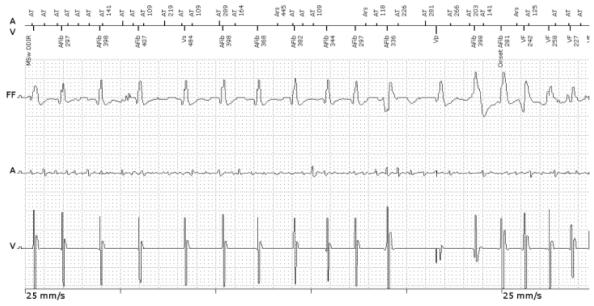
life-threatening arrhythmias, especially in patients with intellectual disability.

Keywords: Home-monitoring, hypertrophic cardiomyopathy, atrial fibrillation, ICD, intellectual disability.

Ventricular fibrillation preceded by atrial fibrillation



The image shows the beginning of an episode of ventricular fibrillation preceded by atrial fibrillation with subsequent sinus pause. There was later an appropriate ICD therapy. We were informed about the event by home-monitoring.

PP-460
Resting 12-lead ecg evolution in pediatric duchenne muscular dystrophy patients

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Background and Aim: Cardiac involvement in Duchenne muscular dystrophy (DMD) is characterized by progressive myocardial fibrosis, left ventricle dysfunction alongside with electrocardiographic changes. With significant differences found in resting ECG dependently on patients age in previous analyses, in current study we aimed to conduct a deepen analysis of the ECG evolution with the disease progression.
Method: The analysis of heart rate (HR) and P, QRS amplitude and duration in resting 12-lead ECG recordings in patients with genetically confirmed DMD diagnosis was performed in the presented single-centre observational study. Reference values by Rijnbeek et al. were used for z-score calculations. Finally, a comparison between younger (≤ 9 years old) and older (>9 years old) patients was performed. Statistical analysis was performed Wizard 2.0 (Evan Miller, Chicago, IL. Z-score values, mean (±SD) are presented.
Results: A hundred and twenty-five studies from 84 patients (83, 97,6% male) were analyzed. Based on the age at the time of the recording patients were assigned to "early course" or "late course" group, n=63, aged 6.25 ±2.00 years old and n=62 aged 12.99 ±2.35 respectively. HR was found to be significantly higher in the older group of patients, z-scores 1.17 ±1.24 vs. 1.63 ±1.28, p=0.04. Later in the course of the disease Q wave was found to be lower in V6 (1.09 ±1.21 vs. 0.69 ±0.79, p=0.03), R wave higher in aVL (1.31 ±1.62 vs. 2.26 ±2.76, p=0.02), and V1 (0.53 ±1.24 vs. 1.19 ±1.40, p=0.01) but lower in V6 (-1.05 ±1.14 vs. -1.54 ±1.17, p=0.02). S wave was found to be higher in aVL (-0.05 ±0.77 vs. 0.33 ±0.69, p=0.01) and III (0.46 ±2.02 vs. 1.91 ±2.99, p=0.01), but lower in V2 (-0.86 ±1.39 vs. -1.62 ±1.18, p<0.01)
Conclusions: ECG in DMD undergoes significant evolution throughout the course of the disease in childhood. It is yet to be determined if novel approach using z-scores applied to ECG will reveal early prognostic factors of more adverse cardiac outcomes.

Keywords: Duchenne Muscular Dystrophy, Resting ECG, QRS amplitudes

Mean ± SD of z-score values based on Rijnbeek et al. stratified by age and p-value for the corresponding comparisions

	n	early	late	p
Age	125	6.25 ±2.00	12.99 ±2.35	
S_aVF_amp	104	-0.15 ±0.78	0.02 ±0.84	0.26
R_aVF_amp	124	-0.13 ±1.00	-0.42 ±1.26	0.17
Q_aVF_amp	95	0.86 ±1.89	1.05 ±1.40	0.59
S_aVL_amp	116	-0.05 ±0.77	0.33 ±0.69	0.01 H
R_aVL_amp	125	1.31 ±1.62	2.26 ±2.76	0.02 H
S_aVR_amp	109	0.38 ±1.84	0.41 ±1.94	0.93
R_aVR_amp	123	0.25 ±2.28	0.69 ±3.29	0.39
S_III_amp	95	0.46 ±2.02	1.91 ±2.99	0.01 H
R_III_amp	125	-0.03 ±1.07	0.09 ±1.43	0.62
Q_III_amp	87	1.09 ±2.31	0.82 ±1.83	0.55
S_II_amp	108	0.13 ±1.90	0.35 ±1.58	0.50
R_II_amp	125	0.19 ±1.11	-0.10 ±1.32	0.18
Q_II_amp	108	0.88 ±1.84	0.78 ±1.46	0.75
P_II_amp	125	0.03 ±0.69	0.28 ±0.89	0.08
S_I_amp	114	0.07 ±0.85	0.45 ±1.26	0.07
R_I_amp	124	1.17 ±1.56	1.65 ±2.26	0.18
S_V6_amp	112	-0.75 ±0.35	-0.59 ±0.68	0.13
R_V6_amp	125	-1.05 ±1.14	-1.54 ±1.17	0.02 L
Q_V6_amp	120	1.09 ±1.21	0.69 ±0.79	0.03 L
S_V4_amp	122	-0.93 ±0.69	-1.11 ±0.68	0.14
R_V4_amp	123	-0.10 ±1.75	-0.49 ±1.44	0.18
S_V2_amp	123	-0.86 ±1.39	-1.62 ±1.18	0.00 L
R_V2_amp	125	1.44 ±1.99	1.47 ±1.49	0.91
P_V2_amp	124	-0.43 ±1.31	-0.64 ±1.08	0.35
S_V1_amp	125	-0.33 ±1.05	-0.68 ±0.94	0.05
R_V1_amp	125	0.53 ±1.24	1.19 ±1.40	0.01 H
P_V1_amp	125	-0.66 ±0.76	-0.35 ±1.04	0.06
HR_m	125	1.17 ±1.24	1.63 ±1.28	0.04 H
P_duration_m	125	-2.27 ±1.01	-2.74 ±0.87	0.01 L
PR_interval_m	125	-1.26 ±0.79	-1.47 ±0.74	0.14
QRS_duration_m	125	0.71 ±1.04	0.66 ±0.98	0.76

Fig. 1. Mean ± SD of z-score values based on Rijnbeek et al. stratified by age and p-value for the corresponding comparisions

PP-461
Exploring neonatal cardiac arrhythmias: A dual case study of permanent junctional reciprocating tachycardia

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Background and Aim: Permanent junctional reciprocating tachycardia (PJRT) is one of the causes of refractory supraventricular tachycardia (SVT), rarely diagnosed in neonatal period. It presents with long RP tachycardia, inverted P waves in the inferior leads and is

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characterized by incessant tachycardia and a slower ventricular rate (around 200 beats/min). The risk of tachycardia-induced cardiomyopathy is significant.
Method: We present 2 cases of fetal PJRT shedding light on the particularities surrounding diagnosis and treatment.
Results: Case 1: A 34-year-old pregnant had an uneventful pregnancy until 32 weeks' gestation when fetal tachycardia was detected. Fetal echocardiography revealed no structural cardiac anomalies. The male neonate was delivered via cesarean section due to persistent tachycardia (max 180 bpm), and exhibited inverted P-waves in DII, DIII and aVF on ECG, suggesting PJRT. Propranolol was initiated on D4. The infant was discharged on D16 with a normal ECG. Currently, at 20 months of age, has no recurrence, under propranolol.
Case 2: A 29-year-old pregnant had fetal tachycardia detected at 24 weeks. Transplacental therapy with flecainide and digoxin transitorily controlled fetal tachycardia. An elective cesarean section was performed at 33 weeks and 6 days due to non-reassuring fetal status. At birth, the female newborn presented refractory SVT. with a heart rate of 220 beats/min. The diagnosis of PJRT was confirmed. After numerous treatment attempts, tachycardia was controlled with high dose propranolol and flecainide. She was discharged on D31. Flecainide was stopped at age 2 and propranolol at age 4. No recurrence at 6 years' follow-up.
Conclusions: The importance of the ECG pattern in diagnosis of PJRT is highlighted. The response to therapy was achieved in both cases. However in case 2 management was challenging, suggesting that the earlier development of PJRT may result in a more severe disease and involved a multidisciplinary approach including obstetrics, pediatric cardiology, electrophysiology and neonatology.

Keywords: neonatal cardiac arrhythm, permanent junctional reciprocating tachycardia

PP-462
Presentation and outcomes of paediatric radiofrequency catheter ablation for supraventricular tachycardia according to sex and age

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Background and Aim: In paediatric age, supraventricular tachycardia (SVT) is the main indication for an electrophysiological study (EPS) with ablation. We aimed to describe the clinical presentation and outcomes of SVT submitted to radiofrequency catheter ablation (RFCA) in paediatric age, according to sex and age.
Method: A retrospective analysis of patients aged ≤18 years submitted to EPS and RFCA between January 2008 and December 2019 was conducted in a non-surgical center. Demographic data, symptoms at presentation, immediate and late success rate, and incidence of complications were evaluated. Follow-up data was obtained via routine visits or telephonic contact. Recurrence was established by electrocardiography or EPS. Patients were divided in two groups, based on the median age: ≤15 years and >15 years.
Results: We included 59 children submitted to RFCA. The mean age was 14.3±2.9 years (between 7 and 18 years) and 25.4% were female. The majority had structural normal heart except for 2 cases: 1 pulmonary valve stenosis, and 1 bicuspid aortic valve. Mean follow-up was 6.6 years The most frequent indication was the presence of accessory pathways (AP) in 79.7% (n=47), mostly of left lateral location (n=22), or atrioventricular nodal reentrant tachycardia (AVNRT) in 20.3% (n=12). Most patients were symptomatic (85,1%), with palpitations as dominant symptom. Presentation was similar regardless of age and sex (Figure 1). The immediate success rate was 97% (AP 96%, AVNRT 100%); the 2 subjects with procedural failure were male, aged 12 and 15 years. A repeat ablation occurred in both with subsequent success. Overall, after primary successful RCA, the long-term success rate was 84.2% (AP 82.2% AVNRT 91.7%) with all recurrences occurring during the first year (mean time to recurrence of 77 days). Long term outcomes were not significantly different between sex and age groups (Log rank p=0.807 and p=0.767, respectively, Figure 2). No immediate or late complications were reported.
Conclusions: These findings support the use of RFCA as an effective and safe treatment for most tachyarrhythmias across pediatric age with a high success rate and no reported complications. Age and sex do not seem to influence symptomatic presentation.

Keywords: radiofrequency catheter ablation, supraventricular tachycardia, outcomes, sex, age

Figures

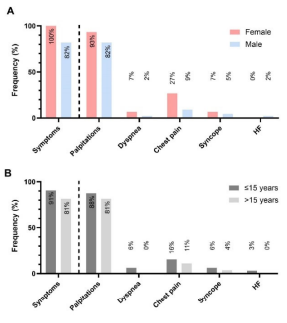

Figure 1: Symptomatic presentation of supraventricular tachycardia according to sex (A) and age (B)

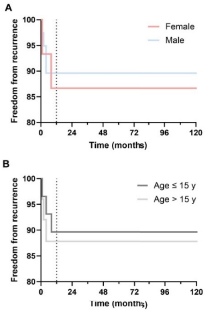

Figure 2: Freedom from supraventricular tachycardia 10 years after percutaneous ablation according to sex (A) and age (B)

Figure 1: Symptomatic presentation of supraventricular tachycardia according to sex (A) and age (B) Figure 2: Freedom from supraventricular tachycardia 10 years after percutaneous ablation according to sex (A) and age (B)

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PP-463

Value of exercise testing in risk stratification of pediatric patients with arrhythmogenic cardiomyopathy

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Background and Aim: High-intensity training can lead to earlier and more severe expression of arrhythmogenic cardiomyopathy (ACM). Moreover, sudden cardiac death in patients with ACM can frequently occur during exercise. Thus, exercise testing is routinely performed in the diagnostic evaluation and during the follow-up in this particular setting of patients.

The aim of the study was to evaluate the arrhythmogenicity of exercise testing (ET) in our pediatric population affected by ACM.

Method: This is a retrospective observational study in which all pediatric patients (<18 years) diagnosed with ACM since 2009 at our institution were enrolled and the results of ET, performed in the absence of any medical treatment, were evaluated.

Results: Occurrence or persistence of arrhythmias during ET, in the absence of any medical treatment, was evaluated in 21 pediatric patients with a "definite" ACM diagnosis according to the "2020 Padua Criteria". In the 16 patients with premature ventricular contractions (PVCs) at rest, 10 showed a complete suppression of PVCs during ET. In all of them, PVCs occurred again during recovery: in 8, with the same morphology, and in 2 with different morphologies. In the other 6 patients, PVCs were observed during exercise and recovery as well.

In 4 patients without arrhythmias at rest, ventricular arrhythmias occurred during exercise: sustained monomorphic ventricular tachycardia (VT) in 1, non-sustained polymorphic VT in 1 and polymorphic PVCs and couplets in the others.

Monomorphic PVCs occurred during recovery in one patient, without arrhythmias at rest.

Conclusions: The arrhythmogenicity of exercise pediatric patients with ACM is highly variable and unpredictable. Exercise-induced polymorphic and/or repetitive ventricular arrhythmias can occur in patients without arrhythmias at rest and suppression of PVCs during exercise should not be considered reassuring during the diagnostic work-up.

Keywords: Arrhythmogenic Cardiomyopathy, Arrhythmias, Pediatric Patients, Premature Ventricular Contractions, Exercise Testing

PP-465

The efficacy of the use of atropin in children with pallid breath holding spells: Can cardiac pacemaker implantation be avoided?

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Background and Aim: Reflex anoxic seizures are a manifest overreaction of the vagal system, with hypotension and bradycardia or brief cardiac arrest. Because of the benign and self-limiting character of the spells with virtually no complications on short or long term, treatment is only necessary in case of frequent spells or severe clinical presentation. Treatment options are medication such as atropine or the implantation of a cardiac pacemaker, with the latter being invasive and entailing risk for important complications. We investigated atropine treatment and aimed to examine if pacemaker placement can be avoided.

Method: We retrospectively reviewed patients treated in our center for severe reflex anoxic seizures with atropine sulphate from January 2017 until May 2023, and compared our results to those in the literature. Patient selection was thrugh clinical history and holter registration with significant bradycardia or asystole during the spells.

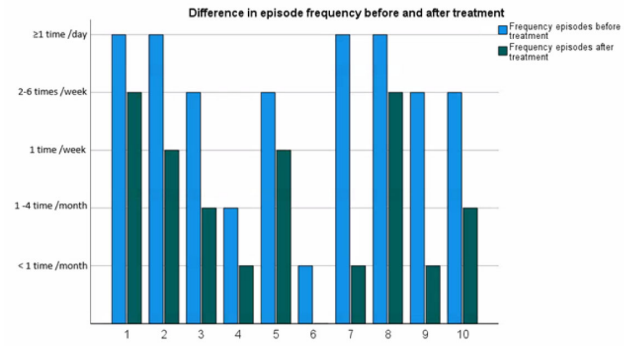
Results: We recruited 10 patients in total. In our population, 100% of the patients reported adequate symptom management after atropine treatment, with complete resolution in 10%. Minor side effects were reported in 70%, with need for change of treatment in one patient (10%). Severe complications were not reported in our population, unlike reported incidences up to 40% of permanent or severe complications in patients which received a pacemaker.

Conclusions: Atropine is a safe and efficient treatment to manage the symptoms, with similar success rate to pacemaker implantation. However, pacemaker implantation entails a substantial risk for complications and is accompanied with morbidity such as scar formation. This might be considered redundant for a benign and temporary condition, certainly given the possibility of other efficient treatment option.

Given the benign nature of the spells and the good long-term prognosis, coupled with the success and complication rates as described, we recommend atropine treatment as the better option to implantation of a cardiac pacemaker in children with severe RAS.

Keywords: Breath holding spells, Atropin, pacemaker

Difference in spell frequency before and after atropin treatment.



All parents reported a significant decrease in symptom frequency after atropin treatment, resulting in a managable frequency.

PP-466

Case report: Everolimus for cardiac rhabdomyomas in neonate with tuberous sclerosis complex and significant arrhythmias

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Background and Aim: Cardiac rhabdomyomas are the most common neonatal intracardiac tumors and the main feature of tuberous sclerosis complex (TSC). Regression without intervention is possible, and surgical resection is reserved for significant arrhythmias or obstruction. The use of everolimus, an mTOR

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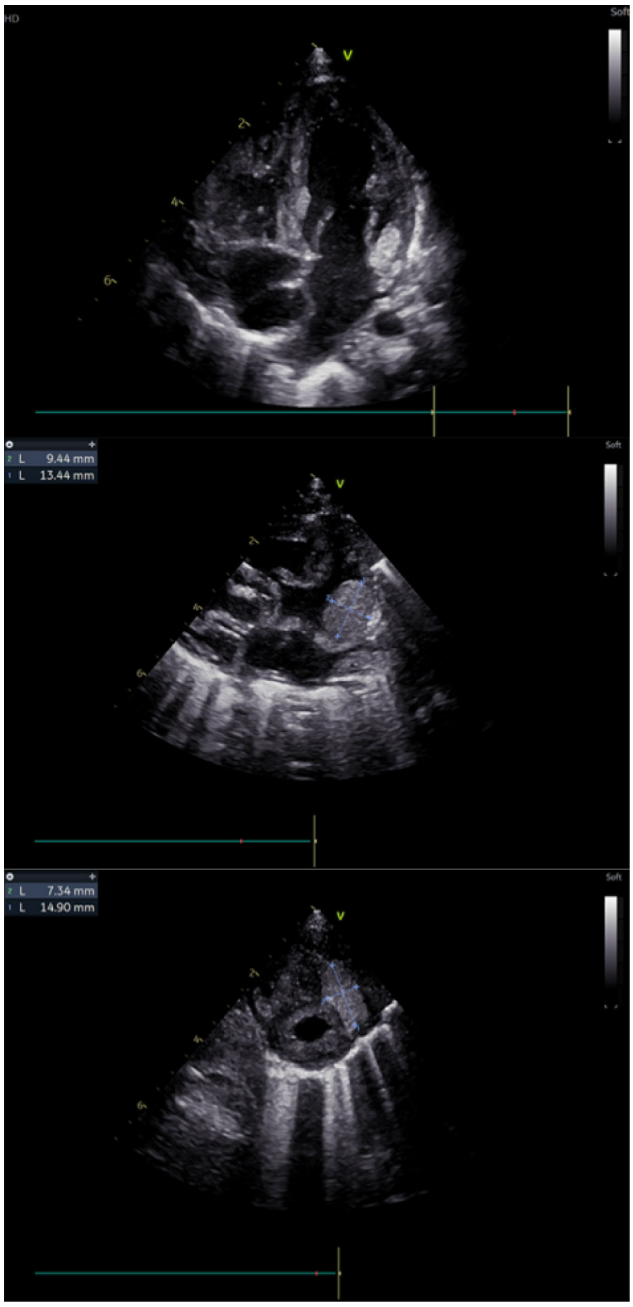
inhibitor, while infrequent, shows potential benefits, particularly for high-risk patients with life-threatening arrhythmias or hemodynamic compromise.
Method: We present the case of a term male neonate delivered via cesarean-section, to a healthy woman, due to fetal bradycardia. Fetal echocardiogram (performed in the third trimester) and all obstetric ultrasounds were normal. The delivery was unremarkable, with Apgar scores of 8 and 9 at 1 and 5 minutes. After birth, the neonate still exhibited bradycardia, rapidly progressing to supraventricular tachycardia (SVT) before spontaneously returning to sinus rhythm (SR) within hours. An electrocardiogram revealed sinus bradycardia (approximately 90 bpm) with no other significant findings. Transthoracic echocardiography revealed multiple rhabdomyomas, including one on the right atrium, at the entrance of the superior vena cava (SVC), near the sinoatrial node, potentially contributing to the bradycardia.
On the same day, a transfontanelar ultrasound identified cortical tuberomas, and an ophthalmological evaluation revealed a retinal hamartoma, leading to the diagnosis of TSC.
Despite an initially stable period, on the seventh day, the neonate resumed frequent SVT episodes (approximately 280 bpm) while maintaining hemodynamic stability. Transient reversion to SR with adenosine suggested the likelihood of atrioventricular reentry tachycardia via a concealed accessory pathway, the most common cause of SVT in infancy.
Results: The presence of baseline bradycardia, likely due to the rhabdomyoma on the entrance of the SVC, posed treatment challenges for the tachyarrhythmia. Management involved low-dose propranolol and flecainide, as well as everolimus. After approximately three months of mTOR inhibitor treatment, complete regression of cardiac rhabdomyomas was achieved, leading to SR with an appropriate heart rate for the patient's age, under antiarrhythmic therapy with propranolol and flecainide.
Conclusions: This case underlines the importance of a comprehensive approach in neonatal cardiac care, showing the potential advantages of everolimus in managing life-threatening arrhythmias and hemodynamic challenges. Early and precise diagnosis is crucial for tailored interventions that yield favorable results in complex clinical scenarios.

Keywords: Cardiac rhabdomyoma, Tuberous sclerosis complex, Bradycardia, Supraventricular tachycardia, mTOR inhibitor, Everolimus

TSC



PP-467

Preliminary results on shortening the duration of antiarrhythmic medication to four months in infants diagnosed with supraventricular tachycardia

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Background and Aim: Infants diagnosed with supraventricular tachycardia (SVT) are treated with antiarrhythmic medication (AM) to avoid recurring arrhythmias. In our recent retrospective study shortening AM duration in infants from 12 to 6 months did not seem to lead to more frequent SVT recurrences. We investigated whether AM duration of four months would be sufficient to allow for resolution of arrhythmias and thus prevent recurrences.
Method: This prospective multicentre national cohort study included infants less than 6 months of age diagnosed with SVT in 13 secondary and five tertiary care hospitals from 2021 to 2023. These preliminary results are a part of an on-going prospective study. From the beginning of 2021, all infants diagnosed with SVT were treated with AM for four months, mostly with propranolol. The patients were followed until the age of one year. A documented episode of SVT after discontinued AM was considered as a recurrence.

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Results: A total of 35 infants were recruited in the study during 2021. Their median age at diagnosis was 12 days (interquartile range, IQR 5-25.3). The majority of infants were male (n=20, 57%). Ten infants (29%) were diagnosed with congenital heart disease and five (14%) had an antenatal diagnosis of SVT. The arrhythmia was diagnosed in 25 of infants (71%) while admitted to or already treated in the hospital for other reason than arrhythmia, and 19 (54%) needed cardioversion with adenosine. Primary AM was propranolol in 33 infants (94%) and 57% of them did well with only one AM. Eight infants (23%) needed two AMs. The median duration of AM was 4.3 months (IQR 4-4.5), but in two cases (6%), AM continued for a longer time due to continuous breakthrough arrhythmias. With the median follow-up time of 8.6 months (IQR 7.7-11.7), two of 33 infants (6%) had recurrence of SVT after the cessation of AM.
Conclusions: Shortening the duration of AM in infants with SVT to four months appears to be safe and recurrence of SVT after discontinuation of AM is rare.

Keywords: supraventricular tachycardia, antiarrhythmic medication, infants

PP-469
Cardiopulmonary exercise testing in complete atrioventricular block. Is it useful in the indication of implanting a pacemaker?

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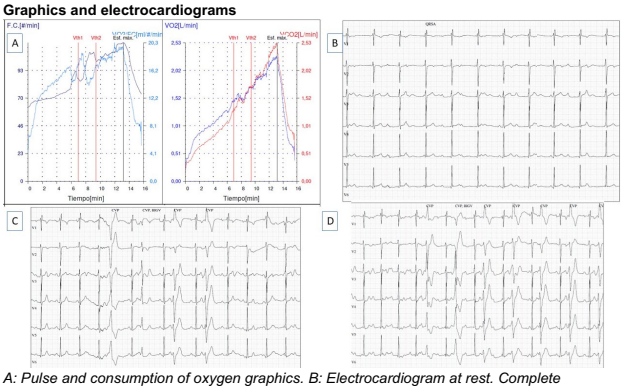
Background and Aim: Permanent pacemaker implantation is controversial in asymptomatic adolescents with isolated complete atrioventricular block (CAVB) and acceptable ventricular rate, a narrow QRS complex, and normal ventricular function. Guidelines recommend pacing these patients based on an individualized consideration of the risk/benefit ratio.
Use of treadmill stress testing is well known in detecting arrythmias, checking the blood pressure and heart rate response to exercise in CAVB patients. On the contrary, cardiopulmonary exercise testing (CPET) is not included in usual follow up protocols in CAVB.
Method: To establish a follow up protocol in CAVB that adds the CPET to the previous tests (ambulatory cardiac monitoring, electrocardiogram and echocardiography) conducted in our hospital. Patients with isolated congenital or acquired atrioventricular block were included. None of them had previous indication to undergo pacemaker implantation.
Results: 2 patients are included. Both are male, active and asymptomatic adolescents. One has a congenital heart block detected before birth, and the other was diagnosed a CAVB in the course of a bronchiolitis. Previous holter and exercise testing were done in the follow up. Both had narrow QRS complex, no pathological pauses, mean heart rate of 58 and 47 bpm respectively in the holter, and tachycardiation, no arrythmias and normal functional capacity in the exercise testing.
CPET in the first patient showed no significant disturbances except for a ventricular premature beat that appeared in minutes 4 and 10 of exercise. Oxygen pulse curve was normal. On the other hand, the second patient presented at minutes 6 and 8 of exercise at least 3 types of ventricular premature complex (right bundle branch block and inferior axis the most common one) that joined in ventricular bigeminy. In that moment, patient's oxygen pulse and blood pressure fell down with no symptoms, patient continued running until he felt tired at minute 13. Oxygen pulse and pressure alterations were recovered when extrasystoles disappeared. Because of these finding, patient 2 will undergo pacemaker implantation.
Conclusions: CPET could be useful in the follow up of CAVB patients. It can demonstrate oxygen pulse alterations and blood pressure repercussion that could guide in establish the indication of implanting a pacemaker.

Keywords: pacemaker, exercise testing, complete atrioventricular block



A: Pulse and consumption of oxygen graphics. B: Electrocardiogram at rest. Complete atrioventricular block. C: Ventricular premature complex at minute 4. D: Ventricular premature complex at minute 10.

PP-470
Ventricular function in children with frequent premature ventricular complexes and structurally normal hearts

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Background and Aim: Premature ventricular complexes (PVCs) are frequent in the pediatric age group. Although PVCs are usually considered benign, frequent PVCs can cause ventricular dysfunction or might be the first sign of cardiomyopathy. The aim of the study: to evaluate left (LV) and right ventricular (RV) function in children with frequent PVCs.
Method: The prospective study of children with structurally normal hearts and ≥20% PVCs per 24 hours was performed in Vilnius University Hospital Santaros Klinikos from 1st January 2022 and 30th August 2023. The exclusion criteria were diagnoses of hemodynamically significant congenital heart diseases, confirmed diagnoses of cardiomyopathies and accessory pathways. Patients underwent 24-hours electrocardiography and echocardiography. LV function assessed with Teichholz (EF-T), Simpson biplane (EF-SB) methods; global longitudinal (GLS) and circumferential strain (CS) by speckle- tracking echocardiography. RV function assessed with RV fractional area change (RVFAC) and RV free wall longitudinal strain (RVFWLS). Statistical analysis performed with R software. Nominal variables tested for normal distribution with Shapiro-Francia test. Nominal values were presented with median, minimum and maximum. Welch test used to compare

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means between normally distributed nominal variables. Fisher exact test used to compare categorical variables. The p value <0,05 was considered statistically significant.
Results: Seventeen children, 4 (23,5%) boys, median age 12,5 years (6-17 years) meet inclusion and exclusion criteria. PVCs in 24-hour ECG were median 27% (20.3 - 37%). LV EF-T median 66.4 (55.5-78)%. LF EF-SB median 61.8 (55-69)%. LV GLS median -21.5 -(17.5-32.1)%. LV CS median -22.2 -(12.9 - 29.3)%. RVFAC median 49 (32-57)%. RVFWLS median -23 -(10,2-27,7)%.
Conclusions: Most children with frequent PVCs had left and right ventricular function within the normal range. One patient had parameters of RV dysfunction. However, patients with ≥30% PVCs had the decrease of the left and right ventricular myocardial strain parameters.

Keywords: premature ventricular complexes, ventricular function, myocardial strain imaging.

PP-471

The efficacy of flecainide compared to metoprolol in reducing premature ventricular complexes. An open label cross-over study in pediatric patients

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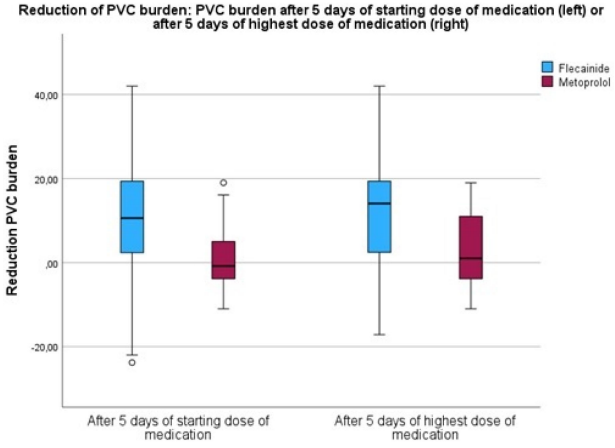
Background and Aim: Frequent premature ventricular contractions (PVCs) in children are rare, mostly asymptomatic and usually considered benign. Symptoms and/or left ventricular dysfunction can develop and are indications for treatment by anti-arrhythmic drugs (AAD). The efficacy of AAD in these children is not well known.
Method: In a prospective open label cross-over trial children with frequent PVCs and a PVC-burden of > 15% on Holter, were successively treated with metoprolol and flecainide or vice versa in a randomized manner. Patients started with metoprolol or flecainide, after 5 days steady state was assumed and basic measurements including PVC burden on Holter were repeated. After a drug free interval of at least two weeks, the drug was switched and measurements were repeated after 5 days of treatment.
Results: In total 62 patients were screened and 19 patients could be included after informed consent. Median age was 13.9 years (range +/- 10.2 years), 47% had symptoms, all had a normal function on echocardiography and a normal pro-BNP. Mean baseline PVC burden was 21.7 % (N=18, SD +/- 14.0) before the start of flecainide and 21.2 % (N=17, SD +/- 11.5) before the start of metoprolol. Mean reduction in PVC burden after 5 days of starting dose of medication was 10.4 percentage-points (SD +/- 17.7) for flecainide and 0.8 percentage-points (SD +/- 8.6) for metoprolol. In a mixed model analysis this led to a significant difference in estimated mean reduction of the PVC burden between flecainide and metoprolol of 8.2 percentage-points (95% CI of 0.86 – 15.46, P = 0.031). Nine out of 18 patients treated with flecainide had a PVC reduction of more than 5 percentage-points to a PVC-burden below 5% PVCs. These flecainide-responders had a mean trough plasma level of 0.34 mg/L (SD +/- 0.22), compared to a mean of 0.52 mg/L (SD +/- 0.37, P = 0.277) in the non-responders.
Conclusions: In children with frequent PVCs flecainide significantly reduces the PVC burden, contrary to metoprolol which does not reduce the PVC burden. Flecainide was effective in only a subgroup of patients, which appears to be unrelated to the trough plasma level.

Keywords: Premature ventricular complexes, anti-arrhythmic drugs, flecainide, metoprolol, child

Figure



Reduction of PVC burden

PP-472

A diagnostic enigma: two rare causes of sudden cardiac death in a child following out-of-hospital cardiac arrest

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Background and Aim: Cardiac syncope and aborted sudden cardiac death (SCD) are rare phenomena in paediatrics. Accurate diagnosis is crucial, can have implications for family screening, and a single identifiable cardiac cause is often detected. This case report describes a child with two rare, co-existing cardiac conditions, both of which are known to cause collapse or SCD.
Method: A case report.
Results: A 12-year-old boy presented with an out-of-hospital cardiac arrest precipitated by trampoline exercises. His initial arrest rhythm was ventricular fibrillation (VF), and he was successfully defibrillated. Prior to this, there was no significant past medical history or family history, aside from an episode of exercise-induced

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syncope that was previously unexplained. His transthoracic echocardiogram was reportedly normal, and his baseline 12-lead ECG was also normal. During further investigation, an ajmaline challenge was positive for a Type 1 Brugada pattern (Figure 1a). In view of his presentation, an implantable cardiac defibrillator (ICD) was inserted. Genetic testing was not performed due to its low diagnostic yield in Brugada syndrome. Three years later, he presented with near-collapse and chest pain whilst playing football. A 12-lead ECG demonstrated dynamic inferolateral ST depression with an elevated troponin T of 1690 ng/L. His ICD interrogation showed no evidence of arrhythmia. His admission echocardiogram raised the suspicion of an anomalous origin of the left coronary artery (LCA) from the right coronary cusp, and this was confirmed on CT angiography with a 5-6mm inter-arterial, intra-mural LCA course with reduced calibre (Figure 1b). He underwent unroofing of the intra-mural LCA and re-suspension of the aortic valve, with a slit-like LCA orifice seen intra-operatively. His post-operative admission was uncomplicated, and he was discharged home with his ICD remaining in-situ.

Conclusions: This is a novel case of co-existent Brugada syndrome and anomalous aortic origin of the left coronary artery in a child, not previously reported in the paediatric population. It demonstrates the importance of thorough investigation after cardiac syncope and the pitfalls of cognitive bias once a potential answer is found. Furthermore, it highlights the dilemmas in management when faced with two aetiologies of sudden cardiac death, considering the risks of invasive interventions in both conditions.

Figure 1

a) ECG demonstrating the characteristic type 1 Brugada pattern seen in the high chest leads V1-V3 (labelled V3R, V4R & V5R), 3 minutes after ajmaline infusion was complete.

b) CT demonstrating the anomalous origin and inter-arterial course of the left coronary artery from right coronary cusp, measuring 1.7 x 2.0 mm at its narrowest point.

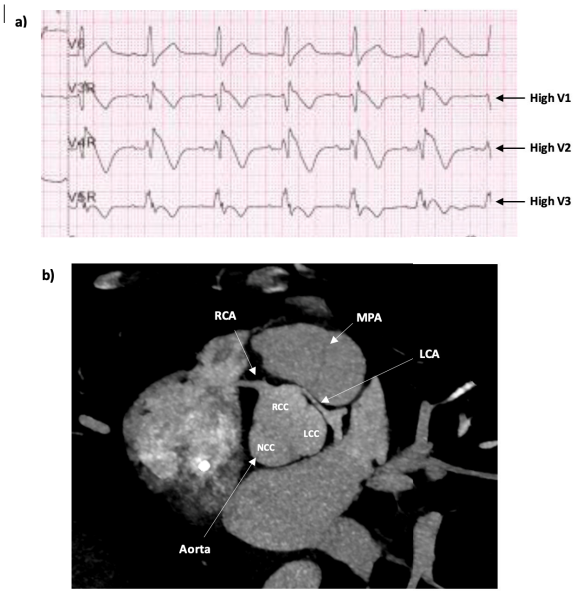

Figure 1 a) ECG demonstrating the characteristic type 1 Brugada pattern seen in the high chest leads V1-V3 (labelled V3R, V4R & V5R), 3 minutes after ajmaline infusion was complete. b) CT demonstrating the anomalous origin and inter-arterial course of the left coronary artery from right coronary cusp, measuring 1.7 x 2.0 mm at its narrowest point.

Keywords: Brugada syndrome, anomalous aortic origin of the left coronary artery, case report, paediatric, sudden cardiac arrest, ICD

PP-473

Unusual symptomatology in a classic atrioventricular nodal reentrant tachycardia pediatric case

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Background and Aim: Atrioventricular nodal reentrant tachycardia (AVNRT) is an arrhythmia caused by the coexistence of two nodal pathways: a slow one and a fast one. AVNRT is one of the most frequent causes of supraventricular tachycardia (SVT) in pediatric population after the first years of life. AVNRT prevalence is higher in older children who often describe palpitations, discomfort in the neck or chest, shortness of breath, dizziness. The aim of this case study is to emphasize the heterogenous symptomatology of arrythmias in pediatric population.

Method: We present the case of a 7 years old male patient with a 2 month history of recurrent vomiting. He had weekly episodes of vomiting that were not associated with any signs of infection nor with bad diet habits. Reportedly, at first, his skin would become pale, cold and sweaty and after 5-6 episodes of vomiting usually he would suddenly feel better. The patient was examined several times by a pediatrician after the vomiting and afterwards by a gastroenterologist who prescribed proton pump inhibitors. Despite the treatment the patient was admitted to the emergency room for a new episode of persistent vomiting. On examination he presented with intractable gagging, heart rate was 235bpm and his blood pressure 75/35 mmHg. Electrocardiogram showed narrow QRS reentrant tachycardia. Vagal maneuvers were unsuccessful, but a single dose of adenosine restored the sinus rhythm. The ECG recording during administration of adenosine showed non sustained monomorphic ventricular tachycardia before sinus rhythm was restored.

Results: The patient received chronic betablocker treatment to prevent recurrences up to 18 months. Afterwards, sudden frequent recurrences imposed a second line antiarrhythmic agent- in this case sotalol with little effect on preventing recurrences. In this clinical setting was referred for electrophysiology study. AVNRT diagnosis was confirmed and the slow nodal pathway was successfully ablated.

Conclusions: Arrythmias in pediatric population can have atypical symptomatology. Adenosine induced ventricular tachycardia is usually non sustained and self-limited. Recurrent episodes of AVNRT are more likely in older pediatric patient due to increased sympathetic tone.

Keywords: tachycardia, adenosine, vagal

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PP-474

Slow & fast - treatment of an AV-node in distress

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Background and Aim: Congenital heart block (CHB) is a rare, but well-known complication among pregnancies of mothers with autoimmune disease (about 2% of autoantibody-positive pregnancies, mainly Anti-SSA/Ro, Anti-SSB/La). Mortality rate is approx. 20% in newborns and 64% of patients will require a pacemaker over time. Clinical spectrum ranges from asymptomatic AV-block I° to (more often) high grade AV-block or other arrhythmias. Hydrops fetalis, endocardial fibroelastosis or valvular disease are feared comorbidities. Histologically an inflammation with subsequent fibrosis of the conduction system (especially the AV-node) has been demonstrated. Optimal management strategies are a matter of ongoing debate.
Method: At 23+3 weeks gestational age a 2:1 AV-Block (V-Rate 87 bpm, A-Rate 165 bpm) was observed in the foetus of a women with known Sjogrens-Syndrome. No other cardiac pathologies were noted and the diagnosis of CHB was made. Mother's therapy consisted of Hydroxychloroquine + ASS. SSA/Ro and SSB/LA antibodies were significantly elevated. Delivery in 39+3 weeks, uneventful postnatal transition. Heart rates were 60-80 bpm (AV-Block II°) with short episodes of intermittent sinus rhythm (150-170 bpm). The child was clinically asymptomatic and a wait-and-see approach was taken. In a regular control (1 month after birth) intermittent tachycardia up to 230 bpm was noted. Junctional ectopic tachycardia (JET) was suspected and therapy options were difficult putting the low baseline heart rate into account.
Results: Therapy with IVIG (2g/kg) was initiated and tachycardia improved rapidly. After another relapse (2 weeks later) another course of IVIG (1g/kg) was administered and no further JET-episodes occurred. Until today (3 years later) the patient is well and not taking any medication. In accordance with the current guidelines no pacemaker has been implemented yet. In the meantime, with repetitive immunoadsorption in pregnancy, the mother gave birth to a healthy girl.
Conclusions: Autoantibody induced inflammation of the conduction system might destroy AV-nodal cells and also induce ectopic tachycardia. In the difficult situation of AV block-related bradycardia and concomitant tachycardias therapy with IVIG can be a feasible option. More than 50% of the patients with CHB require a pacemaker and the time of implantation should be carefully determined. Furthermore, Immunoadsorption seems to be a preventive option in high-risk pregnancies.

Keywords: Congenital Heart Block, Pacemaker, IVIG, Immunoadsorption

PP-475

Outcome for children with inherited channelopathies in northern ireland between 2005 and 2023

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Background and Aim: The inherited Channelopathies are a diverse range of conditions caused by genetic mutations that predispose patients to arrhythmia and Sudden Cardiac Death (SCD). Examples include Congenital Long Qtc (LQT), Brugada (Br) and Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT). Most patients are identified with genetic screening, allowing early intervention. The inherited Cardiac Conditions Clinic (ICCC) was set up with this goal in mind, it serves the paediatric population of Northern Ireland
Method: A retrospective review of children attending the ICCC. Primary Outcomes evaluated included timing of diagnosis, method of presentation (screening vs acute referral), SCD, Arrhythmia/ Events, Genetic diagnosis, Medications and use of Automated Implantable Cardiac Defibrillator (AICD)
Results: 217 patients were identified. 194 had LQT (112 KCNQ1, 36 KCNH2, 13 SCN5A (LQT3 phenotype), 29 KCNE1, 2 Combo and 2 undefined). 18 Patients had Br, 5 had CPVT and 1 complex patient. The average age of diagnosis of LQT was 50 months (range 0-192). 95% of patients were diagnosed via screening. The average age of diagnosis of Br was 120 months (36-216), 88% were diagnosed via screening, 55% of patients with Br had a genetic diagnosis. The 5 patients with CPVT were all diagnosed via acute presentation/ post mortem. There were five SCDS (3 CPVT patients, 2 LQT Patients) all of these patients were diagnosed post-mortem and were not previously known to the ICDC clinic. LQT patients received monotherapy with Beta blockers primarily, conversely 33% of the SCN5A group received combination therapy with a Beta blocker and Mexiletine. The Brugada cohort were primarily unmedicated and the CPVT group varied. Two patients had AICDS implanted (1 LQT and 1 Complex patient).
Conclusions: The majority of patients with Channelopathies at ICCC clinic are initially referred following diagnosis of a proband. They receive an accurate clinical and genetic diagnosis that prompts treatment where indicated. They also receive lifestyle advice appropriate to their diagnosis and have ongoing telephone access to specialist nurses.
Reassuringly the vast majority of these children remain free of serious adverse events. They rarely require acute intervention or placement of AICDS. Unfortunately, there is a cohort of patients (primarily CPVT) who are mostly diagnosed post-mortem.

Keywords: Channelopathies, Arrythmias, SCD

PP-476

Substrates for ventricular tachycardia in patients after repaired tetralogy of fallot

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Background and Aim: Patients after repaired Tetralogy of Fallot (rTOF) have an increasing risk of ventricular tachycardias (VT). Monomorphic VT in this patient population is in the majority of cases related to slow conducting anatomic isthmuses (SCAI). We report our data of risk stratification in this patient population.
Method: All adult patients with rTOF undergoing electrophysiological study (EPS) with risk stratification for VT from January 2019 until September 2023 were included. In all procedures right ventricular voltage and activation mapping during sinus rhythm was performed to identify SCAI (conduction velocity <0,5 m/s) prior to VT induction (up to S4 180 ms). Ablation was performed with an open irrigated tip catheter.

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Results: 41 procedures in 39 patients (mean age 43 yrs, 15 females) were performed. In primary procedures SCAI was present in 29 patients (74%). SCAI was present more frequently if EPS was performed later after intracardiac repair (37.5±9.3 yrs vs. 28.6±6 yrs, p=0.02). VT was only inducible in patients with SCAI (15/29 (52%) p=0.006), thereof 2 different VTs in 4 patients. Indication for primary EPS were sustained VT in one patient, non-sustained VT in 19 patients, and risk stratification only in 19 patients. Ablation was performed in 32 procedures (29 primary procedures with SCAI, one procedure without SCAI, but extended scar and 2 secondary procedures). Ablation was performed most frequently of isthmus 3 (between the pulmonary valve and the VSD patch) in 31/32 procedures and isthmus 4 (between the VSD patch and the tricuspid anulus) in 30/32 procedures. Ablation of all VTs was possible in 13/15 patients (87%). In the remaining two patients after unsuccessful procedure and in one other patient ICD-implantation was recommended/performed. During a mean follow-up of 19.5 months one patient had recurrence of sustained VT/VF (a patient with non-successful ablation of SCAI 3 and consecutive ICD implantation). No complications occurred.
Conclusions: SCAI were present in the majority of patients after rTOF. The most frequent SCAI were isthmus 3 and isthmus 4. The success rate of 87% was high and recurrence of VT/VF occurred only in one patient after unsuccessful ablation. ICD implantation was indicated in 3/39 patients.

Keywords: Tetralogy of Fallot, Ventricular Tachycardia, Slow conducting anatomic isthmus, Risk stratification, Ablation

PP-477

Mobile ECG devices in pediatric cardiology: Efficacy and accuracy in healthy children

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Background and Aim: Pediatric arrhythmias require accurate electrocardiogram (ECG) assessment for diagnosis and management. Mobile ECG devices, such as the Apple Watch and Cardia Mobile 6L, offer potential advantages in terms of portability and accessibility. This study aimed to compare the efficancy and diagnostic accuracy of ECG tracings obtained using these mobile devices with standard 12-lead ECG tracings in healthy children.
Method: ECG data were acquired from healthy pediatric subjects aged 4 to 18 years using the Apple Watch, Cardia Mobile 6L, and standard 12-lead ECG (n=30). Single leads were acquired sequentially. The time required for ECG acquisition was recorded for each device. Subsequently, ECG leads were analyzed for voltage, duration, and morphology of P waves, QRS complexes, T waves, PR intervals, and ST segments. Assessment was performed by two blinded pediatric electrophysiologists.
Results: We found significant differences in the time required to acquire ECG tracings between the Apple Watch, standard 12-lead ECG, and Cardia Mobile 6L, with the Apple Watch being notably slower (median 15 min, 4 min, 1 min, respectively). However, the analysis revealed that ECG leads obtained with the Apple Watch demonstrated comparability to standard 12-lead ECG tracings in terms of key parameters, including voltage (mV), duration (ms), and morphology of cardiac waveforms. Qualitative and quantitative assessment was performed for all leads.
Conclusions: This study suggests that mobile ECG devices offer a diagnostic accuracy comparable to that of standard 12-lead ECGs in healthy children. While the Apple Watch may be slower in ECG acquisition, acquisition of all 12 standard ECG leads is feasible in this population. Its potential for pediatric usage in specific clinical scenarios, such as remote monitoring or emergencies, is promising.

Keywords: Pediatric Arrhythmias, Mobile ECG Devices, Diagnostic Accuracy, Pediatric Cardiology, ECG Tracing Efficiency

PP-478

Facilitation of transseptal puncture by means of sharp transseptal guidewire – single center experience in children undergoing catheter ablation

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Background and Aim: Transseptal puncture (TSP) for catheter ablation of left-sided arrhythmogenic substrates is in adults preferably performed under intracardiac echocardiographic (ICE) guidance. However, the large 11F sheath necessary for ICE catheter is challenging in children. On the other hand, left atria (LA) in children are rather small and therefore more prone to complications. Transseptal guidewire (TSG) of 0.35 mm thickness facilitates the TSP by using a J-shaped retroflecting very sharp tip. This design ensures smooth septal penetration with immediate protective retroflection of the tip after entering a LA in order to minimize risk of injury. Such approach increases safety of TSP and comfort of the operator. Study summarizes experience with TSG (SafeSept, Pressure Products, Inc.) for accessing left sided arrhythmogenic substrates in children.
Method: Retrospective analysis of 18 children who underwent TSP using TSG during a period of 2 years (3/2021- 3/2023). 16 children had ablation of left-sided accessory pathways and 2 children with intraseptal pathways had mapping of the LA before eventual ablation from the right side. The median age of children was 13.5 years (6 – 19 years), median weight 57,5 kg (19 – 111 kg). TSG was inserted into the lumen of standard Brockenbrough needle and then introduced into fixed curve or steerable sheath (SL-1 or Agilis, Abbott Inc., USA). Position of the sheath at the presumed fossa ovalis was verified in left and right oblique fluoroscopic projections. TSG was pushed out of the needle and dilator to perforate interatrial septum. After entering LA cavity, TSG tip "retrocurls" to create an atraumatic J-shape. Radiopaque coil at the shaft was visualized by fluoroscopy and distal J-shaped guidewire tip was inserted into left superior pulmonary vein. After removing

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TSG, needle and dilator, ablation catheter was introduced into LA. Neither LA pressure measurements nor contrast injection were used for verification.
Results: TSG facilitated TS puncture was uneventfully performed in all children. There were no acute or late complications.
Conclusions: Our single-centre experience suggests that TSG facilitated TSP is a practical approach for increasing safety of left-sided catheter ablations in children and adolescents especially in low-volume paediatric EP centres, eliminating the need for ICE guidance.

Keywords: catheter ablation, left-sided arrhythmogenic substrate, transseptal puncture, transseptal guidewire

PP-479
Alternative solution in failed endocardial ablation with severe pediatric arrhythmia; epicardial approach

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Background and Aim: Despite the high success rate of endocardial ablation in pediatric patients, conventional ablation may fail in some resistant cases. This failure is likely attributed to an arrhythmogenic substrate originating from an epicardial focus. In such cases, percutaneous epicardial ablation may be necessary. Herein, we present pediatric patients who underwent epicardial ablation at our center over a 10-year period.
Method: Between November 2013 and November 2023, out of a total of 2360 cases undergoing ablation with the 3D electroanatomical mapping method (EnSite System, Abbott ING), five pediatric patients with arrhythmias resistant to medical treatment and failed attempts of endocardial ablation underwent epicardial ablation. In four patients, access to the pericardial space was achieved through a subxiphoid approach using a Tuohy needle, while in one patient, a small incision was made for surgical thoracotomy. Irrigated radiofrequency (RF) energy was used in all cases. Patient data, including pre- and post-procedural records, were obtained from our clinic's database.
Results: The median age of the patiens was 11 years (range: 8-17). Three of the patients were male. Four patients underwent the procedure due to treatment-resistant ventricular arrhythmia (ventricular tachycardia (VT)/premature ventricular contractions (PVC)) and/or repetitive ICD shocks), while one patient presented with a very high-risk manifest accessory pathway (Wolff-Parkinson-White (WPW) syndrome) and aborted sudden cardiac death. Among the VT patients, one had biventricular involvement of arrhythmogenic right ventricular dysplasia (ARVD), one had fulminant myocarditis and resistant VT-associated ICD storm, and the other two had frequent PVCs and non-sustained VTs leading to tachycardiomyopathy (Table 1). All patients had a history of failed endocardial ablation either before or during the same procedure. All procedures were successful without major complications. Blood transfusion was administered in only one patient due to persistent pericardial effusion post-procedure. The pericardial catheter was removed as effusion ceased from the second day after the procedure. The intensive care unit stay duration for patients was 2 days, with a median total hospital stay of 9 days (range: 8-14).
Conclusions: Percutaneous epicardial catheter ablation is a safely applicable method in experienced centers for high-risk pediatric patients with epicardial arrhythmogenic foci where endocardial ablation has proven unsuccessful.

Keywords: arrhythmogenic right ventricular dysplasia, children, epicardial ablation, ventricular tachycardia, WPW

Table 1. Demographic data and procedure characteristics of the patients

Patient no	Age (yr)	Gender	Weight (kg)	Diagnosis	Ablation location	Energy type	Result
1	16	K	41	Fulminant myocarditis + scar-related refractory VT and ICD storm	left ventricle anterior, anteroseptal, posterobasal, apical (endocardial + epicardial)	irrigated RF (Tacticath)	Suboptimal + scar homogenization
2	8.5	E	18	WPW, Aortic valve pathology, Aborted sudden death	left posterolateral (endocardial + epicardial)	irrigated RF	Successful
3	11	K	42	ARVD – VT/ICD shock	RVOT anterior and tricuspid valve anterolateral (endocardial) Left ventricle posterobasal (epicardial)	Irrigated RF	Successful
4	8.5	E	27	Mitral valve plasty + DKMP – CRT-D, Resistant frequent PVCs-VT	left ventricle basolateral (close to mitral valve) + anterolateral (endocardial + epicardial)	irrigated RF (Tacticath)	Successful
5	17	E	56	Tachycardiomyopathy/VT	left ventricle apical posteroseptal (endocardial + epicardial)	irrigated RF (Tacticath)	Successful

ARVD: Arrhythmogenic right ventricular dysplasia, CRT: cardiac resynchronization therapy DKMP: dilated cardiomyopathy, ICD: intracardiac defibrillator, RF: radiofrequency, PVC: premature ventricular contractions, RVOT: right ventricular outflow tract, VT: ventricular tachycardia, WPW: Wolff-Parkinson-White.

PP-480
Early postoperative arrhythmias following complete surgical repair of truncus arteriosus in children

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Background and Aim: This study examines arrhythmia issues emerging in the early postoperative period of patients undergoing surgical correction for Truncus Arteriosus (TA).
Method: Surgical correction operations were performed on a total of 29 patients between the years 2013 and 2023. Data were obtained by reviewing medical records, including the period from the initial consultation to repair surgery, intensive care and service follow-up, and the final cardiology monitoring.
Results: Of the 29 TA cases, 24 (82.7%) were type-1, 4 (13.8%) were type-2, and 1 (3.4%) was type-3. Among the repaired patients, 18 were female (62%). The median age of the patients was 30 days (2-90 days), and the median body weight was 4 kg (2.1-6.9 kg). Postoperatively, arrhythmia developed in 13/29 patients (44.8%) (Table 1). Among them, 6/29 (20.7%) had ventricular tachycardia and ventricular fibrillation, 5/29 (17.2%) had AV block (2 with complete AV block requiring permanent pacemaker implantation), 1/29 (3.4%) had junctional ectopic tachycardia, and 1/29 (3.4%) had supraventricular tachycardia (Focal atrial tachycardia). The average onset time of arrhythmia was 2 days post-surgery (1-16 days). 6 patients (20.7%) required ECMO, all of whom had arrhythmia issues. Cardiac arrest occurred in 5 patients (17.2%), all with concurrent arrhythmias. Postoperative early mortality was 5/29 (17.2%), all of whom had arrhythmias. The median average cross-clamp time for all patients was 171 minutes (99-350 minutes), while for those with arrhythmias, it was 184 minutes. The total aortic clamp time for all patients was 124 minutes (79-172), and for those with arrhythmias, it was 127 minutes.

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Conclusions: Early postoperative arrhythmias after complete repair of TA are seen very frequently. Especially ventricular arrhythmias during the early period appear to be associated with serious morbidity and mortality.

Keywords: Truncus Arteriosus, Early-postoperative, Arrhytmia, Pediatric

Table 1: Patients with early-postoperative arrhytmia after complete repair of Truncus arteriosus, demographic and clinic data

Patient with arrhytmia	Sex(F/M)	Age(day)	Body weight(kg)	Type of Truncus arteriosus	Post-op arrhtmia time(day)	Post-op arryhtmia type	ECMO (Yes/No)	Exitus (Yes/No)
Patient 1	F	18	3,5	Type 1	16	JET	No	No
Patient 2	F	60	4,6	Type 1	7	Complete AV Block	No	No
Patient 3	F	60	4	Type1	2	AV Block	Yes	Yes
Patient 4	F	2	2.4	Type 1	1	VF-Cardiac Arrest	No	Yes
Patient 5	F	30	5,1	Type1	1	Complete AV Block	No	No
Patient 6	M	12	2.1	Type2	2	AV Block	Yes	No
Patient 7	F	30	3,3	Type1	11	VF-Cardiac Arrest	Yes	Yes
Patient 8	M	30	3,7	Type1	2	VT-VF-Cardiac Arrest	Yes	Yes
Patient 9	F	45	4,3	Type1	9	VT	Yes	No
Patient 10	F	30	5,5	Type3	8	AV Block-ST depression	No	No
Patient 11	F	21	3,2	Type1	1	VT-VF-Cardiac Arrest	No	No
Patient 12	M	21	4	Type1	1	SVT(FAT)	No	No
Patient 13	M	21	3,5	Type2	1	VT-Cardiac Arrest	Yes	Yes

Patients with early-postoperative arrhytmia after complete repair of Truncus arteriosus, demographic and clinic data

PP-481

Cardiac hemachromatosis: a case with complete av block only

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Background and Aim: Complete AV block may be congenital in childhood or may develop due to acquired reasons. In this case report, a patient who developed complete AV block due to cardiac hemochromatosis, a rare etiology, is presented.
Method: A 7 years and 2 months old, 17 kg (<3 percentile), 113cm (3-10 percentile) male patient was referred to us from Syria due to complete AV block. It was learned that the patient had a diagnosis of beta thalassemia major and ES transfusion was administered to the patient at 3-week intervals in Syria. The patient has a complaint of fatigue that has increased recently. There was no pathology in his physical examination other than growth and development retardation. In the examinations performed, Hb:9gr/dL Htc:% 26 WBC:6360 PLT:322000 AST:214U/L ALT:218U/L BUN:8mg/dL Cre:0.29mg/dL Ferritin >2000ng/mL NTpro-BNP:40pg/ mL detected. The patient's ECG was well compatible with AV complete block and a ventricular rate of 57/min(Figure 1). In the ECHO examination, measurements were normal: LVIDd: 37mm(z score:+1.27) LVIDs:21 mm(z score:+0.26) FS: %42 EF: 73% was detected, 24-hour Holter ECG showed ventricular escape rhythm in 60-70%, also QTc values were 460-480 ms, and polymorphic VES. In Holter ECG, max-min-average heart rates were 84-48-57/min. In CMR, T2*, native T1 and T2 values of the left ventricular myocardium were observed to decrease in accordance with iron accumulation and the T2* value of the left ventricular myocardium was determined to be 15.8 ms. This finding is consistent with mild iron accumulation in the heart.
Results: The patient underwent transvenous single-chamber permanent ventricular pacemaker implantation (Figure 1) because of the patient weight and vascular dimensions. The patient was consulted with the relevant units for follow-up of other visceral organ involvements and thalassemia.
Conclusions: Hemochromatosis due to frequent transfusion is a complication that can be slowed/prevented by iron chelation therapy and often begins in visceral organs, especially the liver. Therefore, the development of cardiac hemochromatosis is a rare condition and presents primarily as asymptomatic or dilated cardiomyopathy. This case re-emphasized that in rare cases, the heart's conduction system can be affected before cardiac functions are affected.

Keywords: Complete AV Block, Hemochromatosis

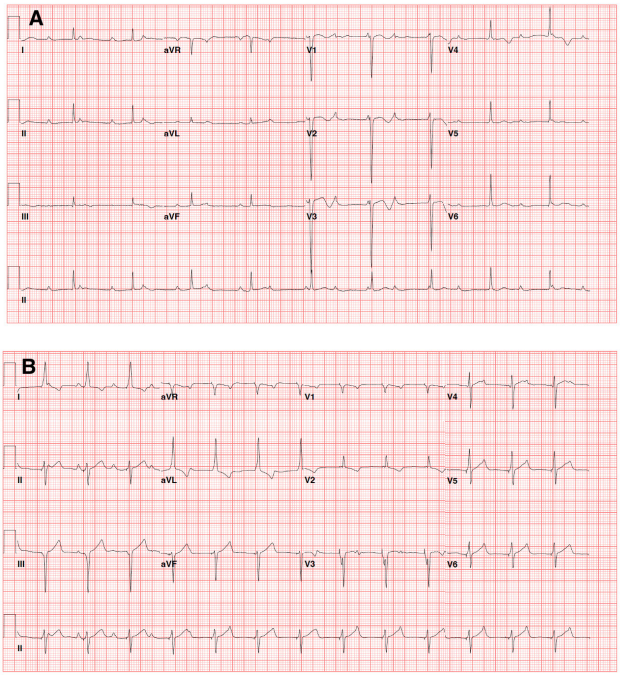

Figure1: Complete AV Block(ECG:A) and After Pacemaker Implantation (ECG: B)

PP-482

Postoperative arrhythmic burden in children after isolated left ventricle outflow tract obstruction surgery

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Background and Aim: This study planned to investigate arrhythmias after left ventricle outflow tract obstruction (LVOTO) surgery.
Method: In this study, 70 patients were enrolled who underwent LVOTO surgery except hypertrophic obstructive

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cardiomyopathy, complex congenital heart diseases (CHD) and Shone complex, between January 2013 and October 2023. Surgery was classified in 3 groups; simple resection(subaortic discrete or fibromuscular membrane or subaortic ridge resection), simple resection+myectomy and agressive resection (Modified Konno, Ross-Konno). Preoperative and postoperative ECGs of the patients were retrospectively scanned from the database.
Results: 42/70 (60%) of patients were male. Mean operation age was 7, 6 years (range 1 months-19,4 years). 53 (75.7%) patients underwent simple resection, 8 (11.4%) patients underwent simple resection+myomectomy, 9 (12.8%) patients underwent agressive resection. 26 (37,1%) patients had a previous surgical history; 6 patients had previous VSD closure, 5 patients had VSD-arcus hypoplasia repair, 8 patients had aortic pathologies surgery and 7 patients had different types of LVOTO surgery before. Preoperative ECGs showed RBBB in 12 patients, LBBB in 2 patients and left ventricular hypertrophy criteria in 30 patients. Sinus node dysfunction (SND) was present in 1 patient and Mobitz type 2 AV block was present in 1 patient. Remaining 24 patients had normal ECG. Postoperative ECGs of patients revealed new LBBB in 18 patients, new RBBB in 4 patients. Junctional ectopic tachycardia (JET) was observed in 1 patient, 3rd degree AV block in 9 patients, the patient with preoperative Mobitz type 2 block rhythm persisted after surgery. Pacemaker implantation was performed in 8 patients with 3rd degree AV block at a mean of 8.1 (range: 5-13) days postoperatively. One patient with sustained VT/VF and AV block was implanted ICD on postoperative 7th day. In 2/18 patients with postoperative LBBB, dyssynchrony and LV-dysfunction developed in the late period. The younger patient is still under follow-up, while the other patient underwent CRT-D implantation at postoperative 6th year.
Conclusions: Postoperative AV block is seen more frequent in LVOTO surgery according to other CHD-surgery and frequently irreversible. Myectomy, agressive surgeries like Konno and concomittant mitral surgery may be a risk factor for permanent-AVB. Follow-up of patients with postoperative LBBB is important in terms of the development of dyssynchrony and the need for CRT in the long term.

Keywords: left ventricle outflow obstruction, AV block, subaortik ridge, Ross-Konno

image

Table 1: Demographic and Clinical Features of the Patients

Patients n (%)	71
Operation Age, median in month (range)	91,5(1.233)
Death, n (%)	2(2,8)
Sex	
Female, n (%)	28 (40)
Male, n (%)	42(60)
Diagnosis	
Isolated LVOTO	38 (54.2)
LVOTO + aortic stenosis	26 (37.1)
LVOTO + VSD	4 (5.7)
Previous Surgery	
VSD closure, n(%)	6(8,5)
VSD closure+Arcus reconstruction, n(%)	5(8,5)
CoA and arcus repair ,n(%)	8(11,5)
LVOTO surgery, n(%)	7(11,4)
Preoperative ECG n (%)	
Normal	24 (34.3)
RBBB	12(17.1)
LBBB	2(3,3)
VSD	1(1,4)
Mobitz Type 2 AV Block	1(1,4)
Surgery	
1. LVOTO surgery	
Simple resection n(%)	53(75.7)
Simple resection+myectomy n(%)	8(11.4)
Agressive resection n(%)	9(12.8)
2. Concomitant surgery	
Aort valve repair n(%)	19(27.1)
Mitral valve repair n(%)	3(4.2)
VSD closure n(%)	4(5.7)
Postoperative Arrhythmias	
LBBB	20(28.5)
RBBB	15(21.4)
SND	1(1.4)
Mobitz Type 2 AV Block	1(1.4)
Complete AV block	8(11.4)
Junctional Ectopic Tachycardia	1(1.4)
Ventricular Arrhythmia (VT/VF)	1(1.4)

Table 2: Demographic and Clinical Features of the Patients with post-op AV block

Patient No	Sex(Female:F, male: M)	Surgery age(month)	Diagnosis	Previous surgery	Current surgery
1	M	51	LVOTO	VSD closure+Arcus repair	Subaortic ridge resection(single)
2	M	77	LVOTO	LVOTO closure+Arcus reconstruction+ 2Subaortic ridge resection	Subortic Ridge resection + Myectomy
3	F	13	LVOTO+VSD	-	LVOTO repair + septal myectomy + VSD closure
4	M	31	LVOTO+Mitral stenosis	CoA repair	Subaortic ridge resection +myectomy+ Mitral valve repair
5	F	214	LVOTO+Mitral pathology	VSD closure	Subaortic ridge resection+myectomy+mitral repair
6	M	13	LVOTO+Aort stenosis	CoA repair	Ross-Konno procedure
7	M	128	LVOTO	-	Subaortic membran resection+septal myectomy
8	M	103	LVOTO+ Aort stenosis	-	Modified Konno+AVR+myectomy
9	F	31	LVOTO+ Aort stenosis	VSD closure+arcus reconstruction	Modified Konno+subaortic ridge resection

Table 1: Demographic and Clinical Features of the Patients Table 2: Demographic and Clinical Features of the Patients with post-op AV Block

PP-483

Management of unpredictable complication after transcatheter vsd closure; CRT for left bundle branch block induced dyssynchrony

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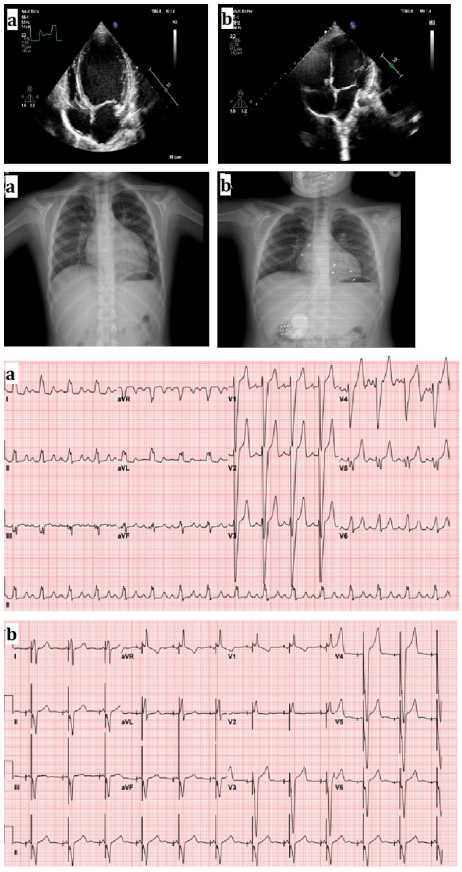
Background and Aim: Percutaneous closure of perimembranous ventricular septal defects (VSD) is increasingly favored over surgical approaches due to its minimally invasive nature. However, this technique can lead to arrhythmias, including left bundle branch block (LBBB), causing dyssynchrony and left ventricular dysfunction. This abstract presents three pediatric cases demonstrating the effectiveness of cardiac resynchronization therapy (CRT) in managing these complications.
Method: Case 1: A 7.5-year-old, 35 kg male with a history of ventricular septal aneurysm and VSD, underwent closure using a 12*10 mm Amplatzer Duct Occluder. 26 months post-procedure, he exhibited fatigue, breathlessness, and exercise intolerance. Electrocardiogram (ECG) revealed LBBB with a QRS duration of 150 ms. The echocardiogram revealed interventricular and intraventricular dyssyncrony, left ventricular dilation with systolic dysfunction (LVEF 37 %). Post-CRT implantation (after 6 months), significant improvement in systolic function (LVEF 68 %) and dyssynchrony (QRS duration 102ms) was observed.
Case 2: A 12-year-old, 40 kg male with a history of VSD closure using an Amplatzer muscular device at age five experienced progressive aortic insufficiency and developed LBBB three years post-procedure. He referred to our center with shortness of breath, weakness, and fatigue. At presentation, the QRS duration was 170 ms, and echocardiography showed significant aortic valve regurgitation, left ventricular dilatation, significant dyssynchrony and systolic dysfunction (LVEF 30 %). The patient underwent aortic valve

Pre- and post-procedural echocardiography, PA chest X-ray, and ECG of the third patient



a:Pre- procedural echocardiography, PA chest X-ray, and ECG of the third patient b:Post-procedural echocardiography, PA chest X-ray, and ECG of the third patient

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surgery with CRT implantation. 30 months post-surgery the exhibited improved cardiac function (LVEF 65 %, QRS duration 110 ms). Case 3: A 6.5-year-old, 21 kg male, with a history of perimembranous VSD closed using an Amplatzer device two years prior, presented with fatigue, and exercise intolerance. Diagnosis of LBBB (QRS 132 ms) and left ventricular dysfunction (LVEF 29 %) was made via ECG and echocardiography. The patient underwent successful epicardial CRT implantation, showing improvement in systolic function (LVEF 65%) and dyssynchrony (QRS duration 96 ms), post-procedural 9 months.
Results:
Conclusions: CRT is an effective therapeutic intervention in pediatric patients developing LBBB, dyssynchrony, and left ventricular dysfunction following percutaneous VSD closure. These cases highlight the importance of long-term monitoring for arrhythmic complications post-VSD closure and demonstrates the efficacy of CRT in reversing the associated cardiac dysfunctions.

Keywords: LBBB, Dyssynchrony, CRT

PP-484
Early postoperative arrhythmias in children with tetralogy of fallot

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Background and Aim: This study is designed to investigate the occurrence of early postoperative arrhythmias in the pediatric population following complete repair for Tetralogy of Fallot (TOF).
Method: The study enrolled a cohort of 275 pediatric patients who underwent cardiac surgery for TOF between January 2019 and October 2023. This investigation systematically assessed the occurrence of postoperative arrhythmias, delving into diagnostic patterns, identifying potential risk factors, and scrutinizing the strategies employed for effective management within this specific population.
Results: After surgery, the majority of patients exhibited right bundle branch block (RBBB) [isolated or with left anterior hemibloc, 201/273(73%), 35/275, (13%), respectively]. Clinically important early postoperative arrhythmias were diagnosed in 31 patients (%11.2), the prevailing one being junctional ectopic tachycardia (JET). The median age of post-operative JET patients was 9.5 months (IQR:6,7-17,25) and the median weight was 8 kg (IQR: 6,4 -10,0). Of the patients with JET, medical treatment was deemed necessary for 21 out of 22 cases (95%). Treatment modalities included the initiation of amiodarone-alone in 11 patients, ivabradine-alone in 6, and a ivabradine+amiodarone-combination in four. Additionally, the mean duration for the termination of JET rhythm was 34.7 (±13.3) hours (Table 1). After surgery the second frequent arrhythmic complication was complete atrioventricular (AV) block (7/275, 2.5%). The median age was 13 years (IQR: 6-103), and the median weight was 8 kilograms (IQR: 6-20) (Table 2). In five (5/7, 71%) patients, the block resolved spontaneously in ten days follow-up, while one patient required the implantation of a permanent pacemaker on the 14th day and one patient on ECMO awaiting for pacemaker implantation died due to other hemodynamic problems on the 13th postoperative day. The mean duration of intensive care unit (ICU) stay for patients with post-operative AV block was 12.9 (±8.7) days. Additionally, one patient had a VF attack while under ECMO support.
Conclusions: This study demonstrates that clinically significant arrhythmias still occur with a certain frequency (almost 15% of cases) in the early postoperative period of TOF patients in the current era. Early diagnosis and appropriate treatment selection appear to be crucial in reducing mortality.

Keywords: tetralogy of fallot, congenital heart disease, early postoperative arrhythmia

Table 1 and Table 2

Table 1: Demographic and Clinical Features of the Patients with post-op AV block

Demographic and Clinical Features of the Patients with post-op AV block	Total
Patients, n (%)	7 (100)
Age, median (months) (25-75p)	13 (6-103)
Death, n (%)	1 (14,3)
Sex	
Female, n (%)	5 (71,4)
Male, n (%)	2 (28,6)
Weight, median (kg) (25-75p)	8 (6-20)
Duration of ICU stay (day), mean (SD)	12,9 (±8,7)
Cross-Clamp time, minute, mean (SD)	113,1 ± (37,2)
Cardiopulmonary bypass time, minute, mean (SD)	174 (± 49,6)
Hemodynamic instability	4 (57,1)
Need for peritoneal dialysis	4 (57,1)
Need for permanent pacemaker	1 (14,2)

Table 2: Demographic and Clinical Features of the Patients with post-op JET

Demographic and Clinical Features of the Patients with post-op JET	Total
Patients, n (%)	22 (100)
Age, median (months) (25-75p)	9,5 (6,7-17,25)
Sex	
Female, n (%)	9 (41)
Male, n (%)	13 (59)
Weight, median (kg) (25-75p)	8 (6,4-10)
Duration of ICU stay (day), median (25-75p)	5,5 (3-7,3)
Cross-Clamp time, minute, median (25-75p)	84 (73-120,5)
Cardiopulmonary bypass time,minute, mean (±SD)	144,3 (± 42,9)
Hemodynamic instability	7 (31,8)
Need for peritoneal dialysis	7 (31,8)
Duration of termination of JET,hours,mean (±SD)	34,7 (±13,3)

PP-485
Feasibility of catheter ablation with zero-fluoroscopy in patients with congenital heart disease

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Background and Aim: Our aim was to compare the safety and outcome of zero-fluoroscopy (ZF) and fluoroscopy (non-ZF) catheter ablation (CA) in congenital heart disease (CHD) patients (pts).
Method: This is a single-center retrospective study of CHD pts who underwent CA at the Gottsegen National Cardiovascular Center between 2006-2020. The ZF and non-ZF groups were compared for the complexity of anatomy, arrhythmia substrate, difficulties of vascular access, transseptal or transbaffle puncture, presence of intracardiac pacemaker or ICD, procedural parameters, complications, and acute outcome. Near-ZF CAs (effective dose ≤1 mSv) were assigned to the ZF group.
Results: The ZF group included 22 pts (mean age: 22±13 years) and the non-ZF group 48 pts (mean age: 31±12 years). The distribution of CHD anatomical complexity (mild/moderate/severe) was 18.2%-68.2%-13.6% in the ZF group and 14.6%-68.7%-16.7% in the non-ZF group. Arrhythmia diagnoses were focal atrial tachycardia (ZF: 4.6%), atrial flutter (ZF: 45.4%, non-ZF: 75.0%), atrioventricular reentry tachycardia (AVRT) via manifest accessory pathway (AP) (ZF: 31.8%, non-ZF: 10.4%), AVRT via concealed AP (ZF: 4.6%, non-ZF: 8.3%), and atrioventricular nodal reentry tachycardia (ZF: 13.6%, non-ZF: 6.3%). Difficulties in vascular access occurred in 4.5% of the ZF group and 18.8% of the non-ZF group (p=0.10). The rate of transseptal/transbaffle puncture was significantly higher in the non-ZF group (22.9% vs. 4.6%, p=0.04). In the ZF group 13.6% and in the non-ZF group 20.8% of pts had pacemaker or ICD (p=0.22). There was no significant difference in procedure time, ablation time and the number of radiofrequency applications (ZF group: 169±53 min, 1205 ±906 sec, 28±15 vs. non-ZF group: 203±74 min, 1459±940 sec, 34±16, p≥0.05). The acute success rate was 100% in both groups. No complications occurred in any group.

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Conclusions: Zero fluoroscopic CA was feasible in 1/3 of the pts with CHD and as safe and effective as fluoroscopic ablation. No significant differences were found in the non-fluroscopy related procedural data between the two groups.

Keywords: catheter ablation, zero fluoroscopy, congenital heart disease

PP-486

Evaluation of cardiac arryhmia in the patients with left ventricule assist device (L-VAD)

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Background and Aim: Pediatric heart failure is an important cause of morbidity and mortality in childhood. Ventricular assist devices(VAD) are used for bridging to transplantation in patients with severe heart failure. In this study, we aimed to evaluate the cardiac arrythmias in the chidren with left ventricular assist device(L-VAD)
Method: The study included 33 children aged 0-18 years who were implanted with L-VAD for advanced heart failure in our hospital between January 2009 and January 2023. we retrospectively evaluated the files of the patients with L-VAD.
Results: Eleven patients (33%) developed atrial or ventricular arrhythmia during follow-up after left ventricular assist device (LVAD) implantation. Demographic findings, echocardiogram parameters, and angiography results were compared between patients with and without arrhythmia to determine their relationship with the risk of developing arrhythmia. Of these, only increased left ventricular end-diastolic diameter was associated with the development of cardiac arrhythmia (p=0.014). For the other parameters, there was no significant difference between the two groups suggesting an impact on the risk of developing arrhythmia.A total of 5 patients underwent cardioversion (n=2) or defibrillation (n=3) because of arrhythmia. Ten patients who developed arrhythmia received medical treatment with amiodarone, mexiletine, beta-blocker therapy, propranolol, metoprolol, or sotalol.
Conclusions: LVADs are life-saving in patients with terminal heart failure, but the risk of complications increases with longer follow-up time. One of the most common of these complications is arrhythmia. Patients with an LVAD should be closely monitored for arrhythmia, even if asymptomatic

Keywords: Arryhmia, L-VAD, children

Cardiac Imaging

PP-487

Lung ultrasound is a useful method for evaluating pulmonary congestion in congenital heart disease

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Background and Aim: Ventricular septal defect (VSD) is a disease that causes excessive pulmonary blood flow resulting in pulmonary congestion. However, estimating its severity to determine the need for surgical repair is sometimes difficult. We tested the usefulness of lung ultrasound in evaluating pulmonary blood flow: systemic blood flow ratio (Qp/Qs).
Method: We prospectively studied 20 consecutive patients with VSD with or without atrial septal defect (ASD) at the age of less than 5 years old who underwent cardiac catheterization at our institution. Echocardiogram and lung ultrasound were evaluated one day before the day of catheterization. The number of B-lines in an intercostal space was measured in 4 areas (right and left, prothoracic and lateral thoracic region) of the chest wall of each patient using a linear probe. We evaluated the correlation between the average of B-line density, which was calculated as the number of each B-line divided by the width of the space, and other hemodynamic variables.
Results: The diagnosis of the patients was isolated VSD in 18 patients, VSD with ASD in 2 patients. Median age at ultrasound and catheterization was 0.54(0.35-4.05) (interquartile range) years, median Qp/Qs was 2.35(1.36-2.70), median mean pulmonary arterial pressure (PAP) was 20.0(14.8-31.5) mmHg, and median B-line density was 0.29(0.12-0.50)/mm. There was a significant correlation between B-line density and Qp/Qs ($p<0.001$, $R^2=0.67$) and mean PAP ($p<0.001$, $R^2=0.51$). The correlation was more pronounced than that between Qp/Qs and brain natriuretic peptide ($p<0.05$, $R^2=0.32$), Qp/Qs and cardiothoracic ratio on X-ray ($p<0.001$, $R^2=0.61$), and Qp/Qs and echocardiographic left ventricular end-diastolic dimension (p=0.73).
Conclusions: Measurement of B-line density on lung ultrasound is useful in evaluating Qp/Qs in children with VSD.

Keywords: Ventricular septal defect, Lung ultrasound, pulmonary congestion

PP-488

Myocardial strain in the healthy neonate

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Background and Aim: Conventional echocardiographic measurements of cardiac function in the neonate have several limitations. 2D speckle tracking echocardiography (2DSTE) derived strain can contribute to the quantification of systolic ventricular function as well as atrial function. Reference values for the pediatric population is available, however data for neonates <30 days of age are scarce. The aim of this study was to describe myocardial strain in healthy neonates during the first day of life and at 3-4 weeks of age and evaluate the feasibility of 2DSTE in this population.
Method: 15 healthy, full-term neonates were prospectively examined twice at first day and at three weeks of age according to a designed echocardiographic protocol. 2DSTE measurements were feasible from all examinations.
Results: Right ventricular free wall strain, left ventricular global longitudinal strain, right atrial and left atrial reservoir and conduit strain was increased at the second examination, p<0.004.
Conclusions: Strain analysis is feasible in healthy neonates and contributes to quantification of atrial and ventricular function with high reliability provided that high frame rate is ensured during acquisition. The increased myocardial strain at 3-4 weeks of age indicates systolic and diastolic improvement during circulatory adaption after birth. Further studies are required to clarify the clinical and prognostic value of myocardial strain in neonates.

Keywords: Myocardial strain, Echocardiography, Speckle-tracking, Neonate

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PP-489

Impact of maternal thyroid disease on cardiac abnormalities in the newborn

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Background and Aim: Maternal thyroid disease has been linked to severe congenital heart diseases (CHDs) in the newborn. Whether it is also associated with less severe structural and functional cardiac abnormalities remains to be further investigated.
The aim of this study was to investigate the association between exposure to maternal hypo- and hyperthyroidism during pregnancy and cardiac structural and functional characteristics in the newborn.
Method: This study is part of the Copenhagen Baby Heart Study (CBHS), a large-scale cohort study including over 25,000 newborns recruited between April 2016 and October 2018. The newborns underwent transthoracic echocardiogram (TTE) and an electrocardiogram (ECG) within the first 60 days of life. Furthermore, comprehensive information about the parents was collected. This study will include all newborns in the CBHS born to mothers with thyroid disease and a matched control group, i.e. non-exposed newborns in the CBHS. Controls will be matched on sex, gestational age at birth, age and weight at time of TTE and ECG. The mothers will be identified using ICD10 codes from the Danish health registries and their medical charts will be manually reviewed to validate the thyroid disease diagnosis. In addition, information on medication, disease onset, characteristics, and severity will be collected through manual review of medical charts. The association between maternal thyroid disease and cardiac structural, functional, and conduction characteristics in the newborn will be investigated.
Results: Extraction from health registries identified 2,017 individual mothers with an ICD10 code for thyroid disease. Validation of the diagnosis is currently ongoing, with final results expected in Spring 2024.
Conclusions: Identification of risk factors for CHDs is important to implement preventive measures and provide relevant medical management and surveillance, to reduce CHD-associated morbidity and mortality. This large-scale study presents a unique opportunity to assess the potential impact of maternal thyroid disease on heart development, and whether newborns in this population should undergo routine systematic cardiac evaluation after birth.

Keywords: Cardiac imaging, Congenital heart disease, cardiac abnormalities

PP-490

A case of neonatal arterial thrombosis mimicking aortic coarctation

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Background and Aim: Thrombotic disease of the newborn is relatively rare compared to that of an adult, mostly representing venous thrombosis. The incidence of arterial thrombosis in infants and children is reported only in a few studies and these are often iatrogenic thrombosis related to vascular access. Spontaneous arterial thrombosis in the neonate is extremely rare but can be life threatening, necessitating emergency systemic thrombolysis and/or surgical thrombectomy.
Method: We describe the case of a newborn baby that presented soon after birth with mild respiratory distress and was found to have severe LV systolic dysfunction due to obstruction of the isthmus by a large mass/thrombus mimicking aortic coarctation. An additional thrombus of 10 mm was revealed in the inferior vena cava. CT angiography confirmed the echocardiographic findings (picture 1) and IV infusion of heparine was started. As the echocardiographic findings remained unchanged and there was no clinical improvement, it was decided to start systemic thrombolysis with r-tPA
Results: After one cycle of tPA infusion there was complete resolution of the thrombus and improvement of the ventricular function. Thrombophilia screen didn't reveal any abnormalities and therefore, in the absence of relevant family history, the etiology of the haemodynamically important thrombosis remains unknown.
Conclusions: Thrombotic disease of the newborn can be a life-threatening complication and in the presence of one important thrombus, all arterial and venous pathways should be screened for additional sites.

Keywords: thrombus, newborn, coarctation, thrombolysis, thrombectomy

PP-491

Correlation study: estimating right ventricle ejection fraction using magnetic resonance imaging/transthoracic three-dimensional echocardiography

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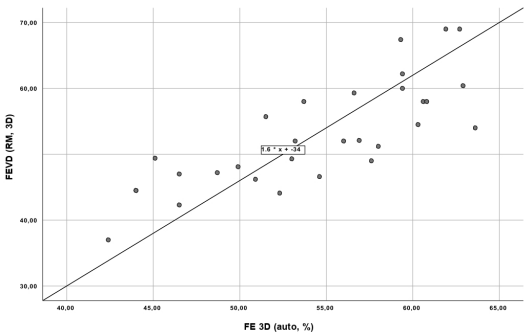
Background and Aim: In recent years, the assessment of right ventricle (RV) function has gained significance as a prognostic marker in Children with Congenital Heart Disease (CHD). Although, Magnetic Resonance Imaging (MRI) is the gold standard for assessing volumes and systolic ejection fraction (EF), its use in children requires sedation and is limited in many hospitals. The advancement of transthoracic three-dimensional echocardiography (TT3DE) presents a promising alternative for RV assessment. The aim is to perform a correlation analysis of diagnostic testing for estimating RV volumes and EF using MRI and TT3DE, along with assessing RV function parameters in 2D/3D in children with CHD.
Method: Data from children with CHD at our center (September 2022-October 2023) were analyzed, excluding those without an optimal acoustic window 1,5 tesla MRI was conducted under sedation, and TT3DE used EPIC CVX ultrasound machines (PHILIPS), both within a 6-month interval.
Cardiac volumes adjusted for body surface área- end-diastolic volume (EDV), end-systolic volume (ESV), and RVEF were evaluated using the Portal ISCV RM program (Philips) and auto-RV (artificial intelligence) TT3DE program. Additionally, RV function data, including tricuspid annular systolic excursion (TAPSE) and 3D RV strain, were analyzed. The correlation between automatic and two-dimensional (2D) measurements, as well as RV EF and volumes by TT3DE and MRI, was assessed using the Pearson correlation coefficient.
Results: 30 children with CHD undergoing MRI and 3D/2D TTE were included (main age 11 years, 34,5% Tetralogy of Fallot, 13,8% pulmonary stenosis). The correlation between 2D TAPSE and automatic 3D TAPSE was very good at 0.80 ($p<0.001$), and the RVEF estimation by TT3DE compared to MRI was also good at 0.793 ($p<0.001$). However, the volumetric correlation of RV and strain (2D-3D) was weaker, with correlations of 0.57 for EDV, 0.63 for ESV, and 0.6 ($p<0,001$) for RV strain.
Conclusions: TT3DE provides a non-invasive, automatic, and accurate method for estimating RVEF and RV longitudinal parameters in children with CHD, offering a quick, simple, and harmless approach. Implementing this technique in CHD units could reduce the need for repeated MRIs in children, facilitating RV function monitoring and providing prognostic data with a technique that has a straight forward learning curve.

Keywords: Right Ventricle, magnetic resonance imaging, transthoracic three-dimensional echocardiography, ejection fraction, volumes, children.

GRAPH



Correlation representation graph between right ventricle (RV) Cardiac Magnetic Resonance Ejection Fraction and that estimated by Transthoracic 3D Echocardiography.

PP-492

Multimodal analysis of hypertrophic cardiomyopathy restrictive phenotype: An uncommon presentation

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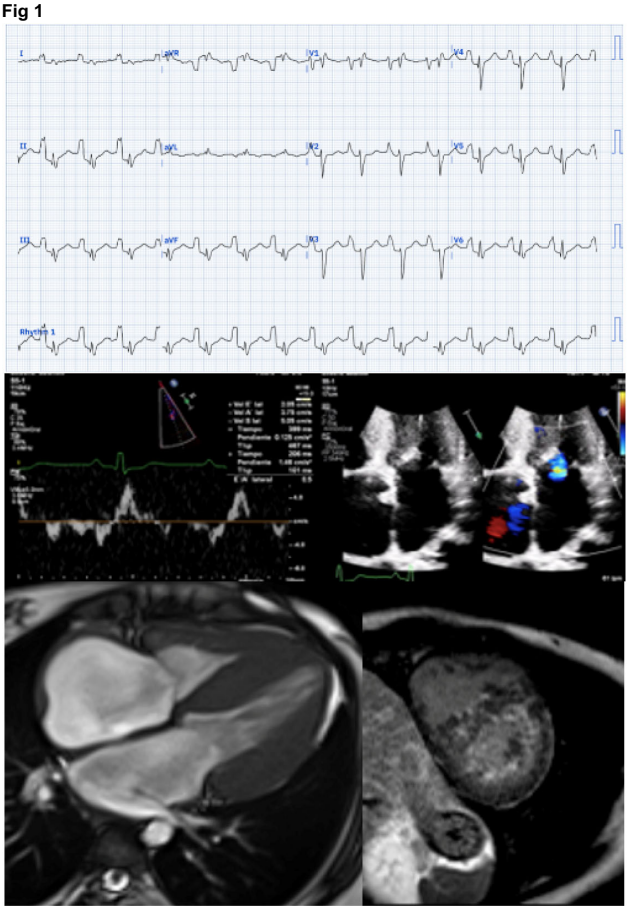
Background and Aim: Hypertrophic cardiomyopathy is the most prevalent genetic cardiac condition, typically occurring in approximately 0.3-0.5 cases per 100,000 children. Within this group, there is a small subset of patients who present severe diastolic abnormalities and restrictive cardiac physiology. Recently a rare subtype of hypertrophic cardiomyopathy, known as the restrictive phenotype, has been identified, constituting only 1.3% of cases. However, the unique characteristics of this phenotype have yet to be thoroughly explored within our population.
Method: Clinical case report describing an unusual presentation of hypertrophic cardiomyopathy through a multimodal analysis.
Results: A 17-year-old female patient first experienced symptoms of heart failure at the age of 12, including orthopnea, precordial pain, and dyspnea. A diagnostic assessment was initiated, leading to the diagnosis of non-obstructive biventricular concentric hypertrophic cardiomyopathy with significant dilation of both atria. The electrocardiogram revealed a prominent 1.1 mV P-wave, marked biauricular enlargement with a distinct "Himalayan" P-wave

Fig 1



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pattern, and a low-voltage QRS complex. Further evaluation included an echocardiogram, which identified diastolic dysfunction of the right ventricle with an E/e' ratio of 10.57 (ZS +6.87) and diastolic dysfunction of the left ventricle with an E/e' ratio of 15.7 (ZS +8.46). Additionally, there was a global longitudinal myocardial strain of -3.5%. Cardiovascular magnetic resonance revealed late gadolinium enhancement in areas of hypertrophy, accounting for 44% of the total left ventricular mass, indicating diffuse intramyocardial fibrosis and severe dilation of both atria. Treatment for heart failure was initiated, and the patient was considered for cardiac transplantation.
Conclusions: It is well known that patients with hypertrophic cardiomyopathy commonly present with diastolic dysfunction as their typical pathophysiology. However, patients with a restrictive phenotype, such as our case, exhibit significantly larger atrial dimensions, distinguishing them from those with classical hypertrophic cardiomyopathy. To ensure accurate diagnosis and appropriate management, further exploration of this rare phenotype is required. This case highlights the importance of recognizing that cardiomyopathies can manifest in various mixed patterns. Therefore, it is essential to consider these findings when approaching their diagnosis and treatment.

Keywords: Hypertrophic, Cardiomyopathy, Multimodal analysis

PP-493
A rare congenital heart disease with high diagnostic complexity: ARCAPA

Luis Felipe Castillo Aguilar[1], Vincenzo Arenas Fabbri[2], Leonardo Rivera Rodriguez[1], Jorge Luis Cervantes Salazar[3], Juan Eberto Calderon Colmenero[1], Regina De La Mora Cervantes[2], Diana Maribel Paz Huox[1], Yessica Alejandra Toro Pastrana[1], Arturo Iván Alonso Vázquez[1], Patricio Anduaga Del Ángel[1], Andrea Carolina Gasca Sánchez[1] and Claudia González García[1]
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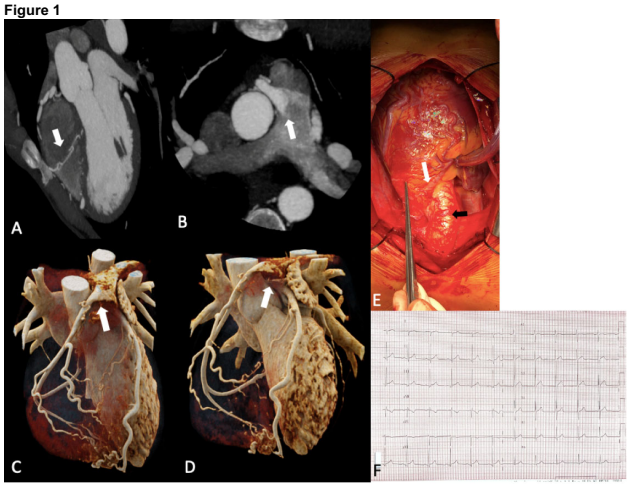
Background and Aim: The anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA) is a rare congenital heart disease, with an incidence of 0.002% and represents 0.12% of coronary anomalies. The presentation of ARCAPA is frequently not specific and angiotomography is the gold standard diagnostic imaging study.
Method: A clinical case report describing a rare congenital heart disease: ARCAPA.
Results: We present a case of a 15-year-old male patient with no significant medical history, who began his current condition with chest pain and syncope during physical activity. He was admitted to hospital for diagnostic and therapeutic approach. Physical examination was no remarkable. Electrocardiogram (ECG) and chest X-ray with no significant findings. Transthoracic echocardiogram showed a dilated right coronary artery of 4.2 mm (Z Score +3.03) in its medial portion. The origin and course of the artery could not be identified. The angiotomography was performed and showed an anomalous origin of the right coronary artery from the lateral aspect of the pulmonary artery trunk and extensive collateral arteries. The patient remained clinically asymptomatic and reimplantation of the right coronary ostium to the ascending aorta and reconstruction of the pulmonary artery with patch was performed, without complications or adverse events. The patient was discharged in good condition.
Conclusions: The diagnosis of ARCAPA in most cases is complex and it requires a high suspicion supported by the appropriate diagnostic tools. The related symptoms are chest pain, dyspnea, fatigue and murmurs, however it is rare that it manifests as syncope, it is necessary to consider this clinical variant as a symptom associated with this rare condition. Taking this into account, it is important not only to perform echocardiogram as a diagnostic tool in the scenario of suspecting coronary anomalies since sometimes may not be enough with the need to perform an angiotomography to establish an accurate diagnostic, as in this case. The definitive treatment is surgery and the prognosis is excellent once corrected.

Keywords: ARCAPA, coronary anomalies, congenital heart surgery

Figure 1



A. Multiplanar reconstruction (MPR) image showing collateral artery crossing right ventricle (RV) cavity on its way to RV free wall (white arrow). B. MPR image showing right coronary artery (RCA) emerging from the pulmonary artery trunk (white arrow). C. Volume rendering reconstruction (VRR) image showing ARCAPA and extensive collateral arteries (white arrow). D. VRR image showing left coronary artery (LCA) with normal origin (white arrow). E. Surgical procedure image showing RCA (white arrow) emerging from the pulmonary artery trunk (black arrow). F. ECG showing not significant findings.

PP-494
Demonstration of streaming with cardiac MRI to unravel a case of desaturation despite high pulmonary blood flow in transposition of the great arteries

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Background and Aim: In transposition of the great arteries, oxygen saturation is determined by effective pulmonary blood flow rather than total pulmonary blood flow. Desaturation secondary to ineffective mixing due to preferential streaming can be difficult to prove in infants and we present a case report of using cardiac MRI with 4D flow data to aid the diagnosis and management.
Method: A 2-month-old, 3.2-kilogram girl with transposition of the great arteries, hypoplastic bipartite right ventricle, unrestrictive ventricular septal defect, mild pulmonary valve dysplasia, and

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coarctation of the aorta had previously undergone surgical atrial septectomy, aortic arch repair, and placement of bilateral pulmonary artery bands at two weeks of age. She had persistently labile oxygen saturations with frequent desaturation despite non-invasive ventilation support and clinical optimisation. Echocardiogram showed unrestrictive, bidirectional atrial and ventricular shunt, good biventricular systolic function, accessory mitral valve tissue in the sub-pulmonary left ventricular outflow tract without obstruction, normal pulmonary valve flow velocity, and well-positioned pulmonary artery bands with left and right pulmonary artery maximum flow velocities 4.3m/s and 3.8m/s respectively. Her hypoplastic right ventricle precluded arterial switch and VSD closure at this stage and her age and weight precluded proceeding with a bidirectional Glenn anastomosis.
Results: A cardiac MRI with 4D flow data was performed to better delineate the underlying physiology prior to potential intervention. This demonstrated high total pulmonary blood flow with a ratio of pulmonary to systemic blood flow of 3:1. Twist angiography was suggestive of preferential streaming of the systemic venous return to the aorta. The indexed right ventricular end diastolic volume was 39ml/m2 with normal biventricular systolic function. She proceeded to have an arterial switch operation, branch pulmonary artery debanding, and main pulmonary artery band placement. She made a good recovery and was able to be discharged home after three weeks.
Conclusions: Cardiac MRI with 4D flow data can be useful to demonstrate streaming in patients with unexplained desaturations and guide further surgical management in complex congenital heart disease.

Keywords: Streaming, 4D flow, cardiac MRI, TGA

PP-495
Neonatal cardiac, sedation free, contrast free feed-and-wrap mri can add useful clinical information for decision making

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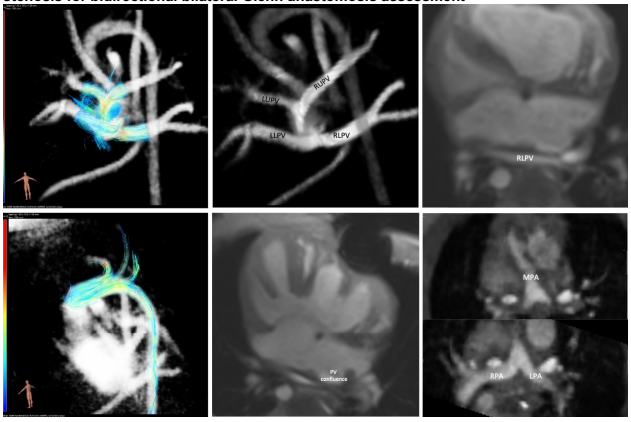
Background and Aim: Traditionally, neonatal cardiovascular magnetic imaging (MRI) requires a general anaesthetic for breath-holding. However, using an MRI safe incubator, free-breathing cine imaging and 4D Flow MRI now allows contrast free and sedation free neonatal feed-and-wrap scans for comprehensive haemodynamic and anatomical assessment.
This study examines the clinical usefulness of neonatal feed-and-wrap MRI scans.
Method: 60 neonates (age range 1 day to 5 months) underwent non-contrast sedation-free feed-and-wrap MRI scan on a 3T clinical scanner (MAGNETOM Prisma, Siemens Healthcare, Erlangen, Germany) using a dedicated 18 channel variety coil (NORAS MRI products GmbH, Hoechberg, Germany) and an MRI-compatible incubator (LMT Medical Systems GmbH, Luebeck, Germany) facilitating seamless transfer into the scanner. The imaging protocol included compressed sensing accelerated 4DFlowMRI research sequence and additional multiple average free-breathing cine imaging and 2D flow assessment if the neonate stayed asleep. Data was analysed using standard commercially available software.
Results: Only one 5-month-old baby failed to stay asleep during the MRI scan and 59 babies successfully completed the feed-and-wrap MRI scan. Clinical questions for CMR assessment included: Arch anatomy (24), pre-Glenn/biventricular repair surgery assessment (21), borderline ventricle (5), pulmonary vs systemic blood flow assessment [Qp:Qs] (5) and others (4). 17/ 58 (29%) MRIs confirmed the findings on echocardiogram (mainly arch anatomy). In 41/58 (71%) cases the MRI added extra information compared to echocardiogram alone. This additional information could have also been obtained with contrast CT in 21/58 (36%) cases but additional information was available unable to be obtained by CT. Additional information gained included: Collateral assessment (13), Qp:Qs assessment (8), patent ductus arteriosus net flow direction (5), arch anatomy (4), branch pulmonary artery flow differential (4), stroke volume of left ventricle (2), ventricular size (2), size of ventricular septal defect suitable for biventricular repair.
Conclusions: Non-contrast sedation-free feed-and-wrap MRI is feasible in the clinical setting and often adds additional information compared to echocardiography alone. Especially in the neonatal population it is a viable alternative to contrast-based CT scan and can often add additional haemodynamic information not available on anatomical CT alone.

Keywords: 4D Flow MRI, neonatal, feed and wrap

3 month old baby with repaired total anomalous venous drainage, right atrial isomerism, bilateral SVCs, dextrocardia, unbalanced AVSD, transposed great arteries, pulmonary stenosis for bidirectional bilateral Glenn anastomosis assessment



PP-496
Intra pericardial congenital tetratoma in a newborn a case report

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Background and Aim: Congenital teratoma is one of the most common fetal tumors. It is most often benign. Pericardial location is unusual and extremely rare. It is usually presents with a variety of cardiac and systemic symptoms in the neonatal period or early childhood. Through a neonatal case report we aim to highlight the difficulties in the management of this rare and challenging condition.

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Method: It's a case report of an intrapericardial congenital teratoma in a male newborn.
Results: We report the case of a male newborn. His fetal ultrasound performed at 33 weeks showed a heterogeneous mediastinal mass associated with a massive pericardial effusion. He was born at 34 weeks by normal vaginal delivery. Physical exam at birth revealed an immediate respiratory distress. Chest radiography showed a Cardiomegaly and a widened mediastinal shadow. Echocardiography showed an anterior intrapericardial tumor measuring 35 mm associated with a massive pericardial effusion. Chest CT-scan showed a multi-walled intrapericardial mass. Emergency pericardial drainage was performed. It drained a citrine yellow liquid. It was removed 18 hours after due to the appearance of pneumopericardium. The pericardial effusion recurred, causing a heart failure and hemodynamic disorders leading to death at 70 hours of life. Post-mortem histopathological exam concluded to an immature teratoma
Conclusions: Pericardial teratoma is a serious and challenging condition. It can be lifethreatening due to the pericardial effusion and heart failure induced by the tumor compression. Prenatal diagnosis and early treatment may improve the prognosis.

Keywords: teratoma, foetal, tamponnade

PP-497
Anomalous left and right single pulmonary vein: A rare benign congenital variant
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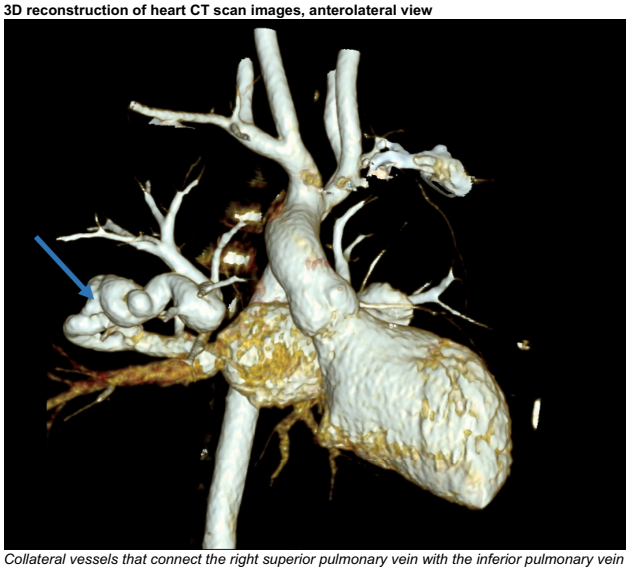
Background and Aim: The development of the pulmonary venous system is a complex process involving multiple embryonic venous systems to establish the normal venous configuration of the lungs. This intricate development can result in various anomalies within the pulmonary venous system, posing diagnostic challenges.
Method: Case description of a rare image finding, incorporating patient history, clinical presentation as well as complementary investigation, which included diagnostic imaging.
Results: We present the case of a 4-year-old female patient born as part of a twin pregnancy at 34 weeks gestational age, complicated by intrauterine growth restriction. At 6 months of age, she began experiencing recurrent episodes of wheezing. At the age of 1 year, she was diagnosed with right upper lobe pneumonia and successfully treated with oral amoxicillin. However, at age 2.5 years, she experienced another episode of pneumonia in the same location, requiring similar treatment. One month later, despite remaining asymptomatic, chest radiography revealed a persistent right paracardial image, and was referred to the pediatric respiratory clinic. On observation, she had adequate peripheral oxygen saturation, heart rate, and blood pressure. Laboratory findings and transthoracic echocardiogram were unremarkable. Simultaneously, the chest X-ray continued to display a right paracardial opacity at the middle lobe level. Two potential hypotheses were considered: bronchiectasis and middle lobe syndrome. Subsequently a Angio-CT scan revealed an anomalous drainage of the right and left superior pulmonary veins into the ipsilateral inferior pulmonary veins through a network of collaterals (Figures 1). Additionally, there was an accessory right lobar bronchus emerging from the right lateral tracheal wall, located 8mm above the common bronchus, which then bifurcated into the middle and right inferior lobes.
Conclusions: To the best of our knowledge, this is the first documented case of an anomalous drainage of the left and right single pulmonary veins, as well as an accessory right lobar bronchus. While typically benign, this condition may also present with concurrent bronchiectasis.

Keywords: pulmonary venous system, anomaly, tomography, single pulmonary vein

3D reconstruction of heart CT scan images, anterolateral view



Collateral vessels that connect the right superior pulmonary vein with the inferior pulmonary vein (blue arrow).

PP-498
Fetal right ventricular pseudoaneurysm: A very rare case in a patient affected by truncus arteriosus
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Background and Aim: We discuss the case of a large right ventricular pseudoaneurysm (RVP) in a fetus with Truncus Arteriosus (TA) type A1. The first reported case diagnosed prenatally was published in 1990, however, no reported cases of fetal ventricular pseudoaneurym in complex congenital heart disease (CHD) exist. Cardiac pseudoaneurysms are defined as any rupture in the wall of the myocardium that is contained by pericardium, thrombus, or adhesions. Pseudoaneurysms present a narrow neck and their wall completely lacks myocardium, having high risk of rupture.
Method: We report the case of a 30 years old pregnant woman.
Results: The first fetal echocardiogram performed at our center at 20 weeks + 6 days of gestational age (GA) showed a TA type A1 according to Van Praagh classification and large circumferential pericardial effusion. At GA of 29 weeks we detected the presence of a large pseudoaneurysm of the right ventricle with a neck originating from the anterior-inferior wall of the right ventricle. It was supplied by the right ventricle with a high-velocity jet. The anterior wall of right ventricle was also dyskinetic. An accessory inferior mid-ventricular septal defect was also found. The fetus showed also a reduction of pericardial effusion during the follow up.

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Conclusions: A RVP associated with TA has never been reported in literature. The diagnosis of pseudoaneurysm is important due to his possible different natural history. Echocardiographic features established for pseudoaneurysms include a neck diameter at end systole that is less than half the maximum diameter of the outpouching. Additionally, aliasing and bidirectional flow in and out of the outpouching's cavity can be visualized using color flow and spectral Doppler. Due to their rarity, the management of pseudoaneurysms is based on limited experience. While the primary goal is to prevent further rupture, the specific approaches vary greatly since the risk of rupture is associated with multiple factors. However, RVP appear to be less likely to further rupture. The multidisciplinary team has a key role to plan birth and minimise risk of pseudoaneurysmal rupture and a catastrophic bleed at birth.

Keywords: Fetal Pseudoaneurysm, Truncus Arteriosus, Right Ventricle

Figure 1&2

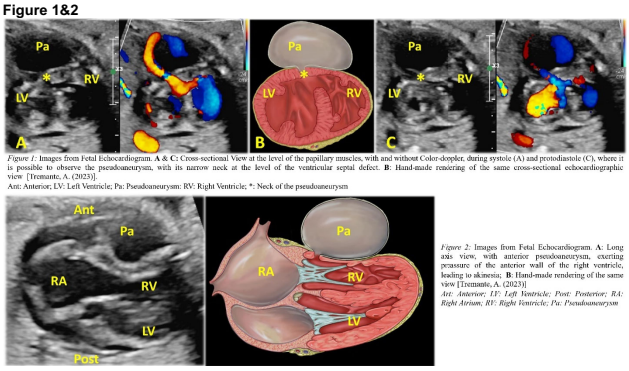

Figure 1: Images from Fetal Echocardiogram. **A** & **C:** Cross-sectional View at the level of the papillary muscles, with and without Color-doppler, during systole (A) and protodiastole (C), where it is possible to observe the pseudoaneurysm, with its narrow neck at the level of the ventricular septal defect. **B**: Hand-made rendering of the same cross-sectional echocardiographic view [Tremante, A. (2023)].
Ant: Anterior; LV: Left Ventricle; Pa: Pseudoaneurysm: RV: Right Ventricle; *: Neck of the pseudoaneurysm

Figure 2: Images from Fetal Echocardiogram. **A**: Long axis view, with anterior pseudoaneurysm, exerting pressure of the anterior wall of the right ventricle, leading to akinesia; **B**: Hand-made rendering of the same view [Tremante, A. (2023)]
Ant: Anterior; LV: Left Ventricle; Post: Posterior; RA: Right Atrium; RV: Right Ventricle; Pa: Pseudoaneurysm

PP-499

A rare case of transposition of great arteries with double aortic ARCH: A highly complex fetal diagnosis with an unpredictable outcome

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Background and Aim: D-transposition of the great arteries (D-TGA) associated with anomalies of the aortic arch occurs in approximately 10-15%. Double aortic arch (DAA) is the most common type of vascular ring. DAA and D-TGA association was described rarely in literature.
Method: The diagnosis was made through a fetal echocardiogram performed by the Perinatal Cardiology Unit at the outpatient clinic of Bambino Gesù Hospital.
Results: Fetal echocardiography at a gestational age of 24 weeks + 6 days showed D-TGA and DAA with ventricular septal defect (VSD) and pulmonary stenosis. A few hours after birth patient underwent a Rashkind procedure. Echocardiography, CT chest, CT angiogram confirmed diagnosis with severe reduction of the tracheal lumen (>85%) and bronchomalacia. The patient underwent posterior tracheopexy and aortopexy and later an Arterial Switch Operation, VSD closure, and resection of part of the infundibular septum, accepting the risk of potential neoaortic obstruction. The patient is currently extubated, with a successful outcome of the surgery, and is continuing their recovery while still admitted to the cardiology department.

Conclusions: D-TGA with DAA remains an extremely rare condition. The literature reported only 8 cases of DAA and D-TGA, just 2 cases with fetal echocardiogram diagnosis and just 1 was diagnosed at a very early GA. Our patient is the third one who had a fetal diagnosis of D-TGA with DAA, but the second one who had a follow-up in itinere, and he is the first ever with a description of D-TGA and DAA with complex intracardiac anatomy, characterized not only by VSD but also by the presence of dual obstruction components at the pulmonary outflow tract, valvar and subvalvar.
The fetal diagnosis still represents a major challenge for fetal cardiologists, although outcome of these patients presents a high degree of variability and is completely unpredictable in prenatal life. Factors influencing this spectrum are related to intracardiac anatomy, the arrangement of the great vessels, tracheal compression and tracheomalacia associated, which represent negative prognostic factors.

Keywords: Transposition Great Arteries, Double Aortic Arch, Fetal Diagnosis

TGA & DAA

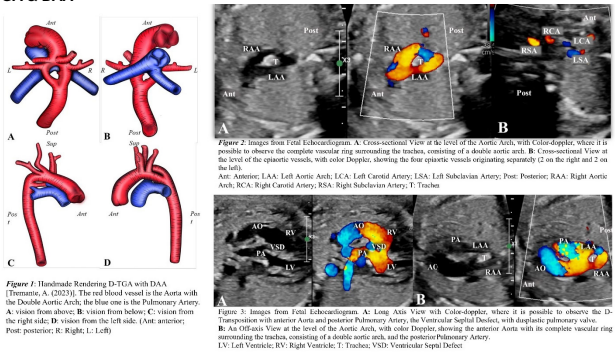

Figure 1: Handmade Rendering D-TGA with DAA [Tremante, A. (2023)]. The red blood vessel is the Aorta with the Double Aortic Arch; the blue one is the Pulmonary Artery. **A**: vision from above; **B**: vision from below; **C**: vision from the right side; **D**: vision from the left side. (Ant: anterior; Post: posterior; R: Right; L: Left)

Figure 2: Images from Fetal Echocardiogram. **A**: Cross-sectional View at the level of the Aortic Arch, with Color-doppler, where it is possible to observe the complete vascular ring surrounding the trachea, consisting of a double aortic arch. **B**: Cross-sectional View at the level of the epiaortic vessels, with color Doppler, showing the four epiaortic vessels originating separately (2 on the right and 2 on the left).
Ant: Anterior; LAA: Left Aortic Arch; LCA: Left Carotid Artery; LSA: Left Subclavian Artery; Post: Posterior; RAA: Right Aortic Arch; RCA: Right Carotid Artery; RSA: Right Subclavian Artery; T: Trachea

Figure 3: Images from Fetal Echocardiogram. **A**: Long Axis View with Color-doppler, where it is possible to observe the D-Transposition with anterior Aorta and posterior Pulmonary Artery, the Ventricular Septal Defect, with duplastic pulmonary valve. **B**: An Off-axis View at the level of the Aortic Arch, with color Doppler, showing the anterior Aorta with its complete vascular ring surrounding the trachea, consisting of a double aortic arch, and the posterior Pulmonary Artery.
LV: Left Ventricle; RV: Right Ventricle; T: Trachea; VSD: Ventricular Septal Defect

PP-500

Case report: Chronic complete occlusion of the left main coronary artery in a pediatric patient with exertion-related symptoms

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Background and Aim: Acute left main coronary artery (LMCA) occlusion can result in anterior myocardial infarction and severe complications such as cardiogenic shock, ventricular tachycardia or sudden cardiac death. In contrast, chronic LMCA occlusion is rare, as patients can only survive if extensive collaterals develop from the right coronary artery (RCA), and these patients are often asymptomatic or may exhibit angina symptoms.
Method: We present the case of a healthy 7-year-old girl admitted to the emergency department after experiencing chest pain and syncope following physical activity. The initial electrocardiogram revealed ST segment depression in the left precordial leads, which resolved simultaneously with the chest pain. Subsequent serial electrocardiograms were normal. Analytical studies revealed transient troponin elevation (maximum 1696.8ng/L) and normal B-type natriuretic peptide levels. A transthoracic echocardiogram demonstrated preserved biventricular function, mild RCA ectasia (3.6 mm, z-score +3.2), and difficult assessment of the LMCA origin and proximal course. A treadmill exercise test was conducted,

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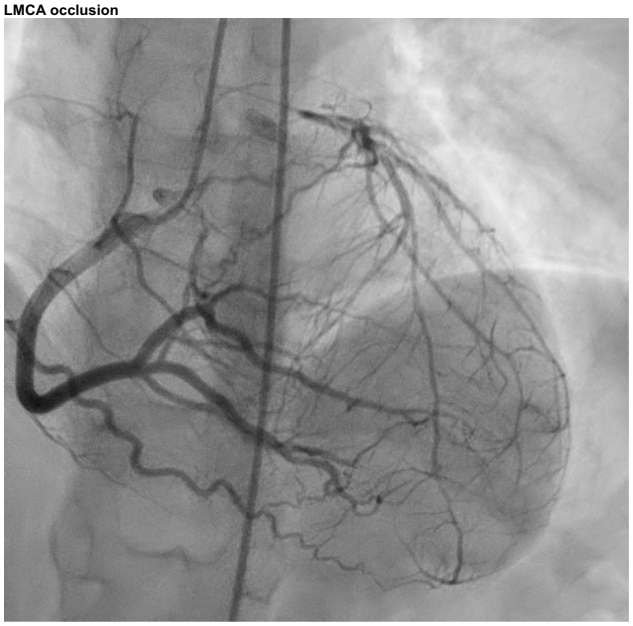
which was maximal, asymptomatic, and negative for ischemia, with troponin levels remaining negative after the test.
For further anatomical characterization, a cardiac CT angiography showed right coronary dominance, and the LMCA was poorly visualized, seemingly originating from the left coronary sinus, and without distal filling defects.
Results: Given lingering uncertainties after these diagnostic studies, additional imaging was deemed necessary. Cardiac magnetic resonance imaging revealed normal biventricular size and function, with no areas of late gadolinium enhancement. Subsequent selective coronary angiographies showed dominant RCA and complete occlusion of the LMCA with extensive collateral filling from the right coronary artery system, without angiographic evidence for coronary atherosclerosis. The patient had no known vascular disease risk factors or a history of Kawasaki disease.
Conclusions: Complete occlusion of the LMCA is an uncommon event during cardiac catheterization due to its high risk of cardiovascular events, including sudden cardiac death, even in the presence of extensive collateral filling from the right coronary artery system. Although coronary revascularization is usually recommended, careful consideration must be taken in a case-by-case basis, particularly in young and small patients.

Keywords: Chronic left main coronary artery occlusion, Syncope, Exertion-related symptoms, Electrocardiogram, Coronary angiography, Pediatric

LMCA occlusion



PP-501

Cardiovascular magnetic resonance T1 and T2 mapping in heart transplant patients: Establishing normative values for rejection-free assessment

Marko Taipale[1], Markku Pentikäinen[2], Laura Martelius[3], Aino Mutka[4], Soili Kytölä[5], Matti Kankainen[6], Juha Peltonen[7], Arttu Lahtiharju[8], Jyri Lommi[2], Timo Jahnukainen[9], Sari Kivistö[3], Karl Lemström[8] and Tiina Ojala[1]
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Background and Aim: The assessment of myocardial tissue in heart transplant patients for possible acute rejection is crucial. T1 and T2 mapping – markers for inflammation, oedema, and fibrosis - by cardiac magnetic resonance (CMR), in conjunction with donor-derived cell-free DNA (dd-cfDNA) analysis, holds promise as a non-invasive approach. For understanding rejection patterns across the heart muscle in CMR, we need comprehensive data of global and segmental T1 and T2 values in transplanted hearts without rejection. However, this data as well as the relationship between CMR mapping and dd-cfDNA remains largely unexplored. Our aim was to establish normative CMR T1 and T2 values in heart transplant patients during the first two years after heart transplantation.
Method: In this blinded prospective study (Trial NCT04311346), we analyzed CMR data from 200 scans in 41 heart transplant recipients within 1 to 24 months post-transplant between March 2020 and November 2023 in Finland. Sixteen myocardial segments were categorized based on their anatomical location and coronary territory. Exclusion criteria to study analysis included EMB acute cellular rejection (ACR) grade ≥ 1 or antibody-mediated rejection (AMR) grade ≥ 1, dd-cfDNA ≥ 0.15 %, left ventricle ejection fraction (LVEF) < 40 %, or history of significant rejection (ACR grade ≥ 2 or AMR grade ≥ 1). *Results:* Our study demonstrated that: 1) over the 24 months post-transplant, global T1 time remained stable, while T2 time biphasically decreased (Figure); 2) segments of right coronary artery territory exhibited higher T1

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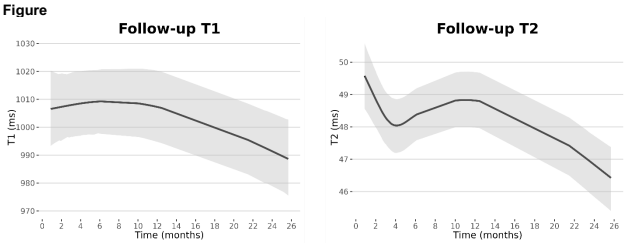
values (1021.4 ms [1006.2-1036.7] vs. 998.3 ms [983.1-1013.5], p < 0.001) and lower T2 values (49.8 ms [48.6-51.0] vs. 50.7 ms [49.5-51.9]; p < 0.001) than segments of left anterior descending artery territory; and 3) increase in dd-cfDNA, decrease in LVEF, and ischemic heart disease as the indication for heart transplantation were associated with elevated T1 values.
Conclusions: In this national 3-year cohort, we demonstrate reference data for CMR T1 and T2 values in heart transplant patients without rejection. The combination of CMR T1 and T2 mapping with dd-cfDNA as a noninvasive tool for rejection detection warrants prospective studies, underscoring the significance of this normal dataset as a valuable reference.

Keywords: CMR, mapping, heart transplantation, rejection, cell-free DNA

Figure



PP-502
3D transthoracic echocardiography assessment in pediatric cardiology: A key in surgical planning of mitral valve

Marta Gambra Arzoz[1], Lucía Deiros Bronte[1], Ángela Uceda Galiano[1], Viviana Arreo Del Val[1], Ángel Aroca Peinado[2], Álvaro González Rocafort[2], Enrique Balbacid Domingo[1], Carlos Labrandero De Lera[1], Diana Sala Mera[1], César Abelleira Pardo[1], Antonio Javier Cartón Sánchez[1], Álvaro Lafuente Romero[1], Mercedes Sampedro Martín[3], Luis García Guereta Silva[1] and Federico Gutiérrez Larraya Aguado[1]
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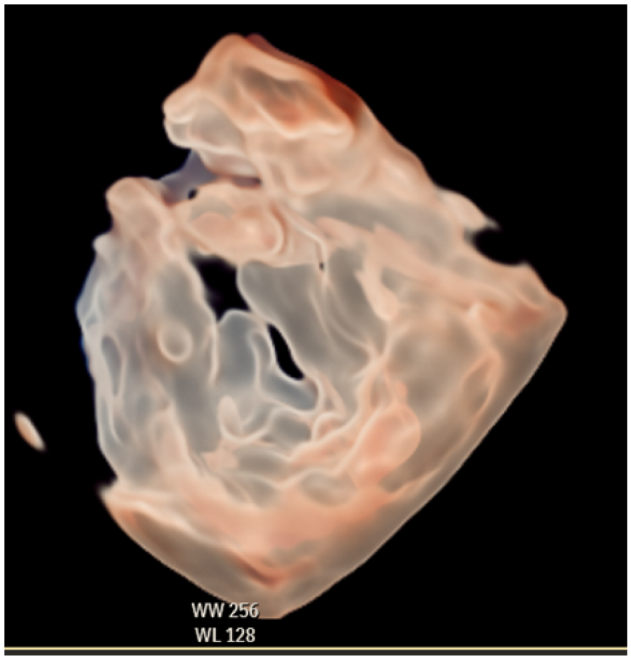
Background and Aim: The mitral valve is a complex structure consisting of the annulus, leaflets, chordate and left ventricular papillary muscles. The description of the morphology of mitral valve disease and its mechanisms is essential for the surgical planning. As mitral valve is a three-dimensional structure, the addition of 3D echocardiography assessment is advantageous. The improvement of 3DE technology and the more feasibility of 3D transthoracic echocardiography in children due to a better acoustic window, makes 3D TTE assessment a key in surgical planning of the mitral valve. We aim to describe the 3D TTE in surgical planning in the last 3 patients operated in a tertiary hospital.
Method: Philips Epic CVX ultrasound system was used. The patient was in left lateral decubitus position when possible, the appropriate matrix probe was selected, 3D volume acquisition was made by activating 3D zoom and transillumination (a new tool that uses a freely movable virtual light source to enhance depth, contours, and image detail), 3D volume was optimized, 3D full volume acquisition was stored and post processed by QLAB.
Results: first patient was a five year old girl, with left atrium enlargement and sever mitral valve regurgitation due to mitral valve cleft. In this patient, transillumination made the diagnosis of isolated mitral leaflet cleft which has not been diagnosed by 2D assessment. Mitral cleft was closured. Patient 2 was a 9 year old girl, 3D TTE assessment showed a mitral valve anterior leaflet prolapsed (A2) and a short posterior leaflet chordate restricting motion. Mitral valve was repaired with artificial chordae replacement. Patient 3 was a neonate with sever mitral regurgitation, the mechanism of mitral disease was similar to patient 2 but in this case mitral reparation failed and a Melody valve was placed in mitral position.
Conclusions: the improvement of the 3DE technology with advances in probe technology and in the postprocessing software optimization (allowing surgeons and cardiologist sharing the same anatomical vision of mitral valve) and the more feasibility of 3D TTE in children, have lead 3D TTE to be a great tool in surgical planning of mitral valve.

Keywords: mitral valve surgery, 3D echocardiography, thanstoracic echocardiography

mitral cleft



transillumination making the diagnosis of isolated mitral leaflet cleft

PP-503
Global longitudinal strain and myocardial work in paediatric patients with type 1 diabetes mellitus

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Background and Aim: Cardiac dysfunction and endothelial damage are complications of type 1 diabetes mellitus (T1DM) that mainly affect adults. However, many studies have shown the appearance of subclinical myocardial changes of the left ventricle (LV) already in paediatric age in patients with T1DM. Recently, the introduction of new techniques to support conventional echocardiography has made it possible to characterize ventricular function in a finer manner, enriching the knowledge of the pathophysiology of cardiomyopathies of various origins.
The aims of our study were to investigate global longitudinal strain and myocardial work in paediatric patients with type 1 diabetes diagnosed from at least 10 years and to compare their myocardial performance parameters to the ones of age and sex-matched healthy volunteers.
Method: We prospectively enrolled 31 patients in between 13–19-year-old with a diagnosis of type 1 diabetes from at least 10 years and in follow-up at our Paediatric Diabetology Unit. For all patients, clinical parameters of glycaemic controls (time in range, glycated haemoglobin, glycaemic variability) and echocardiographic measurements (conventional echocardiography, tissue doppler parameters, global longitudinal strain and Myocardial Work) were recorded. Echocardiographic findings were then compared with data from 31 age and sex-matched healthy volunteers.
Results: In our population parameters of glycaemic control were relatively homogeneous and showed suboptimal glycaemic control (mean glycated haemoglobin 7,4 ± 1,1%; average time in range in between 50-70%; average glycaemic variability >36%). As shown in Table 1, Global longitudinal strain (GLS), Global Work Index (GWI) and Global Work Efficiency (GWE) were significantly lower in the T1DM population compared to the healthy population, whilst Global Wasted Work was significantly higher in the T1DM population (Fig 1).
Conclusions: Both Global Longitudinal Strain and the descriptive parameters of Myocardial Work are significantly lower in the sample examined compared to healthy individuals, highlighting a subclinical cardiac damage already present during adolescence. GLS and myocardial work could therefore become useful tools in assessing cardiac function in diabetic patients, while preventive strategies must be applied to maximize glycaemic control effectiveness

Keywords: myocardial work, global longitudinal strain, type 1 diabetes

PP-504
Correlation between echocardiography and ct angiography in abnormal pulmonary venous return (APVR): A rétrospective study

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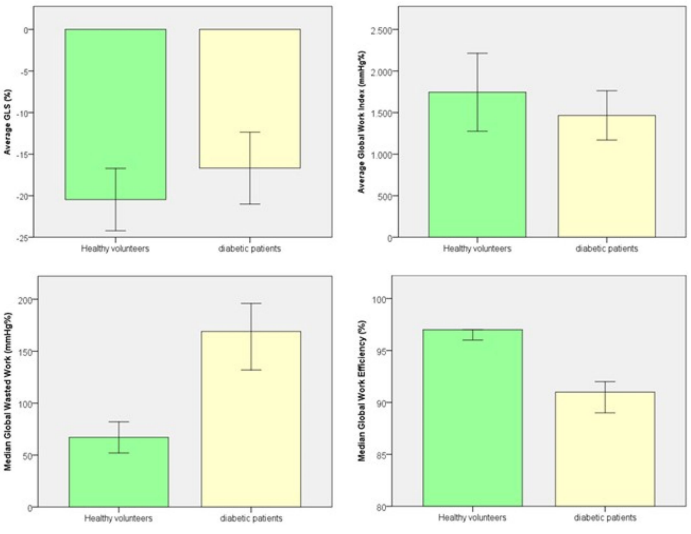
Background and Aim: Abnormal pulmonary venous return (APVR) may result in either partial or complete anomalous drainage back into the systemic venous circulation. A wide spectrum of normal pulmonary venous drainage exists in Partial anomalous pulmonary venous drainage (PAPVD). t is more common on the right especially drainage of the RSPV into the superior vena cava (SVC). Three anatomic types exist, depending on the level of drainage: supracardiac (the most common form), intra cardiac and infracardiac. Given the variability of pulmonary venous anatomy, detailed pre-procedural evaluation of the left atrium and the pulmonary veins is desirable prior to treatment The imaging of pulmonary venous anatomy has traditionally been performed with echocardiography and catheter pulmonary angiography. Multi-detector

Table I and Figure 1

	Diabetic patients	Healthy controls	p value
GLS (%)	-16.7 ± 2.2	-20.5± 1.9	p< 0.001
GWI (mmHg%)	1466 ± 148	1744 ± 234	p< 0.001
GCW (mmHg%)	1941 (491)	2120 (280)	0.097
GWW (mmHg%)	169 (86)	67 (39)	p< 0.001
GWE (%)	91 (4)	96.8 (6)	p< 0.001

Table 1. Comparison in GLS and Myocardial Work parameters among diabetic patients and healthy volunteers. Normally distributed variables are reported as average value and standard deviation, while non-normally distributed variables are expressed as median value and interquartile range.

Fig 1. Comparison between global longitudinal strain and myocardial work parameters in diabetic patients and healthy volunteers is shown.



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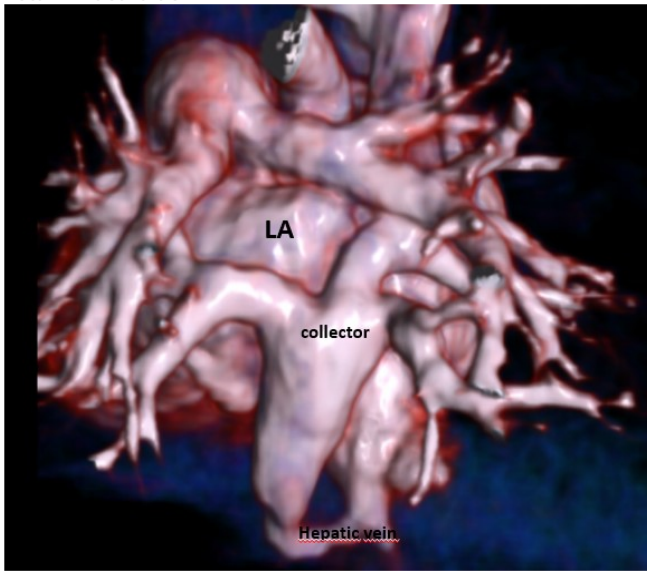
computed tomography (CT) have refined the usual imaging techniques. We aim to assess the role of CT angiography in evaluation of abnormalities in pulmonary venous drainage and compare it with echocardiography.
Method: It is a single center retrospective study, including patients followed in cardiology and radiology departments between 2011 and 2023 for APVR. Echocardiography was performed on all patients. CT angiography provides an alternative diagnostic modality for morphologic evaluation. A correlation between echographic and angiographic data was performed. We enrolled 20 patients with APVR (from 2011 to 2023).
Results: The study included 10 cases of total APVR (6 supracardiac, 2 intracardiac, and 2 infracardiac) and 10 cases of partial APVR (8 supracardiac, 1 intracardiac, and 2 infracardiac). In all cases, CTA confirmed echocardiographic findings (type of APVR and drainage location),but the CT scan allowed to locate more precisely the site of the pulmonary vein drainage.
Conclusions: APVR are rare but potentially fatal pathologies requiring surgical treatment in some cases. Echocardiography is the preferred examination, and CTA is a very efficient method for confirming diagnosis, typing, searching for associated malformations, and guiding the therapeutic strategy.

Keywords: drainage, venous return, echocardiography, CT angiography

Total infracardiac APVR



PP-505
Medium term outcomes of unbalanced atrioventricular septal defect (AVSD) in a single centre

Melonie Johns, Phuoc Duong and Caroline B Jones
Alderhey Children's Hospital, Liverpool, UK

Background and Aim: Unbalanced AVSD remains challenging, with poor long-term outcomes. There has been a shift towards using staged interventions, in order to, "recruit" the smaller ventricle before performing delayed biventricular (BiV) repair. This relies on the premise that the anatomical imbalance of ventricular volumes can be altered by addressing the physiological imbalance atrial, ventricular and vascular shunts can create.
Method: Single centre retrospective analysis of all patients attending a tertiary Cardiology Centre where there has been debate between BiV or UV (univentricular) repair in patients with a modified AVVI <0.4 at presentation from 2018-2023. Patients with TAPVD and VA discordance were excluded.
Results: 11 patients underwent assessment by cross-sectional imaging. The majority had echocardiography and CMR imaging. Mean follow up was 27 months (range 10-59). 3 had left-dominant, 8 right-dominant ventricle. 3 patients had heterotaxy and 4 had genetic abnormalities. 4 patients were deemed unsuitable for BiV strategy due to a modified AVVI <0.35, indexed volume ratio <0.3 or an unfavourable valve anatomy. 4 had a 'recruitment' procedure with the aim to rehabilitate the smaller ventricle prior to BiV repair; of these 1 was converted to univentricular circulation as the ventricular size had not increased sufficiently. There were no early post-operative deaths. No patients following recruitment strategy died. One univentricular patient died at 23 months awaiting transplant for ventricular failure. 2 patients in the BiV repair cohort died with significant comorbidities. Development of moderate-severe left AVVR was significantly associated with mortality (p=0.02). Heterotaxy and genetic diagnosis were not significantly associated with mortality (p=-0.15 and 0.5 respectively). All patients with a PA band applied remained clinically well with no evidence of raised pulmonary pressures by diagnostic catheter. Patients with a fenestrated ASD closure had a greater increase in the modified AVVI than counterparts who did not have their ASD restricted (P=0.06).
Conclusions: Biventricular recruitment is a viable strategy with low medium-term mortality and can delay a decision between BiV and UV strategies without risk of pulmonary hypertension during the interval between surgeries. Patients who developed severe left AVVR had higher mortality. Predicting which patients will develop regurgitation is yet unclear on current imaging parameters.

Keywords: unbalanced atriventricular septal defect, biventricular recruitment

PP-506
A case of anomalous origin of the left coronary artery from the right coronary sinus with a benign course

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Background and Aim: Anomalous coronary arteries originating from the opposite sinus of Valsalva (ACAOS) are a challenge because of their various anatomic and clinical presentation. There exist different clinical presentations and anatomic variants of ACAOS. Most of these variants are considered benign. The ACAOS variant with an inter-arterial course, that is, with the anomalous vessel running between the great arteries, is the foremost potential hemodynamical relevant anomaly.
Method: We are reporting a case of a 2-year-old female. Who was discovered to have anomalous left coronary artery from the right sinus of Valsalva during a routine echocardiogram. She is known to have congenital adrenal hyperplasia under medical therapy. Otherwise there were no cardiac symptoms. There were no murmur or ECG changes. Her echocardiogram showed anomalous left coronary artery origin from the right coronary sinus. The right and left coronary arteries are arising from a common origin. A CT scan

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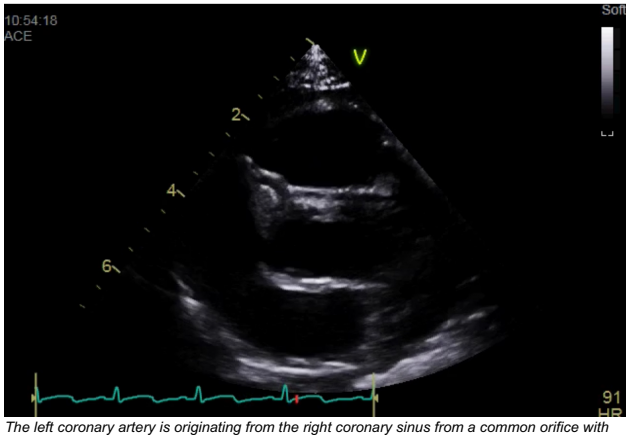
was done which showed Anomalous origin of the long left main coronary artery from the right coronary sinus (common origin of right coronary and left main coronary arteries) with trans-septal course of the left coronary artery, passing between the right ventricular outflow tract and aortic root.
Results: Most of anomalous left coronary from the opposite sinus variants are considered benign and coincidental findings with no hemodynamic relevance and thus not related to cardiac symptoms (ie, prepulmonic, subpulmonic, retroaortic, intraseptal).
In our case the CT angiography diagnosed the anomaly as intraseptal (also called subpulmonic course) and ruled out inter-arterial course, the anomaly was considered benign with no need for surgery.
Conclusions: Anomalous coronary arteries from the opposite sinus are a challenge because of their various anatomic and clinical presentation. Requires high index of suspicion to be diagnosed by echo. Many of the cases has a benign course. A CT scan is required to rule out the inter- arterial (malignant course).

Keywords: Anomalous origin, coronaries, opposite sinus, case report.

Short axis echocardiogram view



The left coronary artery is originating from the right coronary sinus from a common orifice with the right coronary artery.

PP-507
Giant right atrial thrombosis in pediatric restrictive cardiomyopathy: A rare complication

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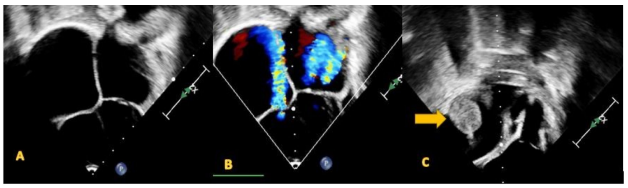
Background and Aim: Thrombotic complications in pediatric RCM (restrictive cardiomyopathy) cases are rare, complicating their management due to the lack of standardized protocols. The scarcity of guidelines emphasizes the need for tailored strategies and further research to address these critical complications effectively.
Method: A 1.5-year-old female presented with post-feeding tachypnea and a familial history of cardiomyopathy. Transthoracic echocardiography diagnosed RCM revealing atrial dilation, normal ventricular dimensions, and insufficiency in both atrioventricular valves. Genetic evaluation unveiled a homozygous TNNI3 mutation, guiding medical management. Subsequent follow-up detected a sizable, previously unnoticed thrombus (26x24 mm) in the right atrium, ventricular dysfunction and valve regurgitation. Anticoagulation was initiated.
Results: Pediatric RCM, characterized by diastolic dysfunction, often implicates genetic mutations, with thrombotic complications being rare.
Conclusions: The complexity of thrombus management in RCM due to its infrequent occurrence challenges the establishment of standardized protocols. Thrombus formation in cardiomyopathies poses risks of thromboembolic events, emphasizing the necessity for tailored and evidence-based guidelines to address these rare yet critical complications. Further research and comprehensive data collection are imperative to guide effective therapeutic interventions in such uncommon scenarios.

Keywords: Pediatric Restrictive Cardiomyopathy, Transthoracic Echocardiography, Atrial Dilation, Atrioventricular Valve Insufficiency, Thrombus

Figure 1



A. Apical 4 chamber view, B. Mitral and Tricuspid valve regurgitation, C. Thrombus in the right atrium

PP-508
Cardiac structure and function in newborns and preschool children with decreased fractional shortening at birth

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Background and Aim: Congenital heart diseases (CHD) constitute the most common type of birth defect and are seen in 0.8% of live births. Despite improvements in prenatal detection of CHDs in the past decades, a considerable proportion of minor defects remains undetected until later in childhood or even adulthood. Furthermore, the implications of abnormal echocardiographic measurements observed in infancy for cardiac function in childhood or later in life are unclear. This study investigated the prevalence of reduced fractional shortening (FS) in newborns, identified potential maternal risk factors for reduced FS, and investigated whether a reduced FS shortly after birth normalizes, persists, or worsens by preschool age.
Method: The study was based on data from the Copenhagen Baby Heart Study, a population-based cohort study that included transthoracic echocardiography (TTE) examination within 60 days of birth. The TTE protocol included measurements of left ventricular (LV) internal diameter in end-diastole (LVIDd) and end-systole (LVIDs) obtained from echocardiographic images from the parasternal long axis view. Reduced systolic function was defined as having FS (calculated as LVIDd − LVIDs/LVIDd × 100%)

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<28%. Potential risk factors for reduced FS at birth included maternal height, weight, pre-pregnancy BMI, parity, smoking and comorbidities, and the child's gestational age at birth, birthweight and length, Apgar-score, and birth-method; data on these factors were collected from medical records. A sample of newborns with FS<28% at birth was reinvited for follow-up examinations at 12 and 36 months.
Results: Results are expected in December 2023.
Conclusions: Identifying risk factors for reduced FS in infancy can provide valuable insights into which children require closer monitoring during the early years of life.
Follow-up examinations of children with subclinical abnormal findings at birth will provide novel information on the evolution and clinical significance of such findings, indicating whether reduced FS persists beyond the first years of life and if so, in whom, helping us to characterize normal variability in infant FS, refining abnormal infant values for these parameters, and suggesting which FS levels require clinical action and which can be managed with routine monitoring.

Keywords: echocardiography, cardiac abnormalities

PP-509

The predictive value of longitudinal strain in assessing the risk of malignant arrhythmias in children diagnosed with myocarditis

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Background and Aim: Myocarditis poses a potentially life-threatening condition in pediatric patients. Malignant arrhythmias, alongside severe heart failure, are a dreaded manifestation and may even occur in patients with preserved systolic cardiac function. Conventional echocardiography faces constraints as an efficient diagnostic tool. Therefore, we aimed to assess the diagnostic and predicitive value of speckle tracking echocardiography-derived longitudinal strain (LS) in children with diagnosed myocarditis.
Method: Pediatric patients with myocarditis, confirmed either through endomyocardial biopsy (EMB) or cardiac magnetic resonance imaging (cMRI) were enrolled in the multicenter, prospective myocarditis registry "MYKKE". Echocardiographic parameters, including M-Mode-derived wall thicknesses, left ventricular ejection fraction (LVEF), LV myocardial mass (LVM) and 4-chamber view-derived endo-/myo-/epicardial LS, were assessed upon initial admission using TomTec Imaging Software. Malignant arrhythmias included non-sustained and sustained ventricular tachycardia (VT), ventricular fibrillation (VF) and third-degree atrioventricular block (AVB).
Results: Echocardiograms from 135 patients included by twelve participating centers were analyzed (2014-2023). Median age was 15.3 (IQR 10.4-16.5) years, with 76 % male. Malignant arrhythmias were monitored in 32 patients (24%; VT n=30, VF n=1, AVB III n=1). Among them, 53% (n=17) had preserved LVEF ≥ 50%, 6% (n=2) had mildly reduced LVEF (41-49%), and 34% (n=13) had reduced LVEF ≤ 40%. Patients with and without malignant arrhythmias showed no differences in echocardiographic parameters LVEF (53% vs. 53%, p=0.367) and LV end-diastolic diameter (LVEDD) Z-score (+0.8 vs. +0.8, p=0.722). However, those with malignant arrhythmias had significantly lower median LS values compared to those without (endocardial LS -14% vs. -21%, p=0.004; myocardial LS -10% vs. -16%, p=0.002; epicardial LS -8% vs. -13%, p=0.001). This was also true when only considering patients with preserved LVEF. As expected, patients experiencing major cardiac adverse events (MACE) including heart transplantation (n=10, 7%), mechanical circulatory support (n=18, 13%), resuscitation (n=18, 13%), and/or death (n=6, 4%) exhibited significantly decreased median LVEF and LS parameters (p<0.001), along with an increased LVEDD Z-score (p<0.001), compared to those without MACE.
Conclusions: In pediatric patients, echocardiography-derived LS is a sensitive diagnostic tool. It not only assesses systolic myocardial function and predicts MACE but also identifies patients at risk for malignant arrhythmias, even in the context of preserved LVEF.

Keywords: Myocarditis, Echocardiography, Longitudinal Strain, Malignant Arrhythmias

PP-510

Optimal timing of surgical intervention for aortic regurgitation based on cardiac mri assessment in congenital patients

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Background and Aim: Indexed MRI right ventricular (RV) volume is the gold standard for defining the correct timing to replace a regurgitant pulmonary valve. On the contrary, there are no cut-off points related to left ventricular (LV) volume indicating the optimum timing for intervention on a regurgitant aortic valve. This study aims to establish MRI indices related to aortic regurgitation that are linked to optimal timing for surgical intervention.
Method: A retrospective review was conducted using data from the Pediatric Cardiology & Adult Congenital Heart Disease Department of MITERA Hospital, between 2013-2023. Forty-seven patients with aortic regurgitation who had undergone cardiac MRI were identified and divided into two groups: those who were listed for surgical interventions based on echocardiographic and clinical criteria and those who did not require intervention, serving as the control group. Relevant MRI indices, including LV end-diastolic index (LVEDi), LV end-systolic index (LVESi), LV ejection fraction (EF), aortic valve regurgitant fraction (RF), and the ratio between LV and RV (LV/RV) were recorded.
Results: Among the 47 patients analyzed (age 16, range 1-64yrs), 26 were scheduled for surgical repair, whilst 21 required no intervention. Of the patients that underwent surgery, echocardiographic characterization of the AR was severe and moderate to severe in 53.8% and 46,2%, respectively. Median LVEDi and LVESi for patients undergoing surgery were 132ml/m2 (range 115-149ml/m2) and 76ml/m2 (range 60-85ml/m2), respectively vs 85ml/m2 (range 76-96ml/m2) and 28ml/m2 (range 26-38.5 ml/m2) for the non-surgical group. Median RF was 44% (range 36-51%) vs RF 26% (range 15-35%) for the control group. Median LV/RV ratio was 1.5 (range 1.3-1.8) vs 1.1 for the control group. The majority of patients (80.7%) had preserved LV EF, except for 5 outliers with LV EF $\leq$46%. All results were found to be statistically significant ($p < 0.05$) compared to the control group.
Conclusions: In this study, MRI indices were observed from studies performed at the time that surgery was decided based on echocardiographic and clinical criteria. Further studies are warranted to validate these indices as reliable predictors and indicators for determining the optimal timing for surgical intervention for aortic regurgitation.

Keywords: Aortic Regurgitation, Cardiac MRI, Pediatric Cardiology, Aortic valve, Left Ventricular Volume

PP-514

Utilization of 3d-virtual models for operative planning of pulmonary atresia, ventricular septal defect and multiple aortopulmonary collaterals

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Background and Aim: The goal of this single-centre study was to evaluate the benefits of 3D virtual versicolor heart models use in clinical management of patients with pulmonary atresia, ventricular septal defect and multiple aortopulmonary collaterals.
Method: Between October 2020 and September 2023, 10 three-dimensional virtual heart models based on computed tomography data were reconstructed. The contribution of 3D virtual heart models for operative planning improvement was not only evaluated in each individual patient, but was also evaluated on the basis of general questionnaire filled independently by 5 pediatric cardiac surgeons. The questionnaire comprised questions relating mainly to the quality of anatomical assessment of multiple aortopulmonary collaterals and native pulmonary arteries by 3D virtual models.
Results: 10 virtual models focusing on imaging of pulmonary blood supply anatomy and its spatial anatomical relationship to the bronchial tree were created. Each model offered an improved anatomical orientation of cardiovascular structures. Preoperative analysis of these models helped to choose the most suitable cardiac surgery in each individual case. Upon the questionnaire results, pediatric cardiac surgeons find the use of 3D virtual versicolor heart models as an excellent imaging tool for anatomical deliniation of origins, courses and communications of multiple aortopulmonary collaterals, native pulmonary arteries, bronchial tree and their spatial anatomical relationships. It is for this reason that the 3D models should be used as a complementary imaging method to catheterization angiography in operative planning in all patients with pulmonary atresia, ventricular septal defect and multiple aortopulmonary collaterals in the future.
Conclusions: The use of 3D versicolor virtual models enabling high-quality spatial imaging of pulmonary blood supply can improve preoperative planning in patients with pulmonary atresia, ventricular septal defect and multiple aortopulmonary collaterals.

Keywords: 3D, models, aortopulmonary, collaterals, pulmonary, atresia

PP-515

Single bolus, dual acquisition, low dose protocol for cardiac computed tomography angiography in children with fontan circulation

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Background and Aim: We present a single bolus, dual acquisition low dose protocol for scanning paediatric patients with Fontan circulation. This offers distinct advantages, including no specific site of injection of contrast bolus, low dose acquisitions, uniform opacification of the entire Fontan circuit with no contrast interface. There is no requirement for sedation or general anaesthesia
Method: We use a dual source scanner (Siemens SOMATOM Force), thereby minimising the radiation dose and acquisition time. 2 mls/kg of contrast variably diluted to 60 to 70% is injected through any available IV access at a rate of 0.9 to 2.5 ml/s depending on the volume of contrast available, to obtain a total bolus duration of 60 to 70 seconds. The contrast is bolus tracked every 5 seconds from the end of injection with the scan acquired immediately after the uniform opacification of the lateral tunnel/extracardiac conduit, which is the region of interest for the monitoring slices. A recent addition to the protocol is a second acquisition approximately 150 to 180 seconds from end of contrast bolus. A low Kv acquisition ensures higher contrast opacification with the later acquisition ensuring absence of interface between the opacified and unopacified hepatic blood in the lateral tunnel/extracardiac conduit.
Results: A total 25 scans, all diagnostic, were performed from January 2019 to July 2022 using the single acquisition protocol with mean contrast dose of 2 ml/kg and radiation dose of 30 mGycm. 4 patients were scanned using the dual acquisition protocol from August 2022 to October 2023. All scans were diagnostic with mean radiation dose of 30 mGycm and contrast dose of 2 ml/kg. We have a cohort of 8 patients scanned using the Bastion

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wheel protocol, which is also used in our centre by another colleague. All scans were diagnostic but with a mean radiation dose of 68 mGycm (one patient scanned using prospective gating) and contrast dose of 2 ml/kg. We plan to prospectively compare the 2 protocols for qualitative and quantitative scan measures
Conclusions: The proposed single injection, dual acquisition, low dose protocol for CCTA in Fontan patients offers a unique alternative for scanning children with Fontan circulation.

Keywords: Fontan, Single ventricle, CT Angiography

PP-517

Can right atrial strain improve the diagnostic accuracy of echocardiography in paediatric arrhythmogenic cardiomyopathy?

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Background and Aim: Progressive fibrofatty myocardial replacement in arrhythmogenic cardiomyopathy (ACM) leads to remodelling of the ventricles and potentially the atria. Current echocardiographic revised Task Force Criteria are not met by the majority of adolescent ACM patients, particularly when indexed to body surface area. Our aim was to study the usefulness of right atrial strain as a marker of right atrial contractile dysfunction in paediatric ACM.
Method: We retrospectively reviewed patients at our institution between 2002 and 2019 with definite, borderline, or possible ACM diagnosed using the 2010 revised Task Force Criteria. While blinded to diagnosis, transthoracic echocardiogram at the time of diagnosis was used for right atrial strain analysis and compared with matched healthy controls (n=28).
Results: We identified 73 ACM patients between 0-18 years of age (23 definite, 24 borderline, 26 possible). The median age at diagnosis was 14 years (IQR 11 – 16 years) and 64% were male. Those with definite ACM had significantly lower reservoir strain (39 ± 8.2%, p=0.011) compared with borderline/possible ACM (45 ± 9.1%) and controls (45 ± 7.2%). Conduit strain was also significantly reduced in definite ACM (26% ± 8.7%, p=0.013) compared with borderline/possible ACM (32% ± 7.7%) and controls (30% ± 6.7%). These parameters were not significantly different between borderline/possible ACM and controls. There was no significant difference between pump strain or maximum strain rate between groups. Two patients diagnosed with left ventricle dominant ACM demonstrated a trend towards higher reservoir strain when compared to right ventricle dominant ACM (48 ± 5.5% vs 38 ± 8.0%, p=0.099).
Conclusions: Paediatric patients with definite ACM demonstrated lower right atrial reservoir and pump strain. Right atrial strain may potentially improve the diagnostic yield of echocardiography for ACM. Further studies are warranted to determine if the severity of atrial dysfunction is correlated to cardiovascular events.

Keywords: arrhythmogenic, cardiomyopathy, strain

PP-518

RV volumetry – from simple 2d echocardiographic measurements to a complex form

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Background and Aim: Assessment of right ventricular (RV) volume during the follow-up of patients with congenital heart defects (CHD) is crucial. Due to its complex geometry, especially in CHD patients, transthoracic echocardiographic RV volumetry might remain challenging and MRI is widely used to assess RV volume. However, MRI does not provide bedside live-imaging, is time consuming, expensive and in infants and small children often not possible without sedation. Therefore, we aimed to establish a reliable, easy and fast to obtain method to calculate RV volume using two-dimensional transthoracic echocardiography (2D echo) and validate it with MRI data.
Method: A retrospective single-center trial was conducted at Deutsches Herzzentrum der Charité. Standard apical four-chamber and parasternal short-axis view was obtained in CHD patients during routine follow-up using 2D echo. RV volumes were calculated using $(\alpha+1)/(\alpha+2)*SS'/d$ formula and compared to RV volumes obtained by MRI.
Results: 22 CHD patients (12 female) with various types of heart defects and a median age of 9.1 years were included. As published before, RV volume quantification by 2D echo traditionally relies on the 2/3*S'*a formula, elucidated by an ellipsoidal shell model. However, in our cohort of CHD patients acquired results indicated a need to augment this formula, particularly in diastolic scenarios, with an additional constant factor for reliable data. This constant factor, unaccounted for in the model, necessitates an alternative approach. Therefore, we propose a model based on a rotational body of the curve $x\alpha$. Accordingly, we calculated a constant correction factor α for systolic volume ($\alpha S = 3.1$) and for diastolic volume ($\alpha D = 3.9$), respectively. The associated formula $(\alpha+1)/(\alpha+2)*SS'/d$ (Fig.1) demonstrates superior performance in our experiments with a mean difference to MRI systolic volume

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of 4.84 ml (13.3%; R2=0.88) and a mean difference to MRI diastolic volume of 16.55 ml (19.2%; R2=0.88) (Fig 1).
Conclusions: The proposed method to determine RV volume in CHD patients using two-dimensional echocardiography allows distinct modeling in systolic and diastolic phases with excellent correlation to RV volume obtained by MRI. Thus, this promising approach provides a feasible, exact and fast method to assess RV volume on bedside, facilitating prompt and precise clinical decision-making

Keywords: RV-Volumetry, 2D-TTE, Non-invasive, Bedside

Required measurements to calculate RV Volume in 2D echo and its correlation to MRI

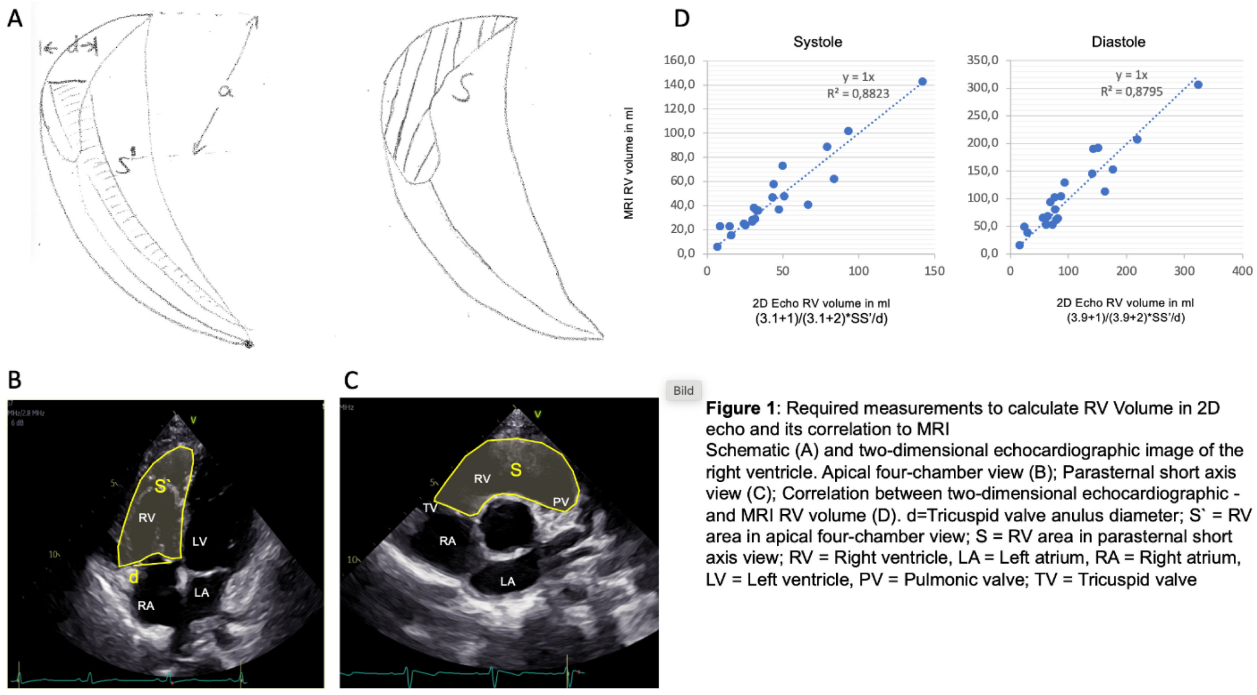

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Figure 1: Required measurements to calculate RV Volume in 2D echo and its correlation to MRI
Schematic (A) and two-dimensional echocardiographic image of the right ventricle. Apical four-chamber view (B); Parasternal short axis view (C); Correlation between two-dimensional echocardiographic - and MRI RV volume (D). d=Tricuspid valve anulus diameter; S` = RV area in apical four-chamber view; S = RV area in parasternal short axis view; RV = Right ventricle, LA = Left atrium, RA = Right atrium, LV = Left ventricle, PV = Pulmonic valve; TV = Tricuspid valve

Schematic (A) and two-dimensional echocardiographic image of the right ventricle. Apical four-chamber view (B); Parasternal short axis view (C); Correlation between two-dimensional echocardiographic and MRI RV volume (D). d=Tricuspid valve anulus diameter; S` = RV area in apical four-chamber view; S = RV area in parasternal short axis view; RV = Right ventricle, LA = Left atrium, RA = Right atrium, LV = Left ventricle, PV = Pulmonic valve; TV = Tricuspid valve

PP-519
Retrospective external validation study comparing prediction models in the neonates with prenatal suspicion of coarctation of the aorta (COA)

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Background and Aim: In new-borns, coarctation of the aorta (CoA) diagnosis is still challenging. In 2013, Soslow et al. developed the first neonatal coarctation prediction model (CPM). Most recently, in 2023, Bartolacelli et al. have also developed a CPM. However, these models have not been externally validated.
This study aims to externally validate these models to evaluate their generalisability and clinical usefulness in predicting CoA reliably.
Method: Retrospective review of 150 neonates who were suspected to have CoA prenatally from January 2016 to December of 2021 was conducted. Same arch measurements that were described by the authors were obtained by two observers who were blinded to the outcome. After calculating the risk scores from the models equations, the outcome was referenced. The external validation process was performed according to TRIPOD checklist (Transparent reporting of a multivariable prediction model for individual prognosis or diagnosis). Key measures of calibration were assessed using calibration in the large (CITL), calibration slope, calibration plot and the expected to the observed outcome ratio (E:O). Also, for the discrimination analysis, the area under the curve (AUC) was assessed. The statistical analysis was performed using StataMP 17 (64-bit).
Results: From the 150 neonates, 139 included in this study. Of these, 59 (42.4%) patients developed CoA and underwent surgery. There were no significant differences in birth weight, gestational age, or ethnicity between neonates with and without CoA. In our cohort, Soslow et al. model showed 93.2% sensitivity and 42.5% specificity, while Bartolacelli et al. model sensitivity was 57.6% vs 85% specificity. Discrimination in Soslow et al. model showed an AUC of 0.96 (95% CI 0.88–0.99), verses in this study, the AUC is 0.786 (95% CI 0.710-0.862). Also, Bartolacelli et al. model AUC

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was 0.9382 (no CI given), compared with an AUC of 0.812 (95% CI 0.737-0.887) in this study. Calibration statistics showed that both models were miss-calibrated and over-fitted. Soslow et al. model (E:O) = 1.323, (CITL)= -1.605, calibration slope= 0.291, while Bartolacelli et al. model (E:O)= 0.695, (CITL)= 1.555, calibration slope = 0.321.

Conclusions: This external validation study indicates that both models are miss-calibrated and associated with false negative rates in our cohort.

Keywords: coarctation of the aorta, prediction model

Figure 1

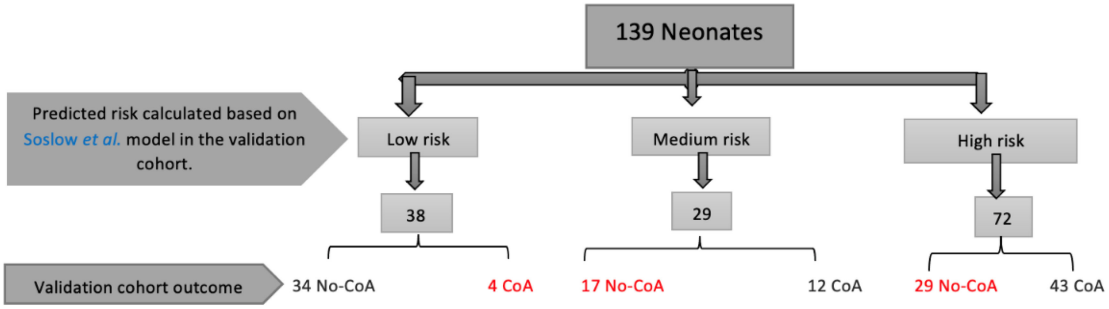

Figure (1): Flow chart of the predicted risk scores according to Soslow *et al.* model, and the validation cohort actual outcome.

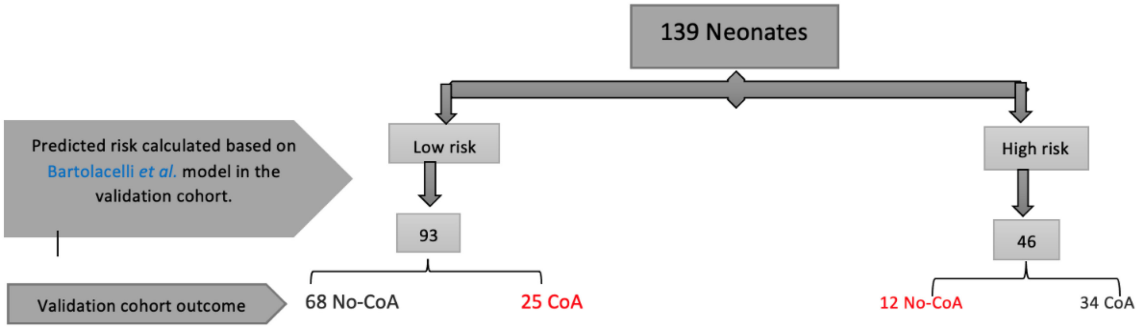

Figure (2): Flow chart of the predicted risk scores according to Bartolacelli *et al.* model, and the validation cohort actual outcome.

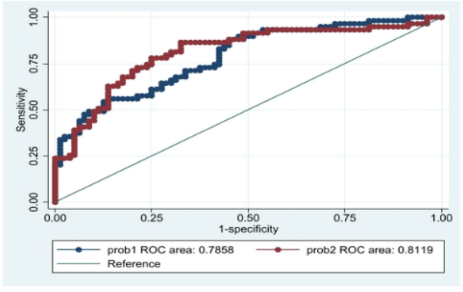

Figure (3): Comparison of the discrimination between both models. (The blue ROC curve: is the model by Soslow *et al.*; while the red ROC curve is the model by Bartolacelli *et al*.

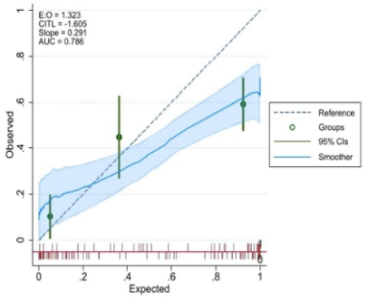

Figure (4): Calibration plot with key measures of calibration assessing Soslow *et al.* model.

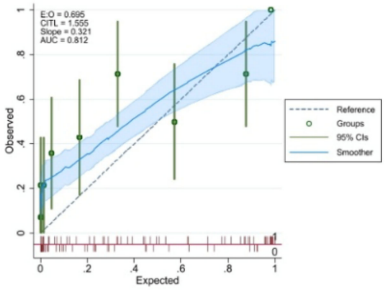

Figure (5): Calibration plot with key measures of calibration assessing Bartolacelli *et al.*[1] model.

CITL: calibration in the large; E:O: expected:observed; AUC: area under the curve.

PP-520

Unveiling subclinical myocardial remodeling in pediatric dystrophinopathies: A study on cardiac magnetic resonance (CMR) parametric mappings

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Background and Aim: Dystrophinopathies, such as Duchenne and Becker muscular dystrophy, often lead to cardiomyopathy, which is the primary cause of mortality in affected patients. Early detection of cardiac dysfunction is crucial, but current imaging methods lack insight into early microstructural remodeling. This study aimed to evaluate the potential of CMR (Cardiac magnetic resonance) parametric mappings for detecting early cardiac involvement and to assess whether any myocardial involvement pattern may serve as an indicator of impending cardiac dysfunction in pediatric dystrophinopathy patients.
Method: A prospective study was conducted on 23 patients with dystrophinopathies who underwent CMR with tissue mappings. To establish a basis for comparison, data from 173 healthy subjects were analyzed. CMR included SSFP sequences for heart function assessment, T2-weighted and T1-weighted sequences before and after gadolinium contrast administration, and tissue mappings for native T1 (nT1), extracellular volume (ECV), and T2 relaxation times.
Results: In this study, significant differences emerged between patients and controls in key cardiac parameters, including LV ejection fraction (LVEF), myocardial mass, and late gadolinium enhancement (LGE), confirming cardiac abnormalities in these patients. Regarding tissue mappings, nT1 ($p<0.001$) and ECV ($p=0.002$), but not T2, displayed substantial variations, indicating their potential as sensitive indicators of myocardial involvement; also a significant asymmetric myocardial involvement between septal and LV posterior wall regions was detected in nT1 (1004.82ms vs 1040.61ms, $p=0.01$) and ECV (24.58% vs 27.18%, $p=0.012$). Notably, higher mapping values did not correlate with reduced ventricular function. However, an asymmetric myocardial involvement, especially in nT1 (correlation coefficient (ρ)=-0.472, $p=0.023$) and ECV (ρ=-0.460, $p=0.049$), exhibited a significant negative correlation with LVEF, highlighting the nuanced relationship between tissue mappings and cardiac function in dystrophinopathies.
Conclusions: CMR parametric mappings demonstrate promise in detecting early myocardial damage in dystrophinopathies. LGE excels at identifying focal fibrosis, but CMR mappings provide valuable insights into diffuse myocardial involvement. Although mapping values may not directly correspond to ventricular function, the observed negative correlation between asymmetric involvement in nT1 and ECV with LVEF suggests their potential as early biomarkers. Larger studies with diverse age groups and longitudinal data are needed to enhance our understanding of CMR changes and improve risk stratification in this population.

Keywords: Dystrophinopathies, Duchenne muscular dystrophy, magnetic Resonance, mapping, dysfunction.

PP-521
The evolution of 3D printing in a large tertiary centre
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Background and Aim: Over the last 10 years 3D printing has become a valuable tool in the pre-surgical planning process for certain types of complex congenital heart disease. Its use in our centre has progressed from isolated test cases with rudimentary models to the standard-of-care in certain lesions with the availability of complex multicoloured models. We sought to review the evolution of the 3D printing service in our department.
Method: We analysed all of our patients who had 3D printed models created since it was first introduced in our department in 2014. We reviewed the indications along with the characteristics of the model including the material(s), the colour(s), the types of cuts through the models and any extra features.
Results: From 2014-2023, 82 models were printed for 78 different patients. 17 were printed by an international manufacturer between 2014 and 2017 with a further 65 printed by a local manufacturer from 2018 onwards. The main indications were in double outlet right ventricle to assess suitability for Rastelli (17), in complex single ventricle prior to Fontan completion (12), congenitally corrected TGA to assess suitability for Hemimustard-Rastelli (6) and in TGA/VSD/PS to assess suitability for Nikaidoh (5). Other indications have involved printing of right ventricular outflow tracts prior to percutaneous pulmonary valve insertion (3) and airways in cases of vascular compression (3). The majority of the models are now translucent and slightly pliable with one or 2 cuts to demonstrate the relevant anatomy and using magnets to connect parts together (49), Older models were either made from softer translucent material with a single cut (24) or rigid plastic with a single cut (13) without the use of magnets. 3 recent models have used more sophisticated techniques that allow different colours to be used for different structures in the heart.
Conclusions: 3D printing is becoming ever more important in the field of congenital heart disease. The service has expanded and evolved considerably in our institution over a 10 year period and given the continuing advancements in the quality of the models, we expect this to continue for the foreseeable future.

Keywords: 3D printing, congenital heart disease

Different 3D Model Types

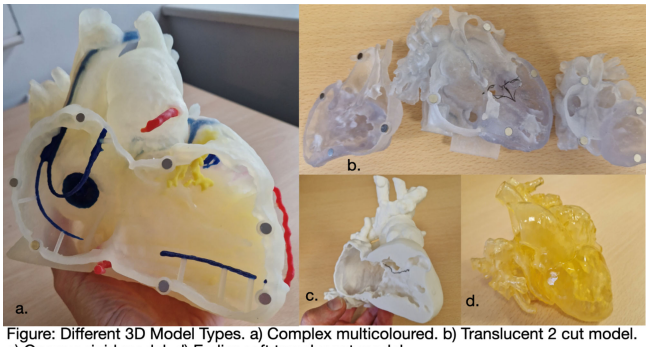

Figure: Different 3D Model Types. a) Complex multicoloured. b) Translucent 2 cut model. c) Opaque rigid model. d) Earlier soft translucent model

PP-522
Cardiac magnetic resonance imaging; assessing myocardial strain and fibrosis as predictors of adverse fontan outcomes
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Background and Aim: In individuals with a univentricular heart, integrating cardiac magnetic resonance imaging (CMR) data with clinical parameters has proven crucial for risk stratification and may offer a valuable tool for improving prognosis. Our objective was to further investigate this by examining specific CMR risk markers—myocardial strain and fibrosis (T1 mapping) —in patients with adverse Fontan outcomes.
Method: This national retrospective observational cohort study included all patients with Fontan circulation undergoing post-Fontan CMR examination between January of 2017 and January of 2023 (N=148) in Finland. Clinical and CMR parameters were analyzed using univariate and multivariate analysis. Primary endpoint was defined as death or (listing for) heart transplant. ROC-curve analysis was used for cut-off analysis.
Results: Median age at Fontan procedure was 3,2 years (IQR: 2,8-3,6 years), and median time from the Fontan procedure to the CMR was 10,8 years (IQR: 6,9 – 13,4 years). Six (4,1%) patients reached an endpoint during follow-up; two patients died, three received a heart transplant, and one was listed for transplantation. Multivariate analysis identified worse global circumferential strain (GCS) (P=0,023, 95%CI 1,04-1,74) and protein losing enteropathy (P=0,007, 95%CI 2,73-588,5) as predictors for the endpoints. The GCS cutoff value for endpoint was – 18% (sensitivity of 83,3%, specificity of 91,0%), with a positive prediction value (PPV) of 28,3% (17,1-43,0%) and a negative prediction value (NPV) of 99,2% (95,5-99,9%). In the univariate analysis worse GCS associated with increased end diastolic (P=0.006) and end systolic volumes (P=<0.001), decreased aortic blood flow (P=0.003), and decreased pulmonary arterial blood flow (P=0.018). Worse GCS was further associated with diffuse CMR myocardial fibrosis (T1-mapping), when adjusting for type of univentricular heart and time from Fontan (P= 0.03).
Conclusions: Worse GCS is a risk factor associated with death as well as (listing for) heart transplant. Moreover, worse GCS is associated with increased myocardial fibrosis on CMR T1-mapping. CMR derived GCS may serve as a valuable tool for predicting adverse Fontan outcomes in single ventricle patients, enabling early intervention strategies, and improving prognosis in this high-risk patient population.

Keywords: Fontan, univentricular hearts, CMR, myocardial strain, myocardial fibrosis

PP-523

A rare case of neonatal asymmetrical hypertrophic cardiomyopathy associated with tetralogy of fallot

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Leeds Congenital Heart Unit

Background and Aim: Although tetralogy of Fallot is a reasonably common congenital heart defect, there are only a handful of cases described with associated neonatal hypertrophic cardiomyopathy. Here we describe a neonate born at our centre with this combination of diagnoses.
Method: The baby was diagnosed with tetralogy of Fallot on fetal echo at 21 weeks gestation. Follow up at 28 and 34 weeks did not show septal hypertrophy. The mother had a normal glucose tolerance test at 30 weeks gestation and her pre-pregnancy BMI was 20.4. The baby was born at full term weighing 3.6kg.
Results: The echo showed normal atrial arrangement with the heart in the left chest and the apex pointing to the left. As expected, anterior deviation of the outlet septum was seen, creating an overriding aorta with a subaortic ventricular septal defect and infundibular pulmonary stenosis with a long narrow right ventricular outflow tract. More surprisingly there was asymmetrical hypertrophy with the interventricular septum measuring 9.8mm (z= +4.5). This was creating biventricular dynamic outflow tract obstruction with the left ventricular outflow tract obstruction originating in the mid cavity. Systolic anterior motion of the mitral valve was also noted.
Conclusions: Neonatal hypertrophic cardiomyopathy is associated with a poor outcome and due to the rapid onset, the prognosis in this case was certainly guarded. The family was extensively counselled regarding long term prognosis including the risk of sudden death and parallel planning was initiated. The baby underwent hypertrophic cardiomyopathy screening and whole genome sequencing revealed a variant of unknown significance in the ACTC1 gene. Following discharge, the baby has been followed up in the outpatient department. Aside from two acute admissions with bronchiolitis he has remained well and is approaching one year of age. His most recent echocardiogram shows that although septal hypertrophy remains, it has not progressed and the interventricular septum now measures 9.4mm (z=+3.5). The baby is now being considered for surgical repair of his tetralogy of Fallot.

Keywords: Hypertrophic Cardiomyopathy, tetralogy of Fallot, Echocardiogram

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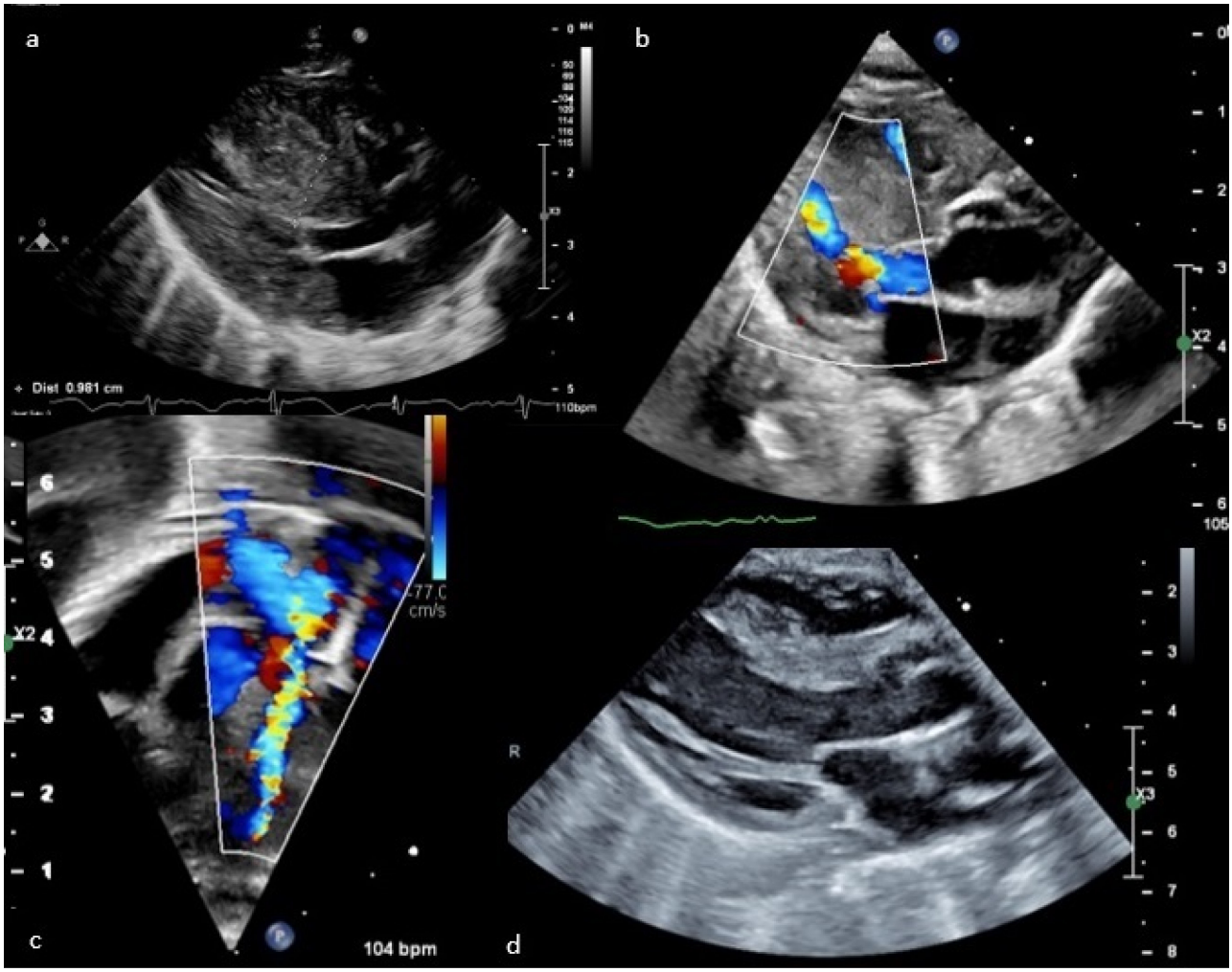
Figure 1



Echocardiogram after birth showed significant septal hypertrophy (a) with resultant near obliteration of the left ventricular cavity in systole. This created left ventricular mid cavity obstruction (b). The right ventricular outflow tract was also obstructed as a result of the anterior deviation of the outlet septum (c). At 11 months the appearance of the interventricular septum is much improved (d).

PP-524

An unusual cardiac phenotype in a neonate with noonan's syndrome

Sally Hall, Grazia Delle Donne, Chris Oakley and Antigoni Deri
Leeds Congenital Heart Unit

Background and Aim: There is a high incidence of congenital heart disease amongst patients with Noonan's syndrome, most commonly pulmonary stenosis and hypertrophic cardiomyopathy. Here we describe a neonate with a more unusual cardiac phenotype: abnormalities of all four intracardiac valves and a hypertrabeculated right ventricle.

Method: The baby was antenatally diagnosed with a CBL gene mutation associated with Noonan's syndrome. Fetal echo revealed dysplastic mitral and tricuspid valves, aortic stenosis, coarctation of the aorta and a persistent left superior vena cava (SVC). The baby was born at full term weighing 3kg. Although she was born in good condition, her respiratory status deteriorated in the first few hours of life requiring intubation and ventilation. The echocardiogram was performed following initial stabilisation and initiation of a prostaglandin infusion.

Results: The echo revealed a normal atrial arrangement with the heart in the left chest and the apex pointing to the left. There were bilateral SVCs with the left SVC draining to a dilated coronary sinus. Both atrioventricular valves were abnormal. The mitral valve had thickened, rolled leaflets and was moderately regurgitant. The posterior leaflet was shortened and partially tethered. The tricuspid valve had significant accessory tissue attached to the anterior leaflet. This accessory tissue moved with the cardiac cycle, protruding into the right ventricle in diastole and refluxing back into the right atrium on closure of the valve. The aortic valve had fusion of the right coronary and non-coronary cusps and the valve domed

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on opening. The pulmonary valve was also abnormal with thickened valve leaflets which prolapsed on closure, supra valvar pulmonary stenosis and mild regurgitation. In addition to the valvar disease the right ventricle was hypertrophied and hypertrabeculated with deep muscular crypts. There was evidence of right ventricular impairment and pulmonary hypertension with tricuspid regurgitation of 4m/s.
Conclusions: Although the echo did not reveal a duct dependent lesion, the baby was continued on the prostaglandin infusion due to her overall clinical instability. Unfortunately, she continued to deteriorate. Multidisciplinary team discussion identified that she was not a candidate for ECMO and so intensive care was withdrawn and the baby passed away peacefully at four days of age.

Keywords: Noonan syndrome, tricuspid valve, hypertrabeculated right ventricle

PP-525

Intracardiac mass such us a cardiovascular association in neurofibromatosis. The role of multimodality imaging in diagnosis and therapeutic decisions

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Department of Paediatric Cardiology, University Hospital of the Canary Islands, Canary Islands, Spain

Background and Aim: Neurofibromatosis subtype 1 (NF-1) is a neurocutaneous disease with multi-system involvement. Clinical features include cutaneous lesions (cafe-au-lait spots), benign dermal and plexiform neurofibromas and ocular malformations. Three cardiovascular associations have been recognized: hypertension, vasculopathy and congenital heart disease. NF-1 is a tumor-predisposing disease and plexiform neurofibromas are often detected in childhood, but intracardiac tumors are exceedingly rare.

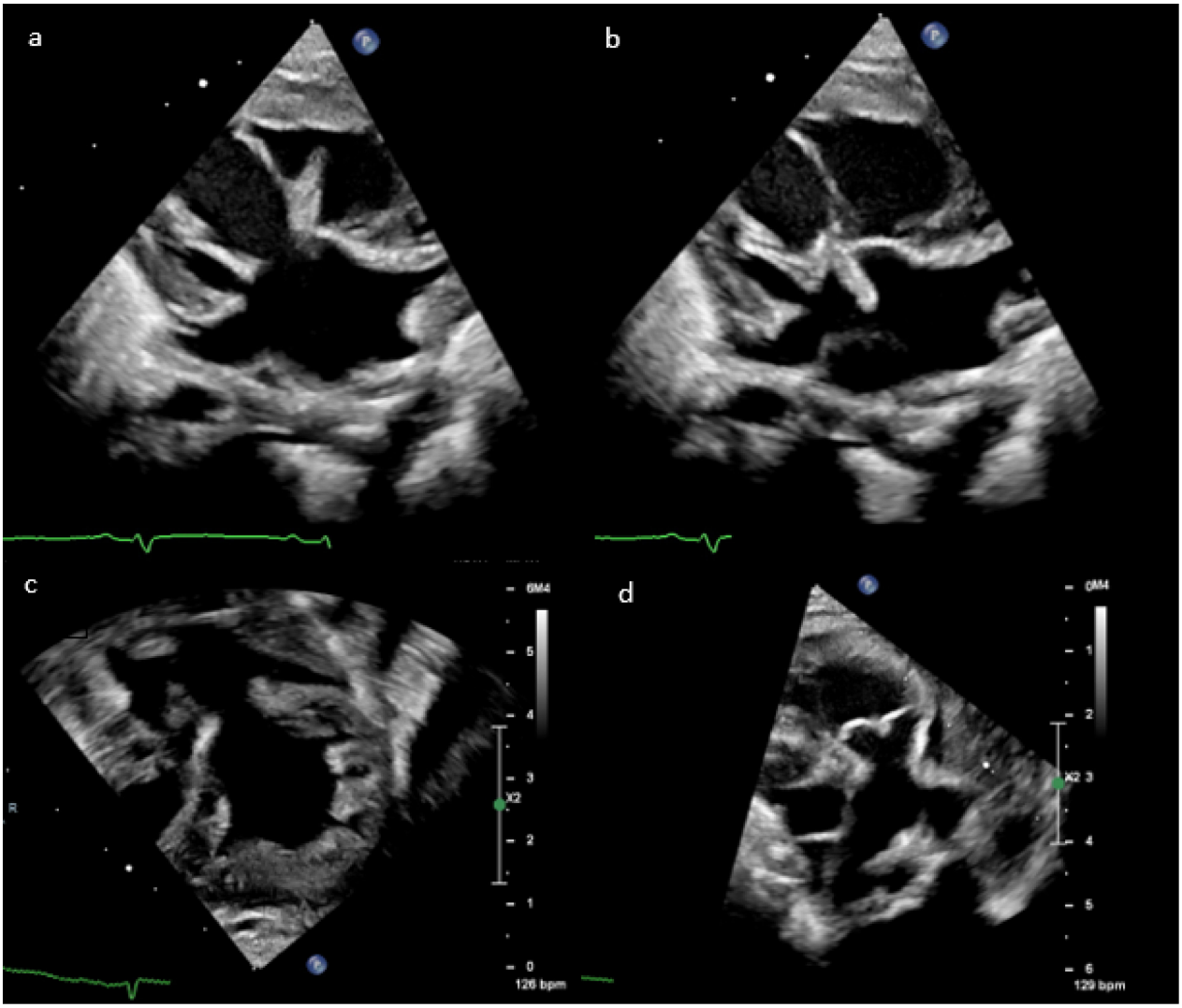

Figure 1: Images taken from the initial echocardiogram. Accessory tissue attached to the anterior leaflet of the tricuspid valve protrudes into the right ventricle on opening (a) and refluxes back into the right atrium on closure (b). The right ventricle is hypertrophied and hypertrabeculated (c). The pulmonary valve is seen to prolapse on closure (d) and there is nipping in of the supravalvar area.

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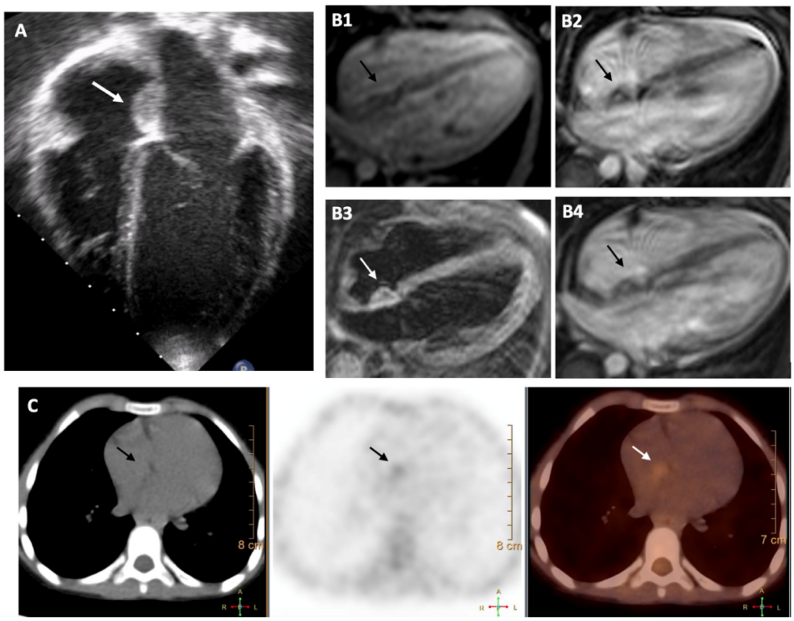
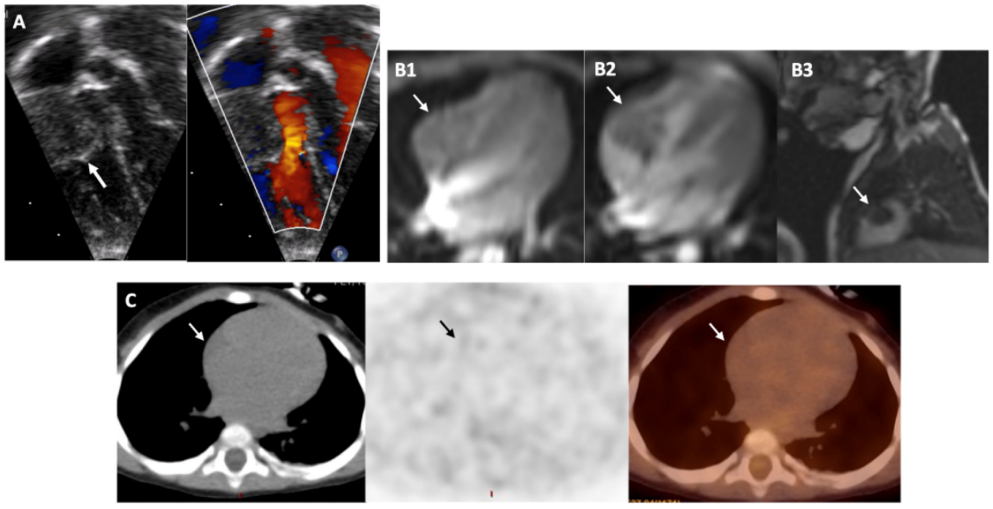
CASE 1. FIGURE 1.



CASE 2. FIGURE 2.



CASE 1. FIGURE 1. A. A well-circumscribed mass (arrow) (measured 13x8 mm) in the interatrial septum without flow obstruction seen on 4-chamber view on transthoracic echocardiography. B. The mass is isointense with the adjacent myocardium in T1 (B1) and T2-weighted (B2) imaging on CMR. No suppression of the mass on fat-water suppression (B3) on CMR. Heterogenous early uptake of contrast without late gadolinium-enhancement (B4) on CMR. C. Coronal image of 18FDG-PET/CT reveal low malignancy potential of the mass. CASE 2. FIGURE 2. A. An intracardiac mass (arrow) with lobed edges, measured about 19x17 mm and located in the lateral wall of the tricuspid annulus seen on 5-chamber view on transthoracic echocardiography. The tumor interferes with valvular flow but does not cause significant obstruction. B. On T1 (B1) and T2-weighted (B2) imaging on CMR, the mass appears slightly more intense than the myocardium. Dynamic contrast uptake study on CMR shows slight uptake in later stages. C. On the PET/CT there's no evidence of 18F-FDG avidity of intracardiac pathological significance, nor in the described lesion dependent on the atrioventricular septum and the basal segment of the right ventricular free wall.

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Our aim is to highlight the importance of the multimodality cardiovascular imaging study in two patients with NF-1 and intracardiac masses.
Method: We present two cases of patients with NF-1 in whom we identified intracardiac masses on echocardiography. To evaluate this further, an assessment by cardiac magnetic resonance (CMR) and 18F-fluorodeoxyglucose positron-emission-tomography (18FDG-PET/CT) was performed to ascertain a tissue diagnosis and to differentiate between benign and malignant characteristics.
Results: CASE-1: A 6-year-old girl with NF-1, bilateral glioma and hypertension from renal artery stenosis. In the echocardiography a homogenous tumor (measured 13x8 mm) was detected in the interatrial septum. CMR confirmed these findings showing a mass isointense with the adjacent myocardium on T1 and T2-weighted imaging and an early gadolinium contrast-enhancement. 18FDG-PET/CT detected low malignancy potential.
CASE-2: A 1-year-old boy under study due to prematurity, hydrocephaly, cafe-au-lait spots and ventricular preexcitation. The echocardiographic findings included a mass (19x17 mm) with lobed edges located in the lateral wall of the tricuspid annulus and without obstruction. On T1 and T2-weighted imaging on CMR, it appears slightly more intense than the myocardium with gadolinium-enhancement in later stages. 18F-FDG-PET/CT showed a benign behavior. Afterwards, a positive genetic result for NF-1 was obtained. In both cases, in the context of the clinical history, the presumed diagnosis is neurofibroma. Due to the mass's benign characteristics, we decided to postpone the endomyocardial biopsy.
Conclusions: - In NF-1, an early diagnosis of cardiovascular associations and an active follow-up of tumors and potential malignant transformation is key. Extensive or repetitive surgical removal for benign NF-1 is not desirable. The integration of clinical data and a multimodality cardiovascular imaging study may provide a comprehensive characterization of tissue properties of masses, and may identify malignancy parameters (growth of the mass, infiltration in surrounding tissues, metabolic activity . . .).

Keywords: Neurofibromatosis subtype 1, intracardiac mass, multimodality cardiovascular imaging, cardiac magnetic resonance, 18FDG-PET/CT.

PP-526
Gender differences in cardiovascular magnetic resonance findings in paediatric marfan syndrome

Sara Moscatelli[1], Federico Fortuni[2], Lydia Meltonlane[3], Nitha Naqvi[4], Raad Mohiaddin[3], Dudley Pennell[3], Inga Voges[5] and Sylvia Krupickova[4]
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Background and Aim: Marfan syndrome (MFS) is an autosomal dominant genetic disorder of connective tissue. In MFS adults, compromised cardiac function has been documented, and gender differences have revealed disparities. However, the full extent of this phenomenon, especially in the pediatric population, remains unclear. The objective of the present study is to investigate cardiac function and gender differences in the MFS pediatric population referred to the Cardiovascular Magnetic Unit (CMR) of the Royal Brompton Hospital.
Method: Patients <18 years receiving a CMR scan between 1st March 1999 and 1st June 2023 were included, and scans were analysed with CVI42 software. Continuous variables were tested for normality, and correspondent parametric and non-parametric analyses were conducted with a 2-sided p-value=0.05.
Results: Thirty-eight patients (median age 15 years, IQR 11-16) were included; 16 (43%) were female.
Indexed left ventricular (LV) end-diastolic volume (EDVi), indexed LV end-systolic volume (ESVi), ejection fraction (EF), and indexed mass did not statistically differ per gender (male vs female): 86±18vs92±23ml/m2, 32±7vs35±12ml/m2, 62±6vs62 ±7%, 66±12vs62±14 g/m2. LVEDVi was increased in 3 males and 9 females, LVESVi in 3 males and 10 females, EF was decreased in 2 females. Indexed right ventricular (RV) EDVi, RVESVi and EF did not statistically differ per gender (male vs female): 88±17vs87±20ml/m2,38±9vs39±13ml/m2,57±6vs56 ±9%. RVEDVi was increased in 3 females and 1 male, RVESVi in 8 females and 1 male, EF was impaired in 5 females. LV global radial strain, LV circumferential strain, and LV longitudinal strain means did not statistically differ per gender (male vs female): 30 ±7vs-29±6, -18±1vs-16±1, 17±3vs-17±3. Two patients had bicuspid aortic valve (1 male and 1 female) and 6 mild aortic regurgitation (4 males, 2 females). 9 patients had mild mitral regurgitation (5 males and 4 females), 2 moderate (1 male and 1 female) and 2 severe (both female), and 11 patients had mitral valve prolapse (9 males and 2 females). Male patients presented with mean larger ascending (22±2vs19±3mm; p=0.0069) and descending (20±2 vs 17±3mm; p=0.007) aorta diameters. *Conclusions:* No differences were found in cardiac function assessment when stratified per gender. However, ascending and descending aorta diameters appear significantly larger in males.

Keywords: Cardiovascular Magnetic Resonance, Marfan Syndrome, CMR strain

PP-527
Cardiovascular magnetic resonance changes before and after pulmonary valve replacement in pulmonary atresia and intact ventricular septum

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[1]Paediatric Cardiology Department, Royal Brompton Hospital, London, UK; [2]Cardiovascular Magnetic Resonance Unit, Royal Brompton Hospital, London, UK; [3]National Hear and Lung Institute, Imperial College, London, UK; [4]Department of congenital heart disease and Paediatric Cardiology, University Hospital Schleswig-Holstein, Kiel, Germany

Background and Aim: Pulmonary atresia with intact ventricular septum (PA-IVS) is a rare congenital heart disease. Patients with biventricular repair eventually require pulmonary valve replacement (PVR) in most cases. However, contrary to patients with Tetralogy of Fallot, currently there are no guidelines for timing of the PVR in this group.
Method: Thirty-eight consecutive patients born with PA-IVS between 1978 and 2011 at the Royal Brompton Hospital, London, who were managed with biventricular repair and underwent PVR between 1999 and 2022, were included into this study. Cardiovascular magentic resonance parameters assessed before and after PVR were: indexed right ventricular end-diastolic volume (RVEDVi), indexed right ventricular end-systolic volume (RVESVi), right ventricular ejection fraction (RV EF), pulmonary

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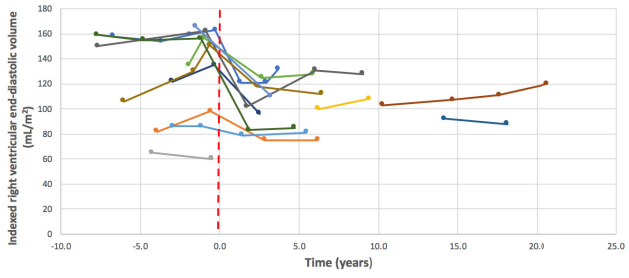
regurgitation fraction (RFPR) and right ventricular to left ventricular end-diastolic volume (RV/LV EDV) ratio. NYHA classification was used to assess symptoms.
Results: Prior to PVR, a gradual increase was noted in RVEDVi (Figure), RVESVi and RV/LV EDV ratio. No obvious trend was identified in either RVEF or RFPR. After PVR, RVEDVi, RVESVi, RV/LV EDV and RFPR decreased. Patients with initially dilated right ventricle (RVEDV ≥150 mL/m2) exhibited significant decrease in RV indexed volumes (p=0.031) when compared to individuals with restrictive physiology (p=0.250). After PVR, no significant difference was noted in RVEF. In general, symptoms reported by patients improved after PVR. Patients initially in NYHA I, continued to be asymptomatic. NYHA improved from class II to I in 7 patients. Two patients in NYHA II before PVR remained in the same class after PVR.
Conclusions: In PA-IVS, RV volumes and cardiac function gradually deteriorated pre-PVR and in some patients improved post-PVR. Two groups of patients based on their RV indexed volume were identified. Those with a dilated RV had reduced RV volume after PVR (similar to Tetralogy patients). Those with restrictive physiology had no volume improvement but did improve clinically after PVR. Guidelines for PVR in PA-IVS could be adopted similar to Tetralogy, recommending PVR in PA-IVS patients with dilated physiology (RVEDV ≥150 mL/m2) but only for symptomatic restrictive patients where RVEDV <150 mL/m2. Longterm likely multicentre observational studies would be required to assess the merits of this approach.

Keywords: pulmonary atresia with intact ventricular septum, cardiovascular magnetic resonance

Serial Measurements of Indexed RVEDV before and after Pulmonary Valve Replacement



PP-528

Myocardial work in patients with congenital heart disease and left to right shunt

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Background and Aim: In patients with left to right (L-R) shunt on ventricular/great arteries level, left ventricle (LV) is subjected to volume overload resulting in increased pump work. The aim was to evaluate LV myocardial work (MW) before and after shunt closure.
Method: MW calculation was performed in 10 paediatric patients with an isolated significant L-R shunt (ventricular septal defect=5, patent arterial duct=4, aorto-pulmonary window=1). Speckle-tracking echocardiography and the GE ECHOPAC version 204.57.0.745 software with the integrated AFI module was used to calculate the global work index (GWI) and global constructive work (GCW) from pressure-strain loops. LV biplane end-diastolic volume (LVEDV) and ejection fraction (EF) were also measured. To account for LV unloading after therapeutic intervention, post-repair GWI and GCW were also indexed to the decreased LVEDV after intervention.
Results: After shunt closure LVEDV decreased and so did the LV EF. Both GWI and GCW also decreased significantly by approximately 25% (unindexed data) and 50% (volume indexed data), resp. (see Table). The change in unindexed GWI was positively correlated with both the decrease in LV EF (R=0.662, P=0.037) and LVEDV (R=0.611, P=0.061).
Conclusions: In patients with congenital heart disease MW decreased significantly after L-R shunt closure. This decrease was proportional to volume unloading. If identical systemic cardiac output is presumed before and after intervention, LV MW expedited to sustain systemic circulation was halved after repair.
(Supported by MHCZ – DRO, Motol University Hospital, Prague, Czech Republic 00064203)

Keywords: shunt, congenital heart disease, myocardial work, speckle-tracking echocardiography, work

Table: Values of MW and other variables in patients with left-to-right shunt before and after shunt closure.

	LVEDV ml	LVEF %	GWI mmHg%	GCW mmHg%	GWI indexed mmHg%	GCW indexed mmHg%
Before (mean, SD)	23.4, 10.7	69.8, 4.2	1720.0, 365.0	2067.1, 449.9	1720.0, 365.0	2067.1, 449.9
After (mean, SD)	17.0, 9.5	51.9, 8.8	1249.2, 391.2	1554.1, 433.6	936.3, 495.4	1158.1, 578.7
P-value	0.004	<0.001	0.017	0.002	0.003	<0.001

Values of MW and other variables in patients with left-to-right shunt before and after shunt closure.

PP-529

Computational modelling of shunt flow dynamics in HLHS palliation i surgery: A comparison between blalock-taussig and sano shunt

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Background and Aim: The Norwood procedure is proposed as neonatal palliation I for hypoplastic left heart syndrome(HLHS). Pulmonary flow is provided by the Blalock-Taussig shunt(BTS) or Sano shunt, yielding different effects on pulmonary and systemic hemodynamics. This study compares the flow dynamics between both shunt types based on a 1D/3D-derived computational model.
Method: A 3D-reconstruction of the ventriculo-aortic and pulmonary artery tree is obtained through segmentation and mesh generation of a MRI of an infant after Norwood surgery. Eight geometrical shunt configurations are implemented: 6 Sano shunts of respectively 4-5-6 mm with 2 different curvatures, and 2 BTS of respectively 3-4 mm with 1 curvature. Pressure-flow profiles are obtained from 1D-cardiac cycle simulation, including arterial resistance, compliance and characteristic impedance as outlet boundaries retrieved from the 3-windkessel model. Three cardiac cycle simulations are obtained with the coupled Solver method, using 1 standard pressure value. Shunt pressure-flow characteristics and wall shear stress(WSS) distribution are calculated for all shunt

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variations, after rescaling anatomical and physiological input variables for neonatal properties.
Results: Qualitative/quantitative analysis revealed a different impact on pressures, flows and WSS as on systemic/pulmonary flow ratio and L/R-pulmonary artery(PA) flow distribution among shunt variations. Larger shunts are associated with increased pulmonary flow, with a superior effect on L/R pulmonary flow distribution for BTS. In contrast, pulmonary artery pulsatility is best preserved for Sano shunt. The impact on aortic flow between shunt types is neglectable, despite the greater pressure drop for BTS. A greater curvature of Sano shunt favors blood flow to the RPA, while BTS favors flow to the LPA. Assuming a Qp/Qs = 1 as optimal, best performance is noticed for 3 mm BTS and 4 or 5 mm Sano shunt with moderate curvature. WSS is highest at the PA inflow of the shunt for both types.
Conclusions: Integrating 1D- and 3D-model derivations, a comprehensive analysis of shunt flow characteristics allowed to differentiate the hemodynamic impact between BTS and Sano shunt, including the effect of size and curvature variations. Increased understanding of shunt dynamics through computational modelling may help to guide the decision-making at the time of palliation I surgery.

Keywords: computational model, HLHS, shunt dynamics

PP-530
Normal functional parameters in myocardial calcification over 5 years
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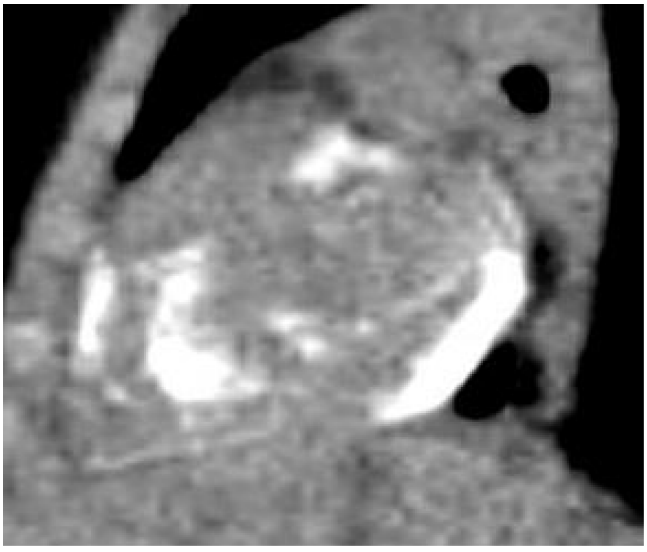
Background and Aim: Myocardial calcification has been described following myocarditis, shock, sepsis and kidney failure. This is extremely rare in children, and little is known about cardiac function. We report 5 years of follow up with normal cardiac function.
Method: Case report
Results: The boy underwent kidney transplantation at the age of three. Seven years later, he had a salmonella sepsis, during which a PET-CT was carried out. Surprisingly, myocardial calcification of the whole left ventricle was noted.

Myocardial calcification - CT findings



Myocardial calcification as seen on PET-CT

Echocardiography showed an abnormal myocardium with echodense aspect. Functional parameters incl. strain were normal.
During 5 years of follow up, neither aspect nor function changed. Clinically, the now 14 year old boy is doing well. No rhythm disturbances occurred.
Conclusions: Little is known about cardiac functional parameters in children presenting with myocardial calcification. Our case demonstrates that function can be preserved completely over years, even with abnormal myocardium.

Keywords: myocardial calcification, echocardiography, strain

PP-531
A rare case of cardiac chest pain in a young person: Minoca in the adolescent
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Background and Aim: Chest pain in children and young people is an anxiety-provoking symptom to the patient, their family and the clinician. In vast majority of children it is benign not requiring further investigation. We present a teenage female with no past medical history present with acute chest pain.
Method: Retrospective Case Review
Results: Her electrocardiogram (ECG) demonstrated inverted T waves in leads II, III, aVF, V5 and V6. Her troponin peaked at 24892 ng/L. Her echocardiogram revealed akinesia of the inferior, inferolateral and anterolateral walls of the left ventricle. She had a Patent Foramen Ovale (PFO) and an otherwise structurally normal heart. Cardiac computed tomography (CT) and coronary angiography did not show coronary artery obstruction. Cardiac Magnetic Resonance Imaging (cMRI) demonstrated extensive near-transmural enhancement of the mid-ventricular to apical inferolateral and inferior myocardium on late gadolinium enhancement. She was diagnosed with Myocardial Infarction with Non-Obstructed Coronary Arteries (MINOCA). The likely underlying causes are coronary artery spasm or transient embolic event.
Conclusions: This case demonstrates features which distinguish ischaemic chest pain from benign causes.
Although rare, it is important to understand the clinical features distinguishing cardiac chest pain from the more common benign presentations. Myocardial Infarction with Non-Obstructive Coronary Arteries (MINOCA) is a syndrome with wide number of causes, requiring careful investigation. Cardiac Magnetic Resonance Imaging (cMRI) is an excellent tool in evaluating function and myocardial injury. Paediatric ECG interpretation is an important skill for all clinicians dealing with children presenting with acute illness.

Keywords: Cardiac MRI, MINOCA, Ischaemic heart disease

PP-532
Serial changes in blood biomarkers correlate with changes in cardiac magnetic resonance imaging in patients with tetralogy of fallot
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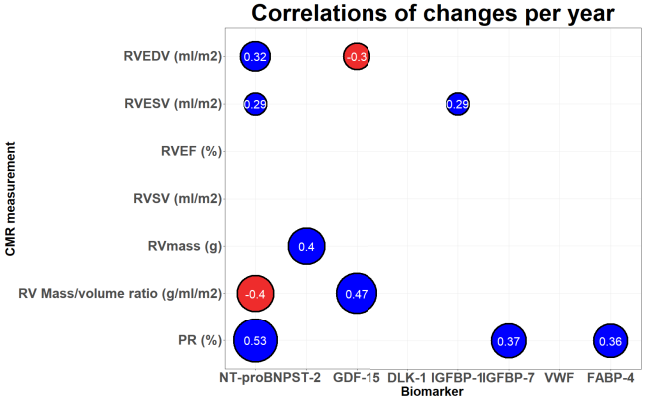
Background and Aim: Patients after surgical correction, of Tetralogy of Fallot (ToF) usually show adverse cardiac remodelling. In order to better understand the underlying biological process, we studied the relation between changes in blood biomarkers and changes in biventricular size and function as assesed by cardiac magnetic resonance imaging (CMR).
Method: This study included 50 ToF patients who underwent blood biomarker and CMR analysis on the same day, twice between 2002 and 2018, in the setting of prospective cohort studies. Time between visits was 7.0 years (IQR 5.23 – 8.27). Biomarkers examined included NT-proBNP, ST2, GDF-15, DLK-1, IGFBP-1/7, and FABP-4, all identified as relevant markers in previous studies. Markers were analysed using a panel of cardiovascular biomarkers (Olink® Target 96 Cardiovascular III, Olink, Uppsala, Sweden). Pearson correlations were used to assess the relationship between changes in biomarkers and CMR measurements of biventricular structure and function.
Results: For serial changes in parameters of right ventricular (RV) size and function we noted correlations with changes in NT-proBNP, ST-2, GDF-15, IGFBP7 and FABP-4, see figure. NT-proBNP changes correlated to changes in end-systolic (ES) and end-diastolic (ED) ventricular size, RV mass volume ratio and pulmonary regurgitation (PR). ST-2 changes correlated with RV mass, GDF-15 with RVEDV and RV mass volume ratio, IGFBP7 and FABP-4 with PR percentage. Nine patients received a pulmonary valve replacement (PVR) between two measurements. Separate analysis of both groups showed that the PVR group drove the correlations between changes in NT-proBNP and changes in RV ED and ES volumes. Correlations were found for changes in NT-pro-BNP, ST-2, GDF-15, and FABP-4 with serial changes of LV size.

Correlation coefficients for statistically significant relations between changes in blood biomarkers and CMR measurements over time.



Conclusions: Serial changes in RV size and function in patients with ToF are associated with alterations in blood biomarkers. Correlations with NT-proBNP and CMR measurements were mainly driven by patients who underwent PVR. Furthermore, correlations of GDF-15, ST2, IGFBP-1, IGFBP-7, and FABP-4 with CMR markers highlights the role of inflammation and immune response and their possible effect in ventricular remodelling in ToF.

Keywords: Tetralogy of Fallot, Biomarkers, cMRI

PP-533
Transcatheter edge-to-edge repair of tricuspid valve regurgitation in patients with systemic right ventricle: Echocardiographic outcome

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Background and Aim: Transcatheter edge-to-edge repair (TEER) provides an alternative option for high-risk patients with systemic tricuspid regurgitation (STR). Preliminary data of the French study has shown the feasibility and safety of this technique, but mid-term echocardiographic outcome is lacking.
Method: The TEER French cohort is a multicentre, longitudinal, descriptive, prospective study of patients with STR. 14 patients with severe or greater tricuspid regurgitation undergoing percutaneous repair with the MitraClip system were enrolled in the study between May 2019 and June 2023. A transthoracic echocardiography was performed at baseline, six months, one year and two years after the procedure. TR was assessed using standard 2-dimensional color Doppler methods and graded regurgitation using the 5-class grading scheme: mild, moderate, severe, massive, and torrential. The number of clips and their localization was analyzed.
Results: A reduction of at least 1 grade In TR was achieved in all subjects. TR grade remained moderate or less in 85% of patients at 1-year follow-up, and 83% of patients at 2-years follow-up. TR reduction was sustained at 2-years follow-up for all the 6 patients evaluated 5 patients had one-clip implantation: in the antero-septal coaptation line for 4 patients and in the antero-posterior coaptation line for 1 patient. 9 patients had two or more clips implantation: antero-septal/postero-septal in 2 patients, antero-septal/antero-posterior in 2 patient, 2 antero-septal in 4 patients, 2 antero-septal/antero-posterior in 1 patient. TR grade seemed to be lower in the group of patient with 2-clips strategy at 2-years follow-up: TR was graduated mild in 67% of patients in 2 or more clips group and moderate in 100 % of patients in 1-clip group.
No hospitalization for heart failure was registered during the 2-years follow up for all patients.
Conclusions: Tricuspid TEER is found to be safe and effective, with sustained effects at 2 years in patients with systemic tricuspid regurgitation. Patients with two or more clips implantation seem to have a better outcome in the regurgitation grade, but additional data and a bigger cohort are needed to predict the outcomes and to define the optimal technical strategy for TEER.

Keywords: systemic right ventricle, tricuspid regurgitation, transcatheter edge-to-edge repair

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PP-534

Exercise-induced pulmonary hypertension in patients with one-lung circulation associated with congenital heart disease

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Background and Aim: Patients with congenital heart disease sometimes have a primary or secondary severe stenotic lesion of a unilateral pulmonary artery or vein resulting in unilateral pulmonary circulation. Although most patients do not have pulmonary hypertension (PH) at rest in childhood, the change in adulthood is not clear. Exercise stress echocardiography (ESE) has recently been shown to detect latent PH at an earlier stage in cardiopulmonary disease of adults. Herein, we show the results of exercise echocardiography in young patients with severe unilateral pulmonary vessel stenosis associated with congenital heart disease.
Method: ESE was performed in two patients with unilateral pulmonary circulation. A prone ergometer was used and the workload was increased by 25 W every 3 minutes. At each stage, heart rate (HR), cardiac output (CO) calculated by velocity-time integral (cm)×left ventricular outflow tract area (cm2)×HR, and tricuspid regurgitation pressure gradient (TRPG) were recorded. Then, mean pulmonary artery pressure (mPAP) was estimated using the equation 0.6×TRPG+7mmHg, and the slope of mPAP/CO was calculated using the least-squares line.
Results: Case 1: 16-year-old female with a history of ventricular septal defect surgery. Although asymptomatic, she was recently diagnosed with severe stenosis of the left pulmonary artery by computed tomography. On ESE, maximum workload was 125W, heart rate increased from 49 to 151 bpm, and TRPG increased from 31 to 53 mmHg. The mPAP/CO slope was 3.1 mmHg/ml, indicating that she had exercise-induced PH. We planned a catheter-based intervention to relieve the stenosis. Case 2: 16-year-old female with a history of total anomalous pulmonary vein drainage. At 1 year of age, she was diagnosed with severe left pulmonary vein stenosis that was refractory to reoperation. She grew up without symptomatic problems. On ESE, maximum workload was 125W, heart rate increased from 70 to 163 bpm, and TRPG increased from 21 to 43 mmHg. The mPAP/CO slope was 3.1 mmHg/ml, indicating that she also had exercise-induced PH. The result suggested the need for regular monitoring throughout life.
Conclusions: In patients with severe stenosis of the unilateral pulmonary vessels, ESE is valuable to detect latent PH and plan patient follow-up.

Keywords: Exercise-induced pulmonary hypertension, One-lung pulmonary circulation, Exercise stress echocardiography

PP-535

Serial follow-up of aortic wall properties in hypoplastic left heart syndrome (HLHS) after norwood operation

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Background and Aim: It has been shown that hypoplastic left heart syndrome (HLHS) patients who underwent Norwood operation have different aortic wall properties compared to the healthy population. However, it is unclear how aortic wall properties change over the long-term and how this affects the clinical outcomes. Therefore, aortic distensibility was assessed in serial cardiovascular magnetic resonance (CMR) imaging studies in HLHS patients.
Method: CMR examinations of 54 HLHS patients after Fontan completion (female: 17) more than 10 years after Norwood operation were retrospectively analysed. Only patients who had two examinations were included. Patients with surgical re-interventions of the aorta were excluded. CMR cine images were used to measure distensibility of the ascending aorta (AAo), isthmus and descending aorta (DAo). Right ventricular (RV) enddiastolic and endsystolic volumes (RVEDV, RVESV), myocardial mass (RVMM) and functional parameters (RV ejection fraction and cardiac index) were measured. Neo-aortic regurgitant fraction (RF) was assessed from phase-contrast flow measurements. Temporal changes of distensibility and its relationship with RV volumes, function parameters and neo-aortic RF were analysed.
Results: The first CMR examination was performed at 4.6 (2.9-25) years and the second at 16.4 (10-27) years. Distensibility of the AAo was significantly lower compared to distensibility at the isthmus and DAo at both examinations (all $p<0.01$). Furthermore, there was a significant decline in distensibility from the first to the second examination for all aortic levels: AAo 3.0±1.2 10-3mmHg-1 vs. 1.9±1.1 10-3mmHg-1, isthmus 10.7±4.2 10-3mmHg-1 vs. 6.7±2.3 10-3mmHg-1, DAo 14.3±4.3 10-3mmHg-1 vs. 8.1±2.7 10-3mmHg-1 (all $p<0.001$). There were no significant correlations between distensibility and RV function parameters. A significant increase in indexed RVEDV (78±23 ml/m2 vs. 111±27 ml/m2) and RVESV (35±15 ml/m2 vs. 57±24 ml/m2) as well as a decrease in RVEF (56±9% vs. 50±10%) and indexed RVMM (68±23 g/m2 vs. 56±21 g/m2) were noted. No changes for cardiac index and neo-aortic RF were observed.
Conclusions: After Norwood operation, AAo distensibility is highly reduced compared to distensibility at other thoracic aortic levels and declines further with age. Although no relationship with RV function parameters was found, our data suggest that aortic and RV surveillance seems mandatory.

Keywords: Cardiovascular magnetic resonance imaging, hypoplastic left heart syndrome, Norwood operation, aortic distensibilty

PP-536

Greater papillary muscle angle and shortened chords are associated with left atrioventricular valve failure in partial atrioventricular septal defect

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Background and Aim: Re-operation for left atrioventricular valve regurgitation (LAVVR) after partial atrioventricular septal defect (pAVSD) repair is associated with significant morbidity. Preoperative risk factors such as LAVVR, double orifice LAVV, single papillary muscle, dysplastic mural leaflet and early age at surgery are well established. A proportion of patients without any of

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these features develop LAVVR and need re-operation. Preoperative identification of these patients is challenging.
This study aimed to utilise quantitative three-dimensional echocardiography (3DE) to explore preoperative LAVV annular, leaflet and subvalvar features associated with postoperative LAVV failure.
Method: A case-control study of patients without established risk factors undergoing pAVSD repair with preoperative 3DE from 2006-2022 was undertaken. Of 82 patients, 26 (31.7%) with post-operative moderate or greater LAVVR at follow-up or LAVV re-operation (LVR) were compared to 26 age and size-matched controls (CON). Quantitative analysis of the LAVV annulus, leaflets and subvalvar apparatus was performed using a custom 3DE software (MATLAB). The LAVV annular dimensions, leaflet area, annular sphericity index, annular bending angle, anterior papillary muscle (APM) angle relative to ventricular centroid, and chordal length were measured. The LAVV was segmented into the superior (SBL) and inferior (IBL) bridging leaflets and mural leaflet (ML) for regional leaflet areas. Areas and circumferences were indexed to BSA0.86 while linear dimensions were indexed to BSA0.45 for comparisons.
Results: No difference in patient age and size. LVR patients had a more circular annulus with greater annular area, greater total leaflet areas and a larger overall IBL compared to CON. The APM angle to ventricular centroid was also greater in the LVR group with associated shorter chord length (Figure 1). No significant differences in annular or leaflet areas, annular bending angle, or papillary muscle length were identified between groups (Table 1).
Conclusions: Quantitative pre-operative 3DE of the LAVV in pAVSD without known risk factors demonstrates that patients who develop post-operative LAVVR have a less ellipsoid annular shape, mild annular dilation and an anterior papillary muscle that has a greater angle with shorter chords. Development of commercially available tools for 3DE quantitative assessment of subvalve apparatus maybe provide important surgical risk stratification of otherwise "normal" pAVSD.

Keywords: 3-dimensional echocardiography, 3D echo, partial AVSD, left AV valve regurgitation, left AV valve reoperation

Cardiovascular Morphology

PP-537

Case report of a newborn with a significant fistula from the right coronary artery to the right ventricle in a Newborn

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Background and Aim: Anomalies of the coronary arteries may lead to potentially life threatening myocardial ischemia. Early detection might help to secure a sufficient coronary circulation, but even if detected early an individual therapy regimen must be performed. Coronary fistulas in infancy have the potential for both, spontaneous regression and growth.
Method: Case Report

TABLE 1:

LAVV CHARACTERISTIC	LVR (N=26)	CON (N=26)	P VALUE
AGE (YEARS)	2.15 (0.42 – 5.4)	2.0 (1.22 – 5.05)	0.65
BSA (M^2)	0.53 (0.32 - 0.79)	0.51 (0.39 – 0.81)	0.56
AGE AT FOLLOW UP (YEARS)	11.04 (6.84 – 16.22)	11.59 (8.04 – 16.48)	0.67
LAVV REOPERATION	12	0	
ANNULAR AREA (CM2/M2)			
SBL	2.37 +/- 0.51	2.27 +/- 0.68	0.57
ML	2.05 +/- 0.63	1.77 +/- 0.81	0.17
IBL	2.15 +/- 0.78	1.57 +/- 0.74	**<0.01**
TOTAL	6.59 +/- 1.27	5.62 +/- 1.80	**0.03**
ANNULAR INDEX	1.18 +/- 0.09	1.25 +/- 0.12	**0.03**
BENDING ANGLE	145 +/- 14	146 +/- 13	0.69
LEAFLET AREA (CM2/M2)			
SBL	2.61 +/- 0.58	2.57 +/- 0.86	0.84
ML	2.26 +/- 0.74	1.87 +/- 0.81	0.08
IBL	2.44 +/- 0.87	1.82 +/- 0.84	**0.01**
TOTAL	7.34 +/- 1.58	6.27 +/- 2.18	**0.05**
SUBVALVAR APPARATUS			
ANTERIOR PM ANGLE TO VENTRICULAR CENTROID	31 (20 – 60)	27 (24 – 35)	0.38
< 36 DEGREES	13	16	**0.04**
≥ 36 DEGREES	10	2	
INDEXED PM + CHORDAL APPARATUS LENGTH	2.26 +/- 0.44	2.4 +/- 0.54	0.41
INDEXED CHORDAL LENGTH	1.12 +/- 0.22	1.32 +/- 0.26	**0.01**

* reported values are median (IQR) or mean +/- SD

FIGURE 1:

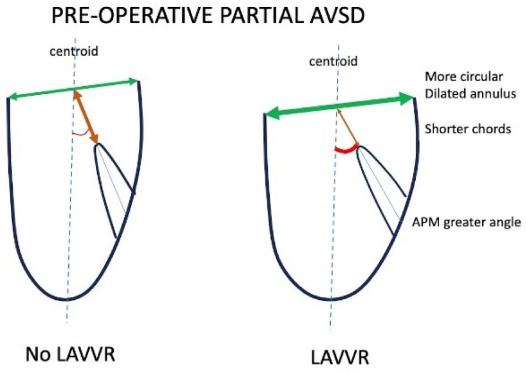

Table 1 - comparison of patient demographics and LAVV characteristics on 3D echocardiography by group Figure 1 - LAVV subvalvar apparatus (anterior papillary muscle angle to ventricular centroid, chordal length) by group

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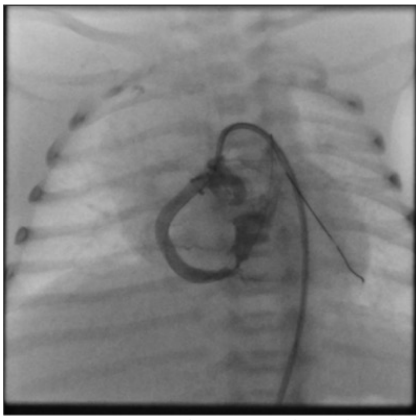
Results: A newborn boy with normal postnatal adaption was referred to our clinic with a heart murmur and a suspected anomaly of the right coronary artery (RCA) in the echocardiography. ECG showed ST lowering in V1-V3, while Troponin was within a normal range. Echocardiography at our clinic confirmed a significant anomalous fistula of the right coronary artery in the right ventricle with a continuous systolic-diastolic flow pattern. Cardiac catheterization proved the coronary fistula of the RCA to the right ventricle (RV) with relevant side branches supplying the right ventricle with blood up to the very RV orifice (Figure 1). Therefore, an interventional closure was not performed, and the patient received a conservative therapy with modulation of the heart frequency with bisoprolol and a platetet aggregation inhibitor (Acetylsalicylic acid). The following check-ups every 4 weeks showed that the fistula became smaller over time and the patient stayed asymptomatic.
Conclusions: Coronary fistulas are rare but can be life threatening if undetected and hence untreated. Symptoms can be absent and develop at a later stage, which makes the diagnosis even harder. In the presented patient, the coronary fistula was detected early, but a percutaneous closure was avoided due to coronary side branches of the fistula up to the orifice providing the right ventricle with blood supply. While surgical or interventional closure can be necessary in cases of large fistulae, a wait-and-see approach in our case seemed to be the right decision for that patient because of decreasing size of the fistula during the follow-up.

Keywords: Coronary artery fistula, congenital, heart defects, neonate

Figure 1



Angiography shows a significant coronary fistula of the RCA to the right ventricle with relevant side branches supplying the right ventricle with blood.

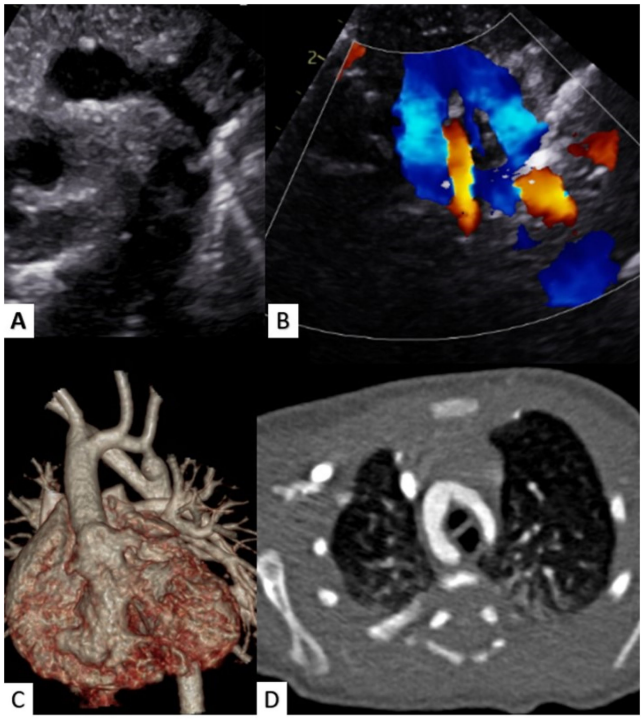
PP-538
Rare case of D-transposition of the great arteries (TGA), ventricular septal defect (VSD), double aortic ARCH (DAA) and bilateral SVC

Melonie Johns, Arul Narayanan, Alexandra Williams and Attilio Lotto
Alderhey Children's Hospital, Liverpool, UK

Background and Aim: The majority of D-TGA with intact ventricular septum have a left aortic arch (96%), whereas, those with associated VSD or PS have an increased incidence of right aortic arch (11% and 16% respectively). Up to 15% of D-TGA have associated arch obstruction, coarctation or interruption, however vascular rings are rarely associated. These can be challenging to diagnose. In D-TGA, this is further complicated by the anterior position of the aorta, and the diminutive arch could be mistaken for an hypoplastic aortic arch.
Vascular rings can be caused by double aortic arch or an arch with contralateral PDA or ligamentum arteriosum, and may not be clear until after the arterial switch operation. There have been 12 cases of vascular ring associated with D-TGA reported in the literature; 9 had double arch. 2 died in infancy from respiratory distress secondary to tracheobronchomalacia. All other patients were discharged home, one with a tracheostomy.
Method: We report a rare case of D-TGA with VSD, double aortic arch, left PDA and bilateral SVC. The patient was felt to have a RAA antenatally. Postnatal transthoracic echo demonstrated a slender left aortic arch with large PDA. After balloon septostomy, prostin was stopped and PDA closed spontaneously. The left arch appeared to have coarctation but with a normal doppler trace and easily palpable femoral pulses. A double aortic arch with ipsilateral carotid and subclavian arteries was diagnosed with right arch dominance, as demonstrated on echo and CT images.
Results: The patient underwent arterial switch operation with division of the left arch posterior to the left subclavian on CPB via median sternotomy. The apical VSD was not reachable and left open. The patient had a calculated Qp:Qs of 3:1 so a PA band was applied during the same setting. The patient had an uneventful recovery and has been discharged home without tracheal or oesophageal symptoms and remains well.
Conclusions: Despite increasing antenatal cardiac surveillance clinicians should remain suspicious if transthoracic echocardiographic imaging doesn't match the clinical picture. CT or MRI should can be used to determine the dominant arch anatomy prior to surgical division at the time of the arterial switch operation.

Keywords: Transposition, Vascular ring, double aortic arch

Image and legend



A) TTE of left arch with distal narrowing. B) Suprasternal TTE demonstrating double aortic arch.
C) Volume rendered 3D and D) axial images from contrast enhanced ECG-gated CT demonstrating double aortic arch with residual left ductal ampulla.

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PP-540

Prognostic factors for hypertrophic cardiomyopathy in newborns of diabetic mothers

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Background and Aim: Maternal diabetes affects the fetal heart. It causes a unique form of hypertrophic cardiomyopathy. It is usually self-limiting, but can be life-threatening in severe and obstructive forms. The aims of our study is to determine the impact of hypertrophic cardiomyopathy in newborns and prognostic factors.
Method: A retrospective descriptive study identifying all cases of hypertrophic cardiomyopathy in infants of diabetic mothers.
Results: 50 cases of hypertrophic cardiomyopathy were identified. 18 % of the mothers had type 1 diabetes, 22 % of the mothers had type 2 diabetes. The rest (60 %) developed gestational diabetes. 41% of the pregnancies were complicated by hydramnios. Macrosomia was noted in 86% of cases. 88% of the newborns presented respiratory distress. Tachycardia was observed in 23% of the neonates. Cardiac auscultation showed a systolic murmur in 54% of cases and a gallop rhythm in 9% of cases. Cardiomegaly was noted on chest X-ray in 19 % of cases. The electrocardiogram revealed an elevated Sokolow index in 3 neonates. Echocardiography showed asymmetric hypertrophic septal cardiomyopathy in 78%, global in 22%, and obstructive in 34 % of the cases. 50 % of the neonates underwent propranolol treatment with a mean treatment duration of 5.7 months. Fifteen newborns benefited from a follow-up echocardiogram after an average period of 5.4 months. No abnormalities were noted. The evolution was favorable. Only one death occurred, caused by severe respiratory distress due to a severely obstructive hypertrophic cardiomyopathy. the severity of hypertrophic cardiomyopathy was correlated with type 2 diabetes and unbalanced diabetes
Conclusions: Hypertrophic cardiomyopathy is frequently observed in infants of diabetic mothers. Prevention is based on promoting diabetes screening and ensuring pre-conception diabetes control in women with pre-existing diabetes.

Keywords: Hypertrophic cardiomyopathy, diabetes, newborn

PP-541

Platypnea-orthodeoxia in a newborn with ductus venosus agenesis

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Background and Aim: Platypnea-orthodeoxia syndrome is characterized by dyspnoea and arterial desaturation, defined by a drop in oxygen saturation of more than 5% while in the upright position, which improves upon recumbency, often associated with an extracardiac or intracardiac shunt including atrial septal defect. It is well-known in the elderly but its description in children has been seldom and it has not been depicted in infants. This syndrome is described here in a neonate.
Method: After informed written consent from the parents, data were collected about the child's medical history and outcome.
Results: A 3.2-kg boy was diagnosed prenatally with ductus venosus agenesis, the inferior caval vein was found to connect normally to the heart. Abdominal ultrasound at birth confirmed the ductus venosus agenesis with a hypoplastic left portal branch, left hepatic hyper-arterialization, no portosystemic shunt and direct connection of the umbilical vein to the right atrium confirmed on computed-tomography-scan. Due to persistent cyanosis, lung perfusion scintigraphy depicted an extrapulmonary shunt. Echocardiography-doppler showed a deformation of the tricuspid valve without significant tricuspid regurgitation, a hypertrophy of the right ventricular, a dilatation of the left ventricle and aortic root, a prominent Eustachian valve, and an atrial septal aneurysm. There was a right-to-left atrial shunt during upright position and crying with a concomitant drop of the pulsed oxygen saturation to 70%, but a left-to-right shunt laying back at rest with normal saturation. No pulmonary arteriovenous malformations were found and haemoglobin was normal. Mean saturation progressively improved from 83% on room air at 1 month of age to 91% at 6 months with normal development and growth, associated with the decrease in ascending aorta diameter Z-score.
Conclusions: Platypnea-orthodeoxia syndrome was depicted for the first time in a newborn and infant, when the upright position echocardiography is rarely performed. Other causes of cyanosis were ruled out by multimodal imaging. Although normally caused by acquired disorders, congenital ductus venosus agenesis with direct umbilical vein connection to the heart led to abnormal heart anatomy favouring right-to-left shunt associated with intermittent increased venous return and right atrial pressure.

Keywords: Paediatric, case report, platypnea-orthodeoxia, Arantius, neonatal

PP-542

A rare case of an infant with congenital giant ductus arteriosus aneurysm and coronary aneurysms during kawasaki disease

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Background and Aim: Ductus arteriosus aneurysm (DAA) is a rare congenital cardiac anomaly with potentially severe complications. Although frequently identified through fetal echocardiography and typically self-resolving, the precise etiology remains unknown. We present a unique case of neonatal giant DAA regression, followed by Kawasaki disease, characterized by the development and subsequent near regression of coronary artery aneurysms. This represents the first documented report of such a phenomenon involving aneurysm dynamics.
Method: We present the case of a neonate with a giant ductus arteriosus aneurysm (DAA) identified shortly after birth. The infant had an uneventful prenatal history with no reported familial congenital heart diseases. Clinical assessment revealed subtle symptoms such as pallor and mild respiratory distress. The infant therefore

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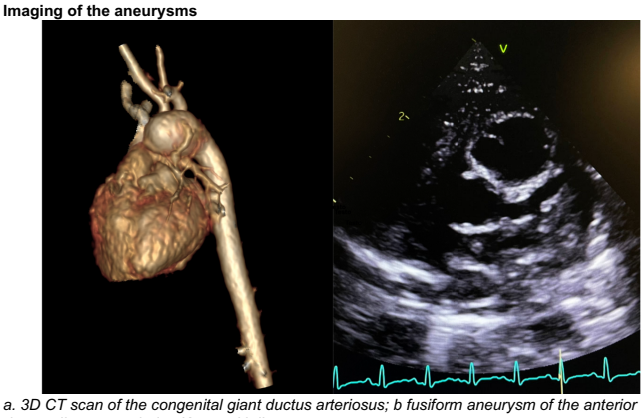
underwent transthoracic cardiac ultrasound, which identified a DAA measuring 23x15 mm. A CT scan was also performed to exclude any signs of tracheal compression or erosion of the DAA. After nearly three months, the DAA entirely resolved spontaneously following intravascular thrombus formation within the aneurysm. However, a few weeks later, the infant displayed symptoms indicative of an incomplete form of Kawasaki disease. Diagnostic evaluation revealed an aneurysm in the right coronary artery measuring 2.4x2.8 mm (z-score 3.4) and a fusiform aneurysm in the anterior descending artery measuring 7x2.94 mm (z-score 7.07). The patient underwent immunoglobulin, prednisone, and antiplatelet therapy, leading to clinical improvement and near regression of the coronary artery aneurysms.
Results: Our case report emphasizes the benign natural progression of DAA in neonates, advocating for a conservative management approach in the absence of critical signs. Additionally, it underscores the possible relationship between DAA and Kawasaki disease, a domain that remains poorly understood due to the limited patient data available in the literature.
Conclusions: Long-term follow-up studies on individuals who experienced DAA during childhood is essential to evaluate the potential long-term vascular comorbidities and the interrelationship between these two medical conditions. Additional future research including genetic analysis, is needed to investigate the potential arterial predisposition for the development of significant vascular aneurysms.

Keywords: Ductus Arteriosus Aneurysm, Coronary Artery Aneurysm, Kawasaki Disease



a. 3D CT scan of the congenital giant ductus arteriosus; b fusiform aneurysm of the anterior descending artery during Kawasaki disease

PP-544

Concomitant pulmonary artery banding as heart failure therapy in patients undergoing congenital heart surgery

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Department of Cardiac Surgery, Fuwai Hospital, Chinese Academy of Medical Sciences, Peking Union Medical College, Beijing, PR China

Background and Aim: The most common primary cardiac diagnosis of pediatric heart failure (PHF) is congenital heart defect (CHD). Pulmonary artery banding (PAB) led to an improvement of LV function by ventricular interaction in young children with dilated cardiomyopathy. However, it remains obscure whether PAB can improve cardiac function in PHF patients with CHD. Hence, we aimed to report our preliminary results of PAB, concomitant to CHD surgery, as PHF therapy.
Method: Since Jun 2021, PAB was performed in consecutive PHF patients with primary diagnosis of CHD in our center. The indication of PAB was severe preoperative LV dysfunction 1) couldn't be fully attributed to primary CHD and 2) with poor response to anti-HF medication. Timing of PAB was concomitant to surgery repairing CHD. Perioperative management and surgical technique was comply with instructions from Giessen.
Results: A total of 2 patients meet the inclusion criteria. One of them was an 8m old female, diagnosed as ventricular septal defect (peri-membranous 5mm) with a 44% of LVEF and 44mm of LVEDv. During surgery, LVEF was not improved after cardiopulmonary bypass for VSD repair (40%). Hence, additional PAB was performed followed with an increased of LVEF (51%). At 6 months follow-up, her LVEF improved to 58%, and left ventricular systolic function recovered well. Another patient was a 4-month-old boy with coarctation (CoA) and severely impaired LV function. However, both the arch imaging and pressure gradient suggest the CoA was mild. Hence, Coarctation repair and concomitant PAB was performed through left posterolateral thoracotomy without cardiopulmonary bypass. Six months later, his LVEF improved from about 20% to 68%. At the latest follow-up, there was no signs of right ventricular overloading or hypoxemia noted in these 2 patients.
Conclusions: Based on our limited experiences, concomitant PAB may be helpful for LV function recover in PHF patients whose primary diagnosis was CHD.

Keywords: Congenital heart defect, Heart failure, Pulmonary artery banding

PP-545

Comparative histology of tissue-engineered and autologous pericardial patches in pediatric heart surgery

Katja Eildermann[1], Joachim Photiadis[2], Kira Weixler[2], Matthias Sigler[1] and Peter Murin[2]
[1]Department of Pediatric Cardiology and Intensive Care Medicine, Georg-August University Göttingen, Göttingen, Germany; [2]Department of Congenital Heart Surgery - Pediatric Heart Surgery, Deutsches Herzzentrum der Charité, Berlin, Germany

Background and Aim: Cardiovascular surgical procedures frequently necessitate the use of patch material. This study compares the tissue responses to various types of pericardial patches, harvested from children with congenital heart defects.
Method: Patches made from tissue-engineered bovine (n=7) and equine (n=7) sources, as well as autologous human pericardium (n=7), were used for vascular reconstruction. These were later explanted either due to functional issues or as part of planned follow-up procedures. To assess defined aspects of biocompatibility, the tissues were stained using Movat Verhoeff to visualize tissue composition and Von Kossa for detecting calcifications. Various immunohistological stainings were conducted to identify specific immune cells and other cellular components within the tissues.
Results: We found distinct, functional differences in the tissue reactions to different patch materials. In bovine patches, degradation was marked by the disintegration of collagen fibers, often accompanied by the accumulation of exudate. Equine tissues displayed edematous swelling and material dissolution. Both bovine and equine patches had marked accumulations of macrophages at the patch-tissue interface, a phenomenon that was minimal in autologous patches. Bovine patches showed luminal thrombus adherence and pronounced calcifications. In equine patches the foreign body reaction was most prominent. Notably, human patches displayed only minimal signs of inflammation, while pannus formation was similar across all patch types.

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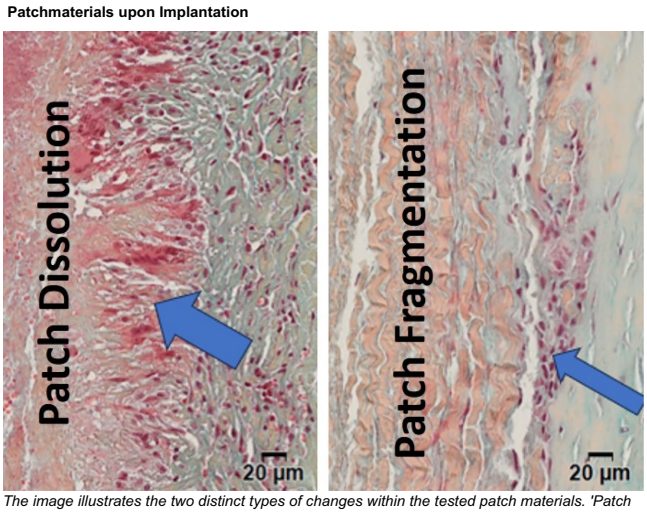
Conclusions: Our findings highlight the superior biocompatibility of autologous human pericardial patch material compared to tissue-engineered alternatives, and emphasize the distinct reactions observed in bovine and equine patches.

Keywords: Pericardial patches, Congenital heart defects, Biocompatibility, Immunohistological staining, Tissue-engineered materials, Vascular reconstruction



The image illustrates the two distinct types of changes within the tested patch materials. 'Patch Dissolution' is depicted, where the patch material appears amorphous and swollen, signifying a loss of structural integrity. Immune cells are unable to infiltrate but accumulate at the material's edge. In contrast, 'Patch Fragmentation' is displayed, characterized by fragmented material with cells infiltrating the spaces in between, leading to the displacement of patch material fragments.

PP-546

A novel biocompatibility scoring system for cardiovascular patches

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[1]Department of Pediatric Cardiology and Intensive Care Medicine, Georg-August University Göttingen, Göttingen, Germany; [2]Department of Congenital Heart Surgery - Pediatric Heart Surgery, Deutsches Herzzentrum der Charité, Berlin, Germany

Background and Aim: We developed a scoring system to facilitate comparison of tissue reactions to various biological patch materials in patients with congenital heart disease (CHD).
Method: Biological vascular patches were retrieved from patients who had undergone corrective surgery for CHD. Explated tissues underwent staining procedures using Movat Verhoeff for tissue composition visualization and Von Kossa for calcification detection. Immunohistological stainings were utilized to pinpoint specific immune cells and other cellular components within the tissues We employed a semi-quantitative scoring system to evaluate multiple facets of biocompatibility, including thrombus formation on the luminal surface, pannus thickness and inflammation, cellular responses at the material-tissue interface, patch degradation, calcification, and neoadventitial inflammation. Each sample received a set of seven aspect scores, with the average of these seven determining the overall biocompatibility score for that sample.
Results: Our scoring methodology enabled nuanced comparisons of biocompatibility between individual explants. Aggregating the aspect scores for each patch type allowed us to compare the performance of different materials.
Conclusions: This scoring system provides a comprehensive framework for evaluating and comparing various facets of biocompatibility among different biological materials. It serves as a valuable tool for both assessing patch types and guiding their development.

Keywords: Congenital Heart Disease, Biological Patch Materials, Biocompatibility, Scoring, Foreign Body Reaction, Histopathology

PP-547

Effects of additional pulmonary flow in bidirectional cavopulmonary connection on subsequent staged total cavopulmonary connection

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[1]Department of Pediatrics, Faculty of Medicine Siriraj hospital, Mahidol University, Bangkok, Thailand; [2]Department of Surgery, Faculty of Medicine Siriraj hospital, Mahidol University, Bangkok, Thailand

Background and Aim: Many debates over whether bidirectional cavopulmonary connection (BCPC) with additional pulmonary blood flow (APBF) is beneficial. We investigated the influence of APBF on pulmonary artery (PA) growth, pulmonary arterial pressure (PAP), ventricular end diastolic pressure (EDP), presence of pulmonary arteriovenous malformation (AVM) and complications post total cavopulmonary connection (TCPC).
Method: Retrospective review of 104 patients who underwent BCPC with subsequent TCPC at Siriraj Hospital between 2006 and 2022 was performed. The patients were categorized into 2 groups by their BCPC physiology; (1) with APBF (n=26) and (2) without APBF (n=78). Changes in the size of PA, McGoon ratio, PAP, EDP, AVM were compared by using pre-BCPC and pre-TCPC cardiac catheterization data. Post-TCPC complications and survival of the two groups were explored.
Results: In group 1 and 2, the median age (IQR) at BCPC were 1.41(1.73), and 1.54 (2.19), respectively (p=0.42), while age at TCPC were 7.62 (3.02), and 8.50 (2.73), respectively (p=0.908). During a median time of 5.08 years between pre-BCPC and pre-TCPC assessment, increased size of RPA and LPA in group 1 (2.90±0.80 and 2.37±0.60 mm, p<0.05) presented, compared to group 2 (1.83±0.33 and 1.04±0.31 mm, p<0.05). However, there was no statistically increase in the McGoon ratio between groups. In pre-TCPC assessment, mean PAP and EDP in group 1 (16.11±2.28 and 12.36±2.39 mmHg) were slightly higher than group 2 (14.48±2.58 and 11.89±3.21 mmHg) (p=0.456, p=0.635). Unexpectedly, oxygen saturation pre-TCPC notably improved in group 1 (81.04%± 7.42) when compared with group 2 (78.88%±6.27) (p=0.05). Pulmonary AVM was reported in 1 patient from group 1 and 7 patients from group 2 (p=0.395). There were no differences in-hospital stay and mortality post-TCPC. At a median follow-up time of 10.37 years (3.4-17.7 years), 2 patients in group 1 and 4 in group 2 deceased

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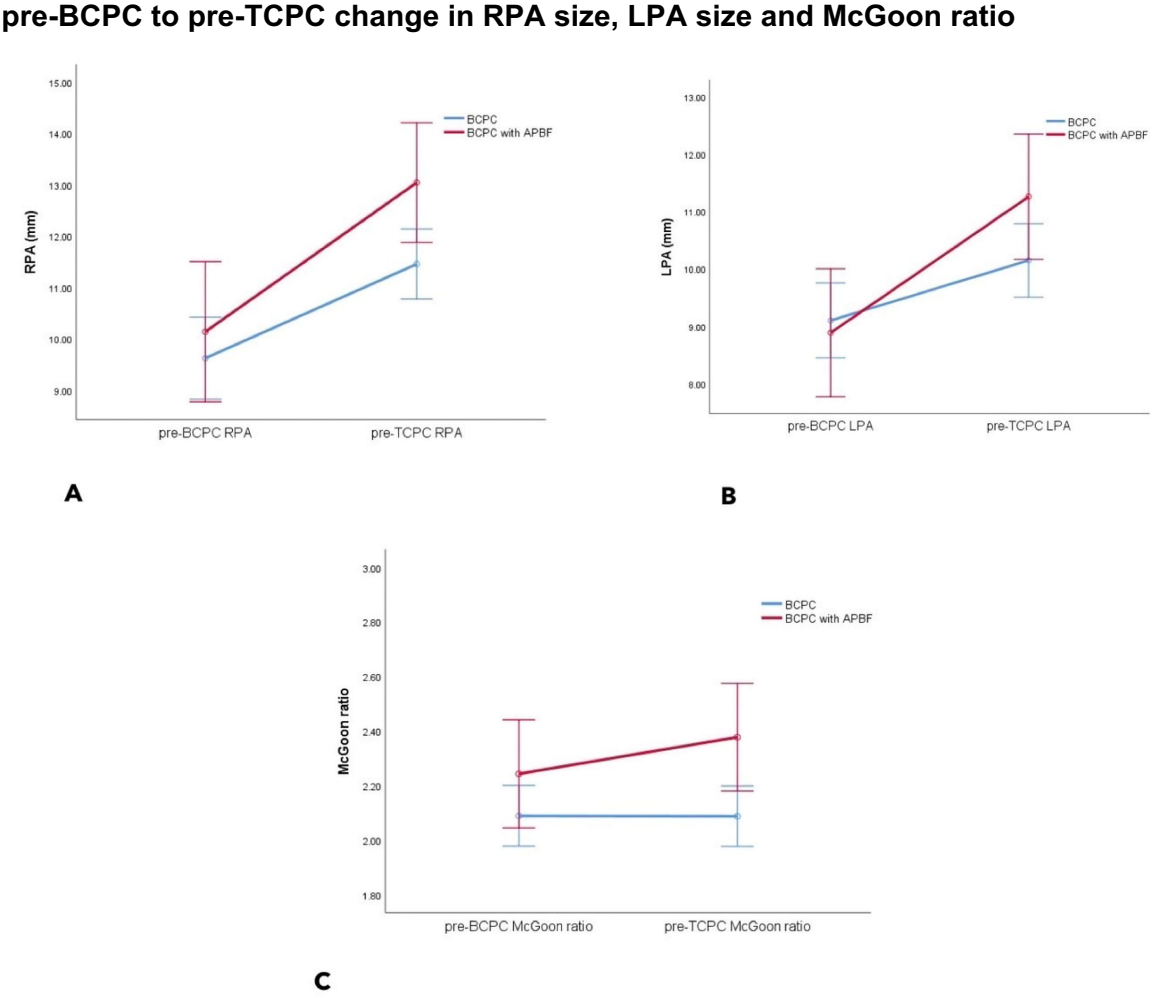

Change in pre-BCPC to pre-TCPC of RPA size (A), LPA size (B) and McGoon ratio (C) between two groups, which are BCPC (in blue) and BCPC with APBF (in red) were shown

(p=0.88). In addition, the 5- and 10-year survival was 96% and 91.4% in group 1, and 97.4% and 95.6% in group 2, respectively (log-rank; p=0.78).
Conclusions: BCPC with APBF meaningfully improves oxygen saturation and pulmonary growth with a cost of mildly increase in PAP and ventricular EDP. Nevertheless, it does not impact on TCPC complications or survival.

Keywords: Single ventricle, antegrade pulmonary blood flow (APBF), pulsatile Glenn, bidirectional cavopulmonary connection (BCPC), total cavopulmonary connection (TCPC)

PP-548
Factors affecting long-term outcomes after fontan: Rationale and design of the pediatric cardiac care consortium fontan cohort study
Lazaros L Kochilas[1], Yanxu Yang[2], Divya Suthar[1], Asaad Beshish[1], Mathew E Oster[1] and Jessica H Knight[3]
[1]Emory University, Atlanta, Georgia, USA and Children's Healthcare of Atlanta, Atlanta, Georgia, USA; [2]Emory University, Atlanta, Georgia, USA; [3]University of Georgia, Athens, GA, USA

Background and Aim: In the absence of a national US registry for congenital heart surgeries, we linked the Pediatric Cardiac Care Consortium (PCCC), a US-based multi-institutional registry, with the National Death Index through December 2022 to understand patient-level factors at time of Fontan that may affect long-term outcomes. Here, we present the rationale and design for the PCCC Fontan Cohort Study.
Method: We queried the PCCC registry for patients enrolled between 1982 and 2003 and underwent Fontan procedure at 1-21 years of age. We included patients with a cardiac catheterization within 6-12 months prior to Fontan and had Glenn physiology at the time of the evaluation. Patients with atrio-pulmonary Fontan were excluded. In addition, we excluded patients with other cardiac interventions between cardiac catheterization and Fontan, chromosomal abnormalities or missing forms.
Collected data encompassed demographics, ventricular morphology, comorbidities, and operation characteristics. Hemodynamic variables underwent statistical tests for normality, with extreme outliers removed. Correlations were assessed using Spearman's rank correlation (rs), and Kaplan-Meier survival plots depicted unadjusted long-term survival estimates. The proportional hazards

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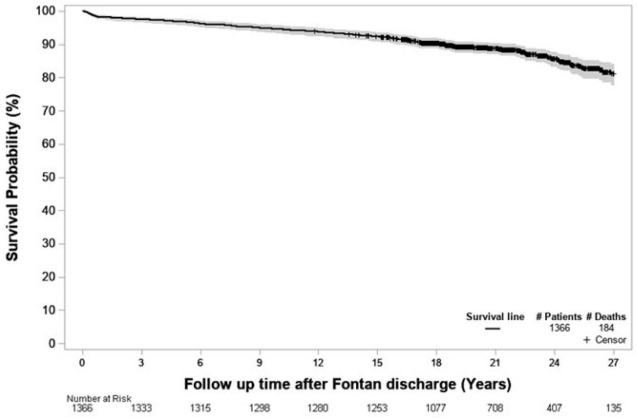
assumption was evaluated graphically using Schoenfeld residuals. We used generalized linear mixed models to assess the associations of risk factors with in-hospital Fontan failure (deaths and Fontan take-downs) and Cox proportional hazard models for post-discharge events reporting hazard ratios (HRs) with 95%CI, considering center as a random effect. The follow-up period commenced upon date of hospital discharge and ended at the date of a death match event or the end of the follow-up period, whichever occurred first.
Results: A total of 1461 patients met eligibility criteria from 43 centers and were included in the study; 1366 were discharged with Fontan physiology. Most hemodynamic variables were slightly only deviating from a normal distribution (skewness range: |0.25-0.98|). There was strong correlation between SVEDP and mPAP (rs 0.542), TPG and PVR (rs 0.714) and CI and SVO2 (rs 0.566). The 25-year survival post Fontan discharge reached 83.7% (95% CI: 81.2-86.3%) (Figure).
Conclusions: The PCCC Single ventricle cohort demonstrates an excellent survival at 25 years after the Fontan procedure for all lesions and offers a unique opportunity for systematic analysis of factors affecting post Fontan long-term outcomes.

Keywords: Fontan, outcomes, single ventricle

Figure:



Kaplan-Meier survival estimate for patients after Fontan discharge in the Pediatric Cardiac Care Consortium cohort.

PP-549

A qualitative review of current-era mortality after bilateral pulmonary artery banding in a single-centre

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Background and Aim: Bilateral pulmonary artery banding to create a "hybrid" circulation is usually performed as a safer temporising measure in patients with borderline left heart structures to allow somatic growth prior to committing to biventricular or univentricular pathways, or in patients with planned single ventricle pathway who are either too unstable, or too underweight to undergo the Norwood operation. Our aim was to qualitatively review recent mortality after bilateral pulmonary artery banding at our centre.
Method: We reviewed the records of all patients who underwent bilateral pulmonary artery banding at our centre in the last five years. Medical records of all cases of mortality were qualitatively reviewed regardless of timing of death in relation to the procedure.
Results: A total of 28 patients underwent bilateral pulmonary artery banding from 2018 to 2023. Of those 28 patients, six died, two of which died within 30 days of the procedure (7% 30-day mortality). Three patients died with hybrid circulation: one patient underwent banding due to unbalanced atrioventricular septal defect and prematurity and died on the day of the procedure following hypotension then cardiac arrest. The second died of Serratia sepsis seven weeks after banding, and the third died following redirection of care after a significant genetic diagnosis. Of the remaining three, the first died suddenly two weeks after Norwood procedure due to BT shunt thrombosis. The second died 18 months after initial banding from intracranial haemorrhage and sepsis following debanding of their pulmonary arteries and re-stenting of their ductus arteriosus while on the heart transplant list as they were unsuitable for either univentricular or biventricular pathways. The third patient underwent biventricular repair nine months after pulmonary artery banding. They died nine weeks post-operatively with autopsy concluding that dense fibrosis and dystrophic calcification in the left ventricle and septum were due to ischaemia during the long operative repair with bypass unable to meet the demands of the hypertrophied myocardium.
Conclusions: One potentially modifiable cause of mortality related to the hybrid procedure may include avoiding the development of significant ventricular hypertrophy prior to subsequent

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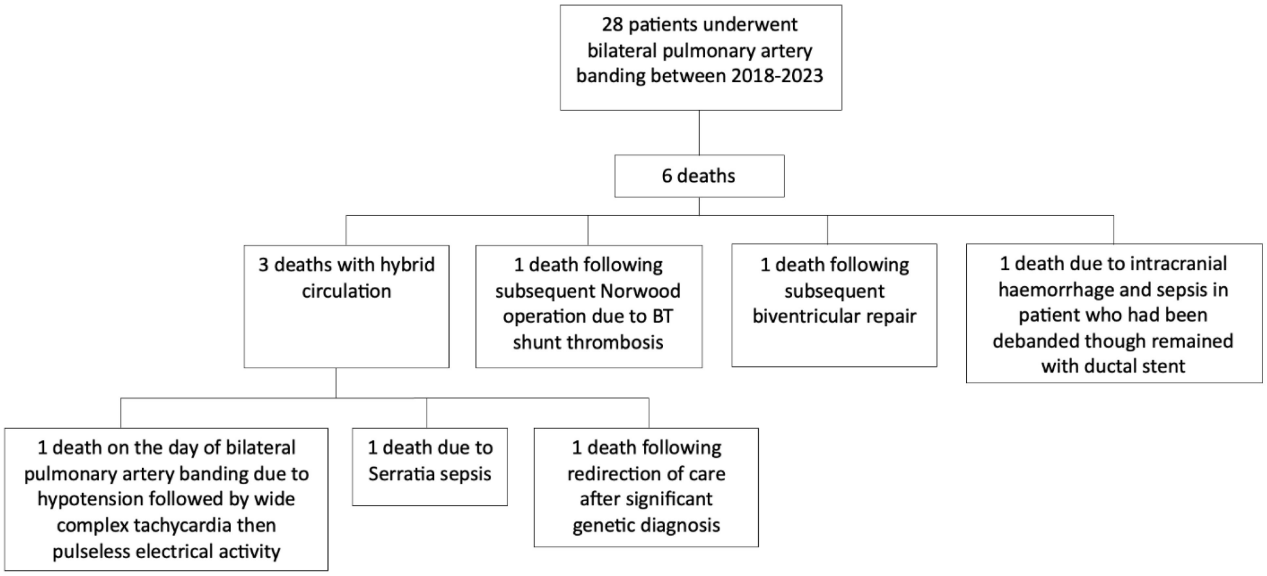
procedures. This may need to be taken into consideration when planning timing of subsequent interventions.

Keywords: Bilateral pulmonary artery band, Hybrid

Image 1



A visual representation to supplement the results

PP-550

Surgery of congenital coronary anomalies in children: 25 years of experience

Luz Polo[1], Juvenal Rey[1], Raúl Sánchez[1], Bunty Ramchandani[1], Blanca Torres[1], Álvaro González[1], Íñigo Velasco[1], Paula Burgos[1], Jorge Utanda[1], Lucía Deiros[2], Montserrat Bret[3], Gemma Campo[4], Juan José Menéndez[5] and Ángel Aroca[1]

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Background and Aim: Congenital coronary arteries anomalies are rare, accounting 2,2% of all congenital heart disorders. If they present clinically during childhood, usually produce cardiac failure and dilated cardiomyopathy. Diagnosis and treatment are challenging. We present our surgical experience with these patients.
Method: Retrospective analysis of 15 children operated in our Unit along 1998-2023. Diagnosis was made with Echocardiography 100%, cardiac catheterization 80%, cardiac CT 40% and myocardial viability test 33%. After diagnosis, surgery was indicated in order to establish a two-coronary system circulation.
Results: Patient age was from 1 month to 12 years, median: 37 months [IQR 5-108], median weight 12 kg [IQR 6-34], 67% females. Two patients (13%) had previous surgeries. Preoperative studies showed anomalous coronary artery origin from the pulmonary artery in 13(86%), anomalous coronary artery origin from aorta in 1(7%) and left coronary ostium atresia in 1(7%). One third of the children were asymptomatic, 47% had symptoms of cardiac failure or acute myocardial ischemia 20%. Median left ventricle ejection fraction (LVEF) was 58% [IQR 28-70], with moderate/severe ventricular dysfunction in 47%. We observed significant mitral regurgitation in 33% (3-moderate, 2-severe). All kids were operated with extracorporeal circulation, moderate hypothermia and aortic cross-clamp. Respect surgical techniques, direct coronary reimplantation are our gold standard, used in 73% (Figure-1). None patient received associated mitral surgery. Median intubation time was 3h [IQR 1-51] and median hospital stay 9 days [IQR 6-18]. Inhospital mortality was 6,7% (a boy, operated 20 years ago, died in the operative room after Takeuchi without ventricular assistance availability). Other girl had an intraoperative myocardial infarction after coronary reimplantation, was supported with ECMO, and later received a heart transplant. These two patients had very low preoperative LVEF. Median follow-up is 11 years [IQR 2-17]. Nowadays 14 patients are alive and well (including the girl with the heart transplant), without reoperation, LVEF is 65% [IQR 61-65]. Mitral insufficiency improved along time (14% moderate regurgitation).
Conclusions: We recommend surgery in children diagnosed of congenital coronary artery anomaly. Direct coronary reimplantation without mitral valve surgery is our preferred technique because it has good short and long-term results.

Keywords: Congenital coronary arteries anomalies, children, surgery

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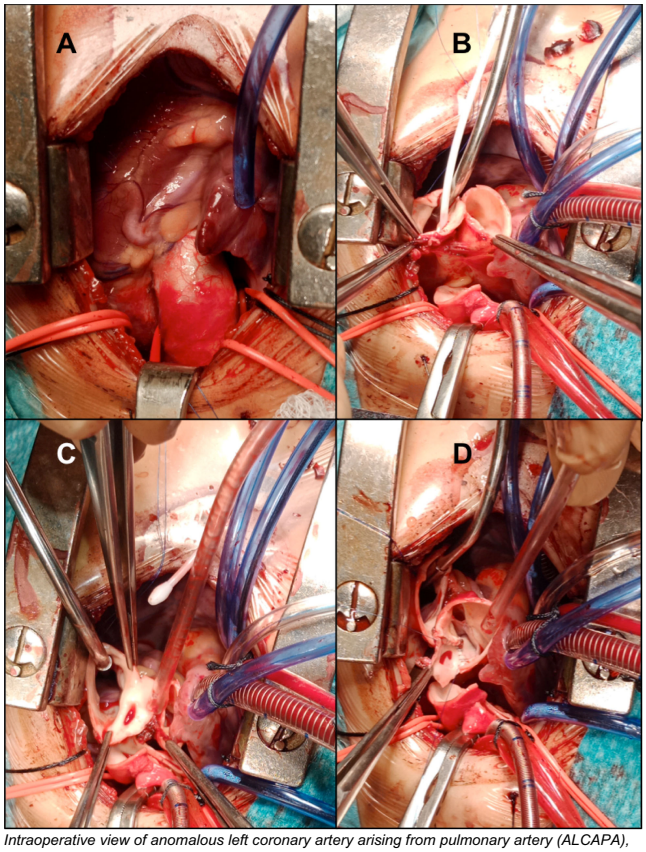
ALCAPA AEPC 2024



Intraoperative view of anomalous left coronary artery arising from pulmonary artery (ALCAPA), before cannulation for cardiopulmonary bypass (A), and with heart arrested and great vessels transected (B). In C we can see the left coronary ostium arising from the pulmonary artery. In D the left coronary artery is being reimplanted with running suture into the aorta.

PP-551
Heart operations in patients with dextrocardia: Our surgical challenge

Luz Polo[1], Raul Sánchez[1], Juvenal Rey[1], Blanca Torres[1], Bunty Ramchandani[1], Álvaro González[1], Iñigo Velasco[7], Jorge Utanda[7], José Ruiz[6], Carlos Labrandero[2], Montserrat Bret[3], Raquel Pérez[4], Paula Burgos[7], Cristina Verdú[5] and Ángel Aroca[1]
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Background and Aim: Congenital heart disease and dextrocardia is a rare entity. Cardiac surgery in this setting is challenging and sometimes associates high inhospital mortality and morbidity. We present our surgical experience with these patients.
Method: Retrospective analysis of 23 patients (11 adults, 12 children) operated in our congenital heart unit along 2009-2023. Statistical study was done with SPSS-20.
Results: Patient age was from 4 days to 57 years, median: 12 years [IQR 0,5-30], median weight 34 kg [IQR 6,5-71], 35% females. Thirteen patients (57%) had previous surgeries. Clinical presentation was cardiac failure-57%, hypoxia-30% and 13% were asymptomatic Preoperative cardiac diagnosis was: single ventricle 9 (39%), septal defects 6 (26%), congenital valvulopathy 5 (22%), transposition of great arteries 2 (9%) and Fallot 1 (4%). The auricular situs was solitus (48%) and inversus (52%). The visceral situs was solitus (43%), inversus (48%) and ambiguous (9%). Heterotaxia was present in 25% patients. Median sternotomy was performed in 22 patients and right thoracotomy in one. All operations were done with extracorporeal circulation and moderate hypothermia, 19 needed aortic cross-clamp. Surgical techniques are detailed in Table-I. Median extracorporeal time was 128 minutes [IQR 85-200] and aortic cross clamp time 60 minutes [IQR 32-150]. Median intubation time was 10 hours [IQR 4-192] and median hospital stay 14 days [IQR 7-22]. Inhospital mortality was 13% (Two patients with failing Fontan died after heart transplant, other died after myocardial infarction complication of Bentall surgery). All were adults, needing ECMO in the postoperative course, and finally died from multiorgan failure. Complications in postoperative stay were seen in 10 patients (44%). Median follow-up is 5,6 years [IQR 2-10], with null late mortality. Five patients (25%) needed reoperation and also two of them needed percutaneous intervention in the follow-up. Nowadays 19 patients are alive and well in functional class I-II, and one patient progressed to functional class III.
Conclusions: Congenital heart surgery in patients with Dextrocardia comprises a wide spectrum of pathologies with different levels of complexity. An individualized approach in a specialized congenital heart unit makes possible to achieve good results with an acceptable mortality, but morbidity remains high.

Keywords: Congenital heart surgery, Dextrocardia

Tabla I Dextrocardia AEPC 2024

Systemic-pulmonary Fistula	2	Ventricular septal defect closure	3
Glenn	3	Atrial septal defect closure +/- pulmonary venous anomalies	6
Extracardiac Fontan	2	Valvular surgery	8 (5 prosthesis, 3 repair)
Heart Transplant in Failing Fontan	3	Fallot repair	1
Stansel	1	Arterial switch operation	1
Epicardial pacemaked lead	4	Right ventricle outflow resection / arterial duct ligation	5

Table-I: Surgical techniques applied in Dextrocardia patients

PP-552
Evaluating techniques of aortic coarctation treatment in children: A 11-year single-centre experience

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Background and Aim: The aim of this study was to utilize and leverage a single center database in order to evaluate both the approach and the outcomes of coarctation of the aorta (CoA) management for patient treatment.
Method: Retrospectively reviewed medical database from January 2011 to December 2021 of 166 patients with CoA. Variables reviewed included type of coarctation, associated cardiac anomalies, type of repair, early outcomes and late outcomes.
Results: The 166 patients (M/F: 68.1%) had a median follow-up of 8.0 years (IQR 4, 11.9 years, mean 8.46 years ± 5.37 years). In 47 patients (28.3%), an isolated type of CoA was detected; in 119 patients (71.7%), a complex type of CoA, associated with other cardiac defects, was reported. Two most common associated cardiac defects were bicuspid aortic valve, found in 80 (48.2%)

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patients and hypoplastic aortic arch, found in 51 (30.7%) patients. With regards to surgical treatment, it was performed in 133 patients (80.1%). The prevailing surgical technique was end-to-end anastomosis (EEA 84, 63.2%), followed by patch (43, 32.3%). Catheter based intervention was performed in 33 patients (19.9%). Moreover, one patient with Turner syndrome died in the early postoperative period, otherwise no deaths were registered. Recoarctation occurred in 74 patients (44.6%), thereof none after initial EEEA, in 35 patients (47.3%) after initial EEA (without patch), in 18 (24.3%) after initial balloon dilatation, in 4 (5.4%) after initial stenting and in 13 (17.6 %) after patch repair. The results also indicate that treatment strategy is not significantly associated with the onset of arterial hypertension (P = 0.0734).
Conclusions: The results point towards the suggestion that the treatment strategy and the overall management of aortic coarctation should be individualized for every patient depending on 3 factors: associated cardiac defects, age at intervention and center experience. Moreover, newer and improved surgical technique EEEA proved to be beneficial, with less recoarctations than the older EEA technique. On the other side, transcatheter repair is a minimally invasive procedure with less complications; nevertheless, the somatic growth of the patient could lead to expected recoartations.

Keywords: Aortic coarctation, Congenital heart disease, Surgical repair, Transcatheter repair, Congenital heart surgery

PP-553
Right ventricular outflow tract reconstruction in patients with fallot-type lesions: Transannular patch insertion versus bicuspid valve construction

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Background and Aim: For patients of Fallot-type lesions with pulmonary valve hypoplasia, reconstruction of the RVOT often requires a transannular patch (TAP). Since 2008, our Institution has used valve-sparing approaches with bicuspid valve(bv) construction with decellularized extracellular matrix (ECM). The objective of this study was to evaluate mid-term outcomes in children using two different techniques.
Method: Retrospective single-center study review evaluating 51 children with TOF-type lesions who underwent palliation with either TAP insertion (TAP group; n=23) or BV construction (BV group; n=28) over 10-year period. The median age at surgery was 6.1 months (range, 11 days to 3.5 years). Reintervention was defined as any surgical or percutaneous catheter procedure on the valve or RVOT. Structural valve deterioration (SVD) was defined as development of a peak pressure gradient equal to or greater than 40 mmHg or/and a moderate pulmonary insufficiency on follow-up echocardiography. Procedure-related morbidity, mortality and reintervention rates were assessed and compared.
Results: ECM was used for RVOT reconstruction as a TAP in 44% of patients (TAP group) and as a bicuspid in 100% of patients (BV group). The two groups were similar in gender, age, weight, previous valve intervention, diagnosis, cardiopulmonary bypass and cross-clamp times, and length of stay (Table). Follow-up completeness was 96%, and the median duration of follow-up was 38 months (range, 0.8-175). The follow-up time of the most recent echocardiogram was similar between the groups (p=0.28). The degree of pulmonary insufficiency was significantly lower in BV group (BV, 2.2+1.2 vs. TAP, 3.3+0.8; p<0.001), however, the peak RVOT gradient was significantly lower in the TAP group (TAP, 19.5+9.5 mmHg vs. BV, 34.3+27; p=0.01). Seventeen patients underwent RVOT reintervention (transcatheter or surgical; TAP, 24% vs. BV, 43%; p=0.14). Nine-year freedom from SVD was similar between the groups (TAP, 30% vs. BV, 43%; p=0.40).
Conclusions: RVOT reconstruction in patients with Fallot-type lesions with pulmonary valve hypoplasia using a TAP or BV construction demonstrated good mid-term clinical outcomes. Both techniques can provide a functional valve in the immediate and mid-term postoperative period.

Keywords: Tetralogy of Fallot, Outcomes, Right Ventricular Outflow Tract

Table

variables	TAP group	BV group	P value
Gender (male/female)	9/12	16/12	0.39
Age at surgery (months)	8.8+10	10.4+10	0.59
Weight at stent (kg)	3.5+1.1	3.8+3.1	0.54
Diagnosis			
TOF	14 (67%)	19 (68%)	0.97
DORV	2 (10%	3 (11%)	1.00
PA/VSD	3 (13%)	2 (7%)	0.85
Others	2 (10%)	4 (14%)	0.77
Preoperative PV repair (#, %)	6 (21%)	11 (39%)	0.55
Cardiopulmonary bypass time (min)	123+50	138+44	0.27
Aortic cross-clamp time (min)	80+36	92+42	0.28
Length of stay (days)	14.9+15	14.9+17	0.99
Follow-Up time (months)	43.2+49	29.2+33	0.28
Gradient on PV (mmHg)	19.5+9.5	34.3+27	0.01
PI grade	3.3+0.8	2.2+1.2	0.001
Branch PS (#, %)	3 (14%)	9 (32%)	0.19
Reintervention on PV (#, %)	5 (24%)	12 (43%)	0.14

LEGEND: DORV – double outlet right ventricle; PA – pulmonary atresia; PI – pulmonary insufficiency; PS – pulmonary valve stenosis; PV – pulmonary valve; TOF – tetralogy of Fallot; VSD ventricular septal defect

PP-554
Identification of collagen-based tissue and an appropriate treatment method for the production of a viable autologous heart valve prosthesis

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Background and Aim: Heart valve prostheses are frequently derived from biological tissue, predominantly of xenogenic or allogeneic origin, necessitating treatment pre-implantation. Regardless of the tissue's origin, the treatment procedure encompasses chemical cross-linking to confer non-antigenicity, augment mechanical strength, and mitigate tissue degradation. Nonetheless, the employment of foreign tissue and chemical cross-linking leads to the implantation of a non-viable structure which in turn leads to several drawbacks, such as calcification, inflammatory degradation, mechanical susceptibility, and a restricted capacity for matrix regeneration. Cumulatively, these factors contribute to the gradual structural degeneration of the prosthesis. To overcome the

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previously mentioned limitations, it is necessary to develop a viable prosthesis, which requires the utilization of appropriate autologous tissue in combination with a gentle treatment procedure. This study assesses various collagen-based tissues as potential candidates and an appropriate treatment method for producing a viable heart valve substitute.
Method: To identify an appropriate material we assessed different collagen-based tissue, namely pericardium, fascia lata, and peritoneum. Therefore, we first quantified the matrix contents and mimicked our desired cross-linking degree utilizing biochemical assays and differential scanning calorimetry. Subsequently, the treated tissues were characterized by comprehensive assessments, encompassing analyses of mechanical properties and fiber alignment. Furthermore, tissue samples were treated with different biological enzymatic agents and assessed appropriately.
Results: Our results show that different tissues can be utilized for the production of a heart valve prosthesis which exhibits mechanical properties within the normative ranges. Compared to chemical cross-linking the assessed alternative methods result in a diminished degree of cross-linking.
Conclusions: With the combination of utilizing autologous tissue in conjunction with a non-chemical treatment method, a durable heart valve prosthesis could emerge. We were able to show that various tissues can be used for this purpose. However, if different tissues are used in the future, their respective composition must be taken into account by adapting the treatment accordingly to achieve the desired cross-linking degree. Furthermore, we were able to demonstrate the capability of biological agents to crosslink collagen-based tissue. However, their specificity needs to be adapted to achieve the desired cross-linking degree.

Keywords: Implantation, Congenital Heart Disease, Biomaterial, Heart Valve Prosthesis, Collagen

PP-555

3D-printed molds to obtain silicone hearts with congenital defects for pediatric heart-surgeon training

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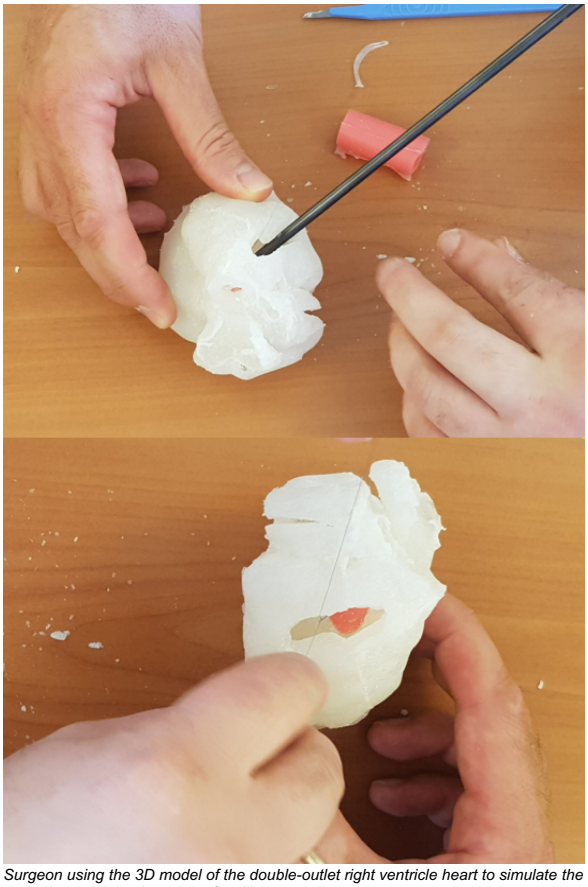
Background and Aim: Many types of congenital heart disease are amenable to surgical repair or palliation. The procedures are often challenging, and require specific surgical training, with limited real-life exposure and often costly simulation options. Our objective was to create realistic and affordable 3D simulation models of the heart and the vessels to improve training.
Method: We created molded vessel models using eights materials, in order to identify the material that best replicated human vascular tissue. This material was then used to make more vessels to train residents in cannulation procedures. Magnetic resonance imaging views of a 23 months-old patient with double-outlet right ventricle were segmented using free open-source software. Re-usable molds produced by 3D printing served to create a silicone model of the heart, with the same material as the vessels, which was used by a heart surgeon to simulate a Rastelli procedure.
Results: The best material was a soft elastic silicone (Shore A hardness 8). Training on the vessel models decreased the residents' procedural time and improved their grades on a performance rating scale. The surgeon evaluated the molded heart model as realistic and was able to perform the Rastelli procedure on it. Even if the valves were poorly represented, it was found to be useful for preintervention training.
Conclusions: By using free segmentation software, a relatively low-cost silicone, and a technique based on re-usable molds, the cost of obtaining heart models suitable for training in congenital-heart-defect surgery can be substantially decreased.

Keywords: surgical simulation, congenital heart disease, 3D segmentation, 3D modeling, 3D printing, silicone model

3D model of DORV



Surgeon using the 3D model of the double-outlet right ventricle heart to simulate the Rastelli procedure, with implantation of a silicone patch

PP-559

Criss-cross heart – a case with dextrocardia, transposition of the great arteries and ventricular septal defect

Nelli Korpela[1], Saara Lehtiranta[1], Ilkka Mattila[2], Teemu Mäkelä[3], Laura Martelius[3] and Tiina Ojala[4]
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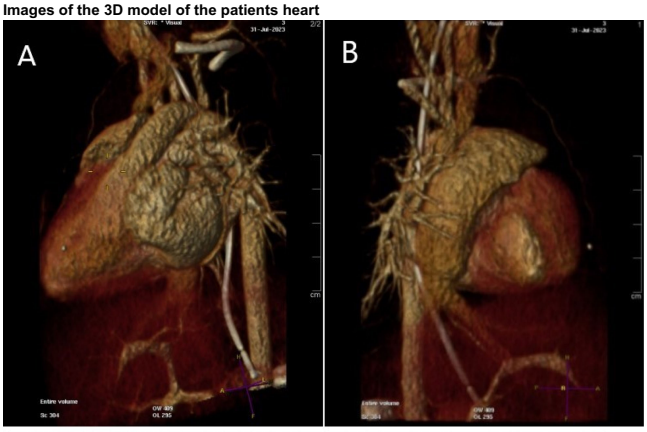
Background and Aim: Criss-cross heart is a rare congenital cardiac malformation characterized by crossing of the atrioventricular connections due to a longitudinal twisting of the ventricular mass and is customarily associated with various additional cardiac anomalies. The complexity of the cardiac morphology in such cases present a substantial challenge for both diagnosis and surgical intervention.
Method: We present a case report of a patient with criss-cross heart and complex additional anomalies.
Results: The patient was suspected to have dextrocardia, criss-cross atrioventricular concordance and ventriculoarterial disconcordance (transposition of the great arteries) as well as a ventricular septal defect (VSD) in antenatal fetal echocardiography. Following delivery, the diagnosis was confirmed by transthoracic echocardiography as well as a computed tomography scan. A three-dimensional (3D) reconstruction of the heart was modeled to facilitate in understanding the unique cardiac anatomy in this rare defect, in this case especially the ventricular sizes. Prostaglandin infusion was started after birth. The patient began to desaturate, and surgical care was initiated at 12 days of age. Arterial switch operation and VSD closure were successfully preformed. The postoperative echocardiography showed an excellent surgical result. The patient has been followed up by a pediatric cardiologist until 2,5 months of age and the clinical condition of the patient has been up to standard. The patient has been asymptomatic with normal growth. Echocardiographic findings have shown a small residual VSD with no effect on the function of the heart, no narrowness at steam points and well-functioning valves.
Conclusions: 3D modeling of the heart was valuable in understanding the unique anatomy of this complex cardiac anomaly and enabled preoperative planning, contributing to a desirable clinical outcome.

Keywords: criss-cross heart, congenital heart defect, echocardiography, three-dimensional modeling, case report

Images of the 3D model of the patients heart



Still images of the 3D model of the patients heart. Different views with the aorta (A) and artrial appendage (B) visualized.

PP-560

The impact of coronary anatomy on early mortality in patients undergoing arterial switch procedure

Ensar Duras[1], Samet Paksoy[1], Okan Yildiz[2], Hacer Kamali[1], Aslıhan Karaman[1], Ismihan Selen Onan[2], Alper Guzeltas[1] and Sertac Haydin[2]
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Background and Aim: This study aims to investigate the effect of coronary artery anomalies on early mortality, intensive care unit stay, and overall hospital length of stay in patients diagnosed with transposition of the great arteries (TGA) and Taussig-Bing anomaly (TBA) who underwent ASO.
Method: This retrospective study included 251 patients who underwent ASO due to TGA with intact ventricular septum (TGA-IVS) or ventricular septal defect (TGA-VSD) and TBA between November 2010 and December 2022. Patients with left ventricular outflow tract obstruction were excluded from the study. Patients were divided into two groups: Group I (n=181, usual coronary artery) and Group II (n=70, unusual coronary artery). Coronary artery anomalies were documented from preoperative echocardiograms (ECHO). Coronary anatomy was defined according to the "Leiden Classification."
Results: The median age in Group I was 8 days (1-1440 days), while in Group II, it was 17.5 days (1-900 days). The rate of coronary anomalies by diagnosis was 23.8% in TGA, 28.7% in TGA-VSD, and 50% in TBA. Coronary anomalies were present in 17.9% of patients on preoperative ECHO. Based on the frequency of coronary anomalies in Group II patients, 28.6% were 1L;2RCx, 21.4% were 2LCxR, 18.6% were 1LCxR, 11.4% were 1LR;2Cx, 8.6% were minor anomalies, and 11.4% were categorized as other anomalies. Three patients had intramural courses of coronary arteries, two of whom died. TThe rate of patients requiring extracorporeal membrane oxygenation (ECMO) support was 11% in Group I and 18.6% in Group II. The mortality rate was 8.3% in Group I and 11.4% in Group II (Table 1). The presence of coronary anomalies did not show a significant impact on early mortality. In patients with coronary anomalies, longer CPB and AXC times and a higher rate of ECMO support were observed. However, no statistically significant difference was noted.
Conclusions: The presence of coronary artery anomalies was not indicative of early postoperative mortality. In cases with a coronary artery pattern featuring a single trunk and intramural course, it may lead to postoperative ventricular dysfunction. This can complicate the surgery, resulting in longer CPB and AXC times.

Keywords: Arterial switch, transposition of the great arteries, coronary anatomy, coronary artery anomaly

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Table 1. Demographic, Preoperative, and Postoperative Data, and Diagnostic Characteristics of Patients

Variables	Group I (n=181)	Group II (n=70)	p
Gender (male)	125 (%69.1)	52 (%74.3)	0.41
Age at operation, days, median (range)	8 (1-1440)	17.5 (1-900)	**<0.05**
Weight (kg), median (range)	3.5 (2.0-13.8)	3.8 (2.3-11.0)	**<0.05**
Diagnosis			
TGA-IVS	99 (%76.2)	31 (%23.8)	
TGA-VSD	72 (%71.3)	29 (%28.7)	**0.05**
TBA	10 (%50)	10 (%50)	
Additional anomalies (CoA, AAH)	11 (%6.1)	12 (%17.1)	**<0.05**
CPB, minutes, median (range)	184 (80-466)	196 (76-423)	0.08
AXC, minutes, median (range)	103 (57-290)	116 (58-212)	0.08
ECMO	20 (%11)	13 (%18.6)	0.11
Length of ICU stay after ASO, days, median (range)	11 (3-90)	11.5 (4-70)	0.90
Length of hospital stay after ASO, days, median (range)	20 (5-120)	21.5 (8-135)	0.94
Mortality	15 (%8.3)	8 (%11.4)	0.43

TGA: transposition of the great arteries, IVS: intact ventricular septum, VSD: ventricular septal defect, TBA: Taussig Bing anomaly, CoA: coarctation, AAH: aortic arc hypoplasia, KBP: cardiopulmonary bypass, AXC: aortic cross clamp, ECMO: extracorporeal membrane oxygenation, ASO: arterial switch operation, ICU: intensive care unit

PP-561
Isolated tricuspid valve dysplasia in children: follow-up and treatment

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Background and Aim: Isolated tricuspid valve (TV) dysplasia with significant tricuspid regurgitation (TR) is a rare cardiac anomaly. In the isolated variant, TR may not manifest itself clinically for a long time. In pediatric population, only severe TR with significant clinical symptoms is considered for surgical treatment.
The purpose of the study was to analyze the natural course and treatment management in children with isolated TV dysplasia and significant TR.
Method: From 2012 to 2023, 39 pediatric patients with isolated TV dysplasia and moderate or greater regurgitation were observed in our center. Patients with displacement of TV leaflets or CCTGA were excluded from our study. In all but one case, this pathology was diagnosed postnatally. All patients were diagnosed and monitored using transthoracic echocardiography (TTE).
Results: The average follow-up period was 72.7 (6-159) months. On TTE, all patients had no coaptation of the TV leaflets due to chordal and papillary muscles thickening and shortening, thickening and reduced mobility of the leaflets. One patient had additional cleft of the anterior leaflet. In all cases there was TV dilatation and right atrial enlargement. Nine patients (23.1%) had severe TR during the initial examination. At follow-up, four patients had regurgitation reduced to moderate without treatment. Conversely, in two patients, an increase in TR from moderate to severe was noted during follow-up. Of the seven cases with severe TR 4 patients (10.3% of 39 children) had significant symptoms and were surgically treated. The mean age at the time of surgery was 74.5 months (from 38 to 152). One patient was re-operated a week after the first surgery, when a prosthetic ring was inserted in addition to the TV plasty. Postoperative results in all patients were satisfactory. All patients survived. TR was significantly reduced to a mild or moderate degree. During postoperative follow-up (mean duration 48.5 months, range 9 to 84), no patient had increased regurgitation on TV. One patient with a prosthetic ring has a mild stenosis 3 years after surgery.
Conclusions: Isolated TV dysplasia with significant TR rarely (10.3%) results in surgery among children. TV plasty has satisfactory early and mid-term results, but requires long follow-up in the future.

Keywords: isolated tricuspid valve dysplasia, tricuspid regurgitation, surgery

PP-563
Early and progressive stenosis of vascular anastomosis in congenital heart diseases infrequently associated with alagille syndrome

Pedro Agudo Montore[1], Marta Yagüe Martín[1], José Joaquín Domínguez Del Castillo[2], María De Los Ángeles Tejero Hernández[1], Elena Gómez Guzmán[1] and Carlos Merino Cejas[2]
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Background and Aim: Alagille syndrome or arteriohepatic dysplasia is an autosomal dominant multisystem condition characterised by cholestasis and bile duct paucity on liver biopsy and variable involvement of the heart, skeleton, eyes, kidneys, and face; and caused by pathogenic variants in the JAG1 or NOTCH2 gene. Congenital heart defects (most commonly involving the pulmonary arteries) are explained by disordered vasculogenesis.
Method: Retrospective chart review of clinical, genetic, echocardiographic and surgical data of patients with Alagille syndrome and congenital heart defects surgically intervened in a tertiary care children's hospital.
Results: We present 3 patients with Alagille syndrome and early and progressive stenosis of vascular anastomosis. They all have chronic cholestasis, positive genetic testing, and underwent heart surgery at the appropriate age and weight for each situation.
Patient 1. Girl with peripheral pulmonary artery stenosis, partial anomalous pulmonary venous return (scimitar syndrome), ventricular septal defect and mild coarctation of the aorta. JAG1 mutation. Heart surgery at 5 months. She developed right pulmonary veins stenosis within 2 months. She needed 2 more surgeries because of restenosis, the last one using a sutureless technique.
Patient 2. Boy with peripheral pulmonary artery stenosis and non-obstructed supracardiac total anomalous pulmonary venous return. JAG1 mutation. Heart surgery at 1.5 months. He developed pulmonary veins stenosis within 2 months. 1 more surgery because of restenosis. Death due to neurological complications.
Patient 3. Boy with transposition of the great arteries with coronary anomaly. NOTCH2 mutation. Heart surgery at 6 days of life. He developed anterior descending coronary artery obstruction and moderate supravalvular neopulmonary and neoaortic stenosis within 1 month. Death due to refractory hepatorenal syndrome.
Conclusions: Stenosis of vascular anastomosis is a rare surgical complication that occurs predominantly in the late postoperative period and in the venous territory. These cases stand out for their association with Alagille syndrome, their progressive and early onset, and the involvement of both the venous and arterial

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territory. Vasculopathy is likely to be the underlying cause and it negatively affects the prognosis of the congenital heart surgery. To our knowledge, this is also the first case reported of transposition of the great arteries associated with Alagille syndrome.

Keywords: Alagille syndrome, transposition of the great arteries, anomalous pulmonary venous return, pulmonary vein stenosis, stenosis of vascular anastomosis

PP-564

Little hearts at home a life enhancing, clinically validated remote monitoring platform

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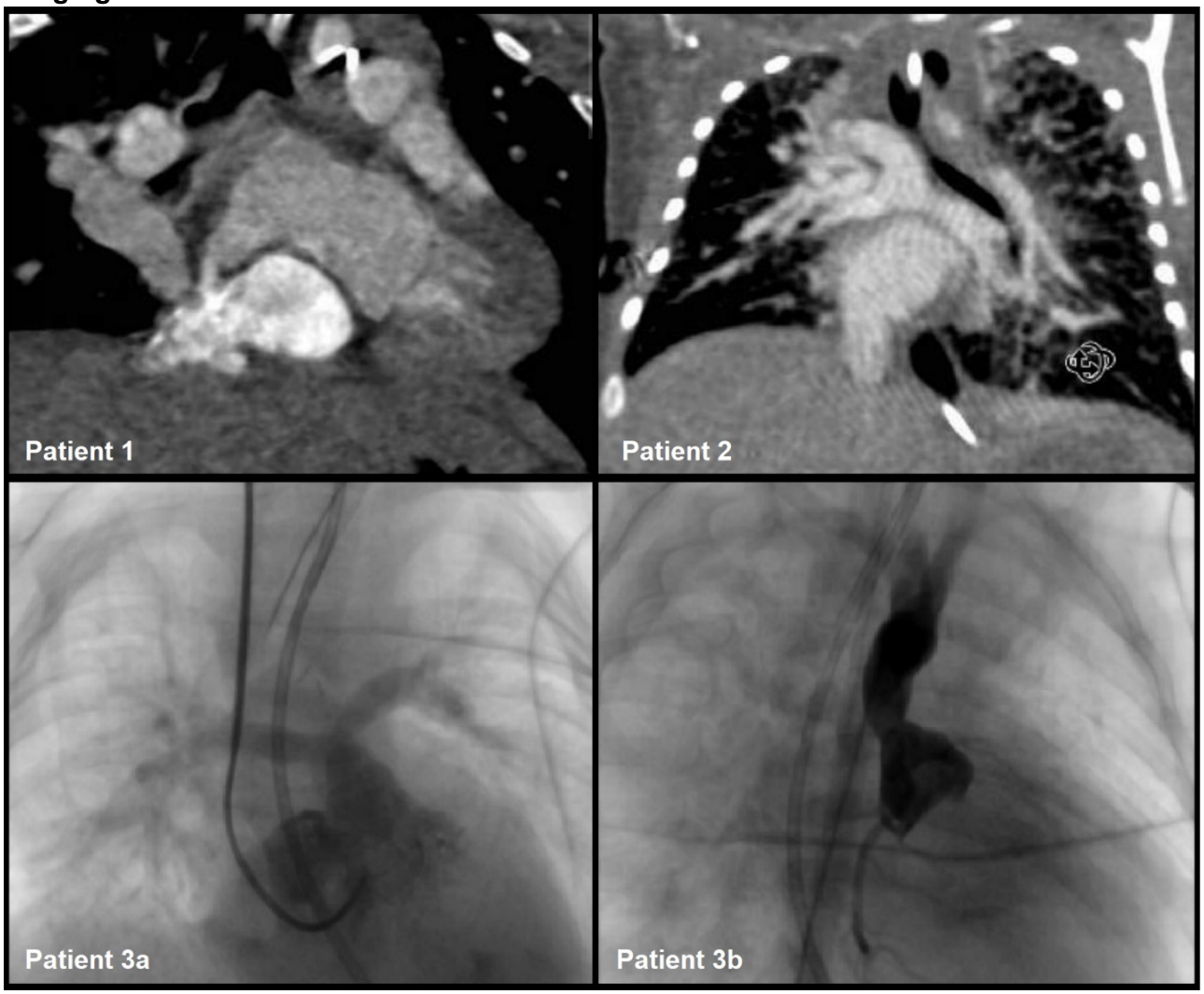
Background and Aim: Healthcare globally is looking to digitisation to help tackle quadruple priorities: improving patient experience, improving overall population health, reducing costs, and

Imaging



Patient 1. Scimitar syndrome Patient 2. Supracardiac total anomalous pulmonary venous return Patient 3. Moderate supravalvular neopulmonary and neoartic stenosis

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improving the experience for health workers. Progress is inevitably restricted by limitations in resources and funding. Therefore, Alder Hey Innovation in collaboration with Alder Hey Heart Centre have developed a clinically validated paediatric RPM (remote patient monitoring) platform, Little Hearts At Home®™. LHAH transforms existing post-operative pathways of fragile infants from a reactive approach to a proactive and preventative model, by integrating automatic alerts into domains of child health and well-being at home, specific to the heart condition. The platform aims to facilitate a safe early discharge, reduce emergency admissions, and reduce unnecessary readmissions.
Method: LHAH platform hosts information of babies born with severe heart defects, such as single ventricle anatomy, providing RPM, connecting patient, parents, community care providers, and hospital staff. We audited real-time data provided by the platform, and counter-checked with hospital visit records of these children over a 12-month period. Contemporary data is compared with historic cohorts from matched period in previous years before the platform is implemented. All patients were managed by a tertiary regional hospital (Alder Hey Heart Centre).
Results: The platform enrolled 56 high risk patients (univentricular circulation pre/post stage I/stage II Norwood, BT shunt or ductal stent/RVOT stent-dependent pulmonary circulation). Health alerts were triggered in 574/1110 entries, with the predominant reason of poor weight gain in 72% instances. Low saturations that prompted hospital admissions was encountered in 7 alerts. We avoided hospital admissions in 131 visits (95% of alerts). We also identified 11 hospital admissions not prompted by alerts, and this occurred predominantly outside working hours of community services. Compared with historic data, average length of stay from initial treatment course dropped by 25% (by 14.5 days). Reduced outpatient appointments were noted by 10.5 visits. Average emergency admissions per patients were reduced by 0.5 per patient.
Conclusions: LHAH platform demonstrates and applies the benefit of active RPM in high-risk patients with congenital heart disease. This can be scaled up to other regions and other well-defined patient population.

Keywords: Remote Monitoring, Paediatrics, Cardiology, Cardiac Surgery, Innovation

PP-565

Residual right ventricular outflow tract obstruction after pulmonary valve-sparing repair of tetralogy of fallot: An echocardiographic evaluation

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Background and Aim: Pulmonary valve sparing repair (PVSR) of Tetralogy of Fallot (TOF) provides good results in selected patients and is ideal, when feasible, to avoid long-term effects of pulmonary valve regurgitation. However, recurrent right ventricular outflow tract (RVOT) obstruction after PVSR can occur and may require surgical revision. We sought to evaluate the course of RVOT obstruction after PVSR, to identify risk factors for progression and need for reintervention by serial echocardiographic assessment.
Method: Retrospective analysis was conducted in pediatric patients who underwent PVSR of TOF at our institution, from May 1999 to May 2023. Demographic, anatomical, surgical and 2D echocardiographic data were collected. The cohort was divided into 2 groups: no reintervention group (group 1) and RVOT reintervention group (group 2).
Results: 91 patients who underwent PVSR of TOF before the age of 24 months with a minimum of 24 months follow up were included in this study; 13 patients (14%) required surgical or percutaneous reintervention. RVOT echocardiographic (ECHO) peak gradient was significantly higher in group 2 at the intraoperative transesophageal ECHO (TEE) evaluation (29.14 vs 21.71 mmHg, p 0.009), at hospital discharge (29.00 vs 22.68 mmHg, p value 0.021), at 6 months follow-up (38.25 vs 20.93 mmHg, p 0.0001) and at 12-36 months follow up (54.25 vs 19.03 mmHg, p 0.0001). A more prevalent subvalvular stenosis was found in the reintervention group at 6 months (p 0.0011) and 12-36 months follow up (p 0.00069). An RVOT ECHO peak gradient of 30 mmHg at intraoperative evaluation (p 0.025) and at discharge (p 0.011) was statistically associated with the need for reintervention. Kaplan-Meier survival curves showed that 86% of our population were free from reintervention at a medium follow up of 115 months.
Conclusions: RVOT peak gradient was significantly higher in group 2 than in group 1 at intraoperative evaluation, discharge and follow-up, with an ECHO peak gradient value of 30 mmHg being predictive of reintervention at intraoperative TEE and at discharge. At follow-up, residual RVOT obstruction was prevalent at subvalvular level in group 2. This information could be useful for intraoperative and clinical decision making in PVSR patients affected by TOF.

Keywords: Tetralogy of Fallot, Valve sparing repair, RVOT obstruction, Echocardiography

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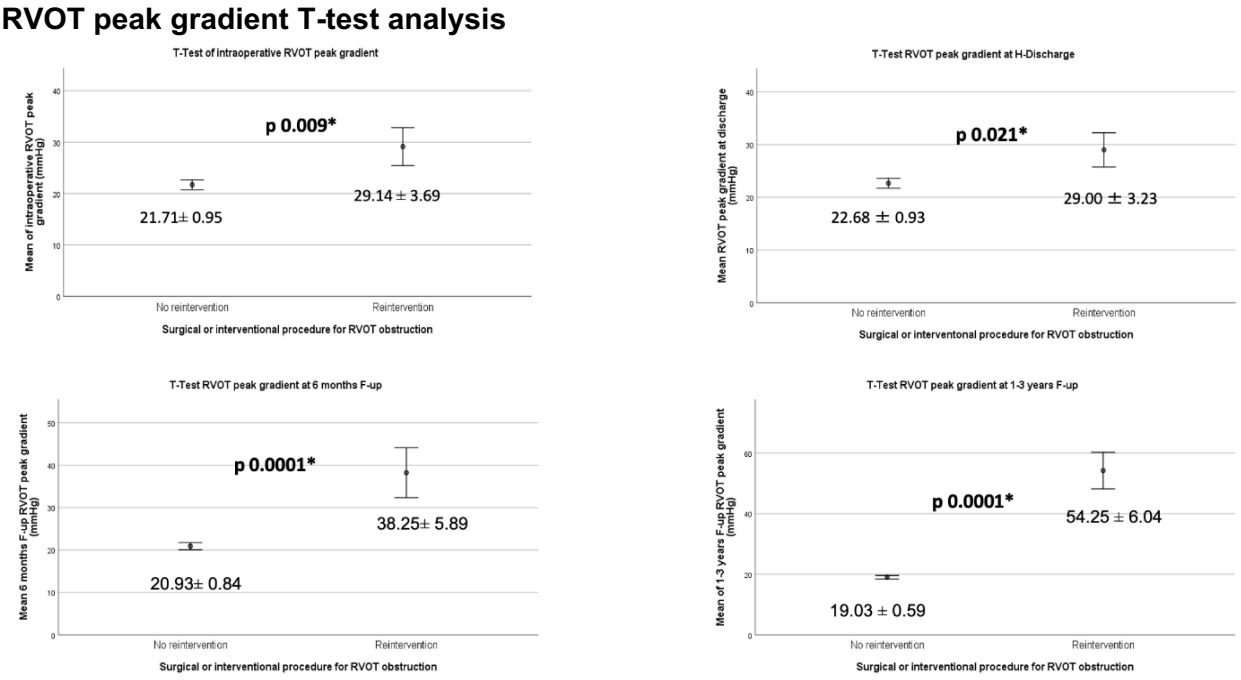

Mean RVOT peak gradient in the two groups at intraoperative evaluation, at hospital discharge, at 6 months follow-up and at 12-36 months follow-up. T-test analysis

PP-566

Surgical management of double aortic ARCH: A systematic 36-year single center study

Sara Kacem, Maroua Messaoud, Nouha Boukhrissa, Myriam Ben Fredj, Sabrine Ben Youssef, Mohamed Sami Sfar and Mongi Mekki
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Background and Aim: Double aortic arch (DAA) is a rare congenital vascular malformation in which the two aortic arches form a complete vascular ring. The trachea and/or esophagus are completely encircled. This study aims to summarize the experience of diagnosis and surgical treatment for congenital double aortic ARCH.
Method: A retrospective study of the clinical data of children treated for DAA was conducted from January 1987 to September 2023.
Results: The data of 11 patients were collected. The sex ratio was 1.75 with a median age of hospitalization in our department for surgical treatment of 11 months (range: 2 to 28 months). The median delay between the beginning of the symptoms and the diagnosis was 11.4 months. All patients had respiratory symptoms, the main symptoms were dyspnea, stridor and recurrent respiratory infections. Gastrointestinal symptoms were present in 3 cases. All patients had a chest X-ray which revealed: a pulmonary infection in all cases, disappearance of the aortic knob on the left in 2 cases. Upper Gastro intestinal opacification has shown double imprint on the esophagus in 8 cases and one simple imprint in 3 cases. CT angiography confirmed the diagnosis of DAA in 9 cases. All the patients were treated surgically. A thoracotomy approach was performed in 6 cases and a thoracoscopy was performed in 5 cases however one patient was converted preoperatively because of the alteration of the hemodynamic status related to a section suture disunion. This patient died 2 days after the operation. Outcomes after surgery was satisfactory in 7 cases, recurrence was observed in 3 cases. No late re do surgery were needed.
Conclusions: Recurrent bronchopneumonia, especially if associated with stridor, should be considered as a diagnosis of DAA. The diagnosis of DAA could be established with the use of CT-scan. The treatment is surgical. Thoracoscopy is the new surgical approach for DAA. Outcomes are excellent after surgery.

Keywords: Double aortic arch, thoracoscopy, thoracotomy

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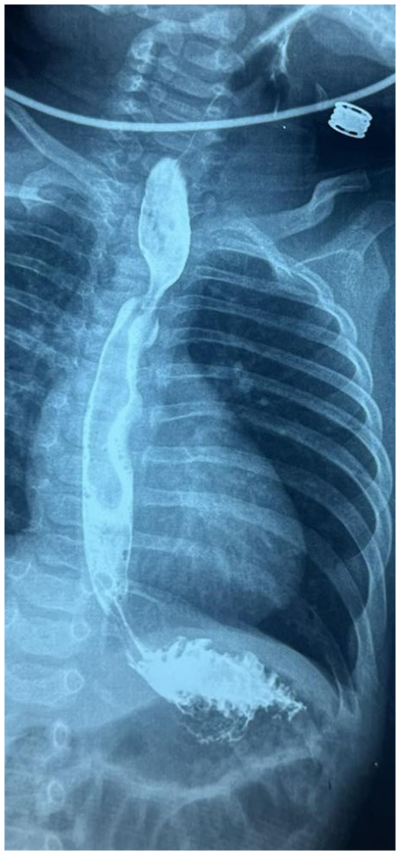
Upper Gastro intestinal opacification



An Upper Gastro intestinal opacification showing a double imprint on the esophagus meaning the presence of a double aortic arch

PP-567

Congenital extrahepatic portosystemic shunt: Diagnostic and therapeutic approach

Sara Kacem, Nouha Boukhrissa, Maroua Messaoud, Myriam Ben Fredj, Sabrine Ben Youssef and Mohamed Sami Sfar
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Background and Aim: Congenital portosystemic shunts (CPSS) are rare vascular malformations, causing changes in hepatic metabolism leading to hepatic encephalopathy and pulmonary arteriovenous shunts with a pulmonary hypertension (PH). The development of radiological exploration techniques has enabled diagnosis and facilitated therapeutic management. The purpose of this presentation is to show the clinical presentation and short-term outcomes of the surgical managment of congenital portosystemic shunts.
Method: We report the observations of 2 patients managed for congenital portosystemic shunts (CPSS).
Results: Case 1: A 3-year-old girl followed for cyanosis. On examination: periorbital cyanosis, digital hippocracticism, O2 saturation: 85%. Biology: ammoniemia: 128UI/L, prothrombin time (PT): 89%. Doppler ultrasound and abdominal angioscanner showed a complete common mesentery, CPSS between the left renal vein (LRV) and the superior mesenteric vein (SMV) with agenesis of the suprarenal vena cava, absence of hepatic portal vein (HPV). Cerebral MRI detected an arteriovenous malformation. Cardiac echocardiography with Microbubble Enhanced Echocardiography revealed an intrapulmonary shunt. The CPSS was surgically closed. Saturation stabilized at 87% postoperatively, but an ultrasound at day 5 postoperatively revealed a small thrombus at the junction of the shunt and the SMV, indicating the need for anticoagulant treatment. Case 2: A 3-year-old girl with known as followed for pulmonary narrowing with spontaneous regression. Incidental disovery of CPSS on abdominal ultrasound. On examination, no cyanosis, liver palpated on the left. Biology: normal. Investigations showed a situs inversus, a porto-caval shunt between the right renal vein and the HPV, which was permeable. Cardiac ultrasound revealed an intra-pulmonary shunt. Surgical ligation of the shunt was performed with good progression, and 2 echocardiography confirmed closure of the CPSS with a permeable HPV.
Conclusions: In the event of suspected CPSS, the diagnosis must be confirmed, the exact location of the shunt must be determined, and a search must be made for associated malformations and complications. Closure of the CPSS is essential to restore portal flow and limits the occurrence of complications.

Keywords: Congenital extrahepatic portosystemic shunt, surgery

PP-568

Is right-sided aortic ARCH challenging for surgeons to repair esophageal atresia ?

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Background and Aim: The management of neonates with esophageal atresia (EA) associated with right-sided aortic arch (RAA) is a rare anatomic finding. Many surgeons would advise for repair through left-chest access. However, RAA is generally discovered peroperatively and makes the repair of EA more challenging for surgeons.
Method: Medical records of infants with EA with and without RAA from January 2005 to October 2023 were retrospectively reviewed.
Results: A total of 347 records were studied, and 11 patients were found to have a RAA. Cardiac ultrasound before surgery showed Tetralogy of Fallot in one case, a 6 mm atrial septal defect (ASD) with repercussions on the right heart chambers in one case, and transposition of the great vessels with an ASD in another case. The right-sided position of the aorta was discovered intraoperatively, and all the 11 patients underwent surgery through a right postero-lateral thoracotomy. All, except one, underwent a termino-terminal esophageal anastomosis with closure of an esotracheal fistula. The distance between the two esophageal pouches allowed for tension-free suturing. Accidental pleural opening occurred in 3 cases. The child who could not undergo an esophageal anastomosis was the one with Tetralogy of Fallot and became unstable intraoperatively. For the other cases, only one patient developed an anastomotic stricture and underwent pneumatic esophageal dilation sessions.
Conclusions: In conclusion, the intraoperative discovery of a RAA during the repair of esophageal atresia, should not induce anxiety in the surgeon. This anatomical variant is possible and may make the dissection of the esophageal pouches and the closure of the esotracheal fistula more challenging but not impossible.

Keywords: Right-Sided Aortic Arch, Esophageal Atresia

PP-569

Congenital sternal cleft: a report on two cases of siblings

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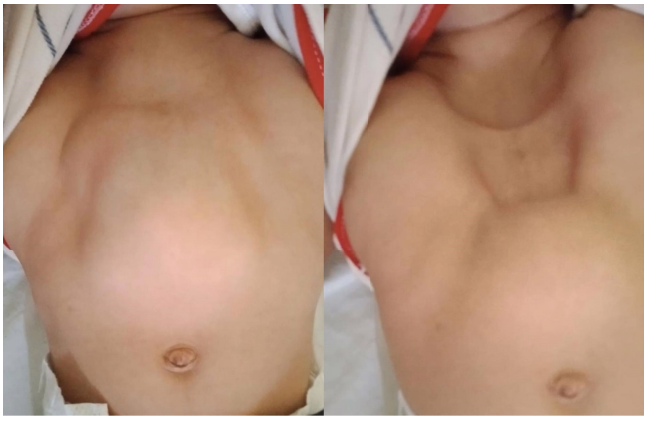
Background and Aim: A sternal cleft is a form of anterior chest wall deformity that is clinically diagnosed by observing paradoxical movement of the chest wall in newborns. Patients with sternal clefts may either be asymptomatic or experience recurrent respiratory infections. =It is a very rare condition and generally sporadic or associated with a syndrom such as PHACES syndrome or Cantrell's pentalogy. This is the first time that a congenital sternal cleft has been described in a brother and a sister, raising the hypothesis of genetic transmission to consider.
Method: We report the observations of 2 siblings followed up for a congenital sternal cleft.
Results: This concerns a 9-year-old boy, referred to our outpatient clinic along with his 11-month-old sister for the management of a complete sternal cleft. The parents were consanguineous. Both children presented with a complete sternal defect during the clinical examination, with visible heart movements under the skin: the thorax swayed with each heartbeat. Their hemodynamic and respiratory conditions were stable. Cardiac ultrasound revealed an atrial septal defect in the girl, and in the boy, there was an Amplatzer device for the closure of a persistent ductus arteriosus in place. Furthermore, the parents reported that the gap of the sternal defect decreased with age for the boy. The surgical management is currently under discussion, including the choice of the technique to be employed, among other considerations.
Conclusions: The sternal cleft is a source of thoracic instability, characterized by paradoxical movements or recurrent respiratory infections and episodes of cyanosis. Cases within a sibling pair have not been reported before. A multidisciplinary approach is necessary for management.

Keywords: Congenital sternal cleft, surgery

Sternal Cleft: visible heart movements



PP-573

Berry syndrome: first successful neonatal single stage repair in India

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Background and Aim: Aortopulmonary window comprises of only 0.1% of all congenital heart defects. Berry et al.(1982) described a rare syndrome with association of distal aortopulmonary window, aortic origin of Right Pulmonary artery (RPA), intact ventricular septum and interruption or co-arctation of aorta. Here we present the first neonatal case of single stage repair of Berry syndrome in India.
Method: A neonate, weighing 3.7kg presented on 3rd day of life with respiratory distress, persistent differential cyanosis of bilateral lower limbs and Grade 3 systolic murmur in left parasternal area. Echocardiography revealed Large aortopulmonary window (APW), RPA from ascending aorta, Interrupted aortic arch type A (IAA), large Patent ductus arteriosus (PDA) supplying descending aorta, PFO, Intact Inter ventricular septum, normal ventricular function, Severe Pulmonary Arterial hypertension. Our echo diagnosis matched completely with the defects comprising Berry syndrome. CT pulmonary angiography confirmed our echo diagnosis.
Results: After stabilization, the defect was repaired on 9th day of life as a single stage surgery in which RPA was translocated to main pulmonary artery (MPA) with augmentation of RPA anterior wall, end to side anastomosis of aortic arch was done and PDA dissected. Arch was repaired under total circulatory arrest. Total cardiopulmonary bypass time was 191 minutes. Post operatively baby was kept in ICU with open sternum for 2 days due to unstable hemodynamics. Gradually baby recovered and discharged on 10th postoperative day with Echo and CT showing well repaired arch, no residual APW, No residual PDA, Mild RPA flow turbulence, normal ventricular function.
Conclusions: Berry syndrome is a rare critical congenital heart disease. Detailed echocardiography and CT evaluation are very crucial to accurate diagnosis. Timely surgery and skilled postoperative care defines the short term and long term outcome. Single staged repair is preferred wherever feasible.

Keywords: Berry syndrome, interrupted aortic arch, single stage repair

PP-574

Giant – aneurysmatic left atrium surgically corrected in a small infant: A case report

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Background and Aim: Giant – aneurysmatic left atrium (LA) is a rare entity in children. Aneurysms can occur in any part of the LA including more frequently its appendage and can be either congenital or acquired. Younger age and lack of valvular abnormalities are supportive factors of congenital etiology.
Method: We report a case of a 4-month-old male infant with a giant LA treated in our Center.
Results: The patient presented with dyspnea and a diagnosis of bronchiolitis was made. A chest x-Ray revealed excessive megalocardia. ECG showed sinus tachycardia with P mitrale. Cardiac echo revealed a giant left atrium and an anatomically normal mitral valve (MV) with mild central regurgitation. No other cardiac or extracardiac defects were revealed. No other congenital lesions were present. The patient was started on diuretics per os. Indication for surgical intervention was the risk for thrombi formation and systemic embolization because of the blood stasis within the giant LA. Via median sternotomy and on cardiopulmonary bypass a giant left atrium was revealed (14cmX10cm) with intact pericardium. LA was opened, no thrombus was found, and an extensive resection of the abnormal atrial wall was done, so that the remaining atrial volume would be of adequate size. MV was found normal. Postoperative course was smooth, without arrhythmias or other complications, and the patient was discharged home with only aspirin for 3 months. Echocardiogram before discharge and at 3 months post surgery showed a mildly increased diameter of the left atrium with normal diastolic pressure of the left ventricle and trivial MV regurgitation.
Conclusions: Congenital aneurysms of the left atrium are the result of congenital weakness and thinning of the atrial wall due to abnormal structure. Diagnosis may be delayed as patients may be asymptomatic, or present with atypical symptoms such as shortness of breath, fatigue, feeding disorders or frequent respiratory infections. Surgical correction should be undertaken for the risk of thrombi formation and systemic embolization, as well as respiratory symptoms because of airway compression.

Keywords: Giant left atrium, aneurysmatic left atrium, congenital, surgical repair

PP-575

The subvalvular patch-enlargement is advisable in the surgical approach of tetralogy of fallot

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Background and Aim: Tetralogy of Fallot (ToF) is a complex congenital heart defect characterized by individual expressions of infundibular stenosis. Reevaluating the outcome of our patients after a 12 year period could lead to paradigm shift in our center.
Method: We retrospectively analyzed 94 patients with simple ToF in our center. Primary endpoints are re-intervention rate and mortality. Both primary repair and staged repair were included.
Results: The data show a significant revision rate in patients who initially underwent subvalvular muscle resection without patch enlargement of the right ventricular outflow tract (RVOT) (40,9%). 24,3 % of patients after transannular patch (TAP) underwent secondary conduit implantation. TAP accounts for 39.1 % of all reoperations. 60% of all re-do´s had an intact valvular anulus. Operative mortality was 2 out of 94 patients (2,1 %).
Conclusions: Primary closure of ventriculotomy after desobstruction of the RVOT lead to higher rate of reobstruction in the follow up. Therefore it is advisable to use subvalvular enlargement with consecutive patch implantation, which may avoid surgical re-do of hemodynamically relevant infundibular re-stenosis.

Keywords: congenital heart surgery, tetralogy of fallot, reoperation

PP-576

Abnormal origin of right pulmonary artery from the ascending aorta in an infant ("Hemitruncus")

Stefanos Despotopoulos[1], Sotiria Apostolopoulou[1], George Vagenakis[1], Panagiotis Zachos[1], Anastasios Chatziantoniou[1], Spyridon Rammos[1], Dimitrios Bobos[2], Meletios Kanakis[2], George Samanidis[1], Ioannis Papagiannis[1] and Alexandros Tsoutsinos[1]
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Background and Aim: Anomalous origin of right pulmonary artery from ascending aorta (AORPA) is a rare congenital heart malformation with a prevalence of 0.33% in patients with congenital heart disease.The term "hemitruncus arteriosus" is described for the same disease, but is considered inadequate, because unlike—truncus arteriosus—two arterial valves, aortic and pulmonary are present. The pathophysiology resulting from this condition affects both lungs and leads to pulmonary hypertension and severe pulmonary vascular disease. AORPA is a lethal condition and 70% of untreated patients are dead by 6months of age and 80% by 1 year.
Method: We present a 2-month-old male infant (weight: 4.1kg) referred to our hospital with failure to thrive, tachypnea, tachycardia, cyanosis, cardiomegaly and consolidation of the right upper lobe in chest X-ray, and palpable liver, with cold extremities. As the infant was intubated under inotropic support, in the transthoracic echocardiography the right pulmonary artery (RPA) could not be visualized especially from the short axis view (Figure 1A,B). The diagnosis of AORPA laterally was suspected (Figure 1C) and confirmed by CT angiocardiography. (Figure 1D,E). There was also significant right ventricle dilatation, compressimg the left ventricle in systole with an estimated right systolic pressure of 70–75mmHg. The infant underwent a median sternotomy. Severe right ventricular dilatation was prominent and the RPA originated from the lateral aspect of the ascending aorta (Figure 1F). PDA was ligated and under cardiopulmonary bypass the RPA reimplanted to the main pulmonary artery. A PFO was deliberately left due to the existing high pulmonary artery pressure.
Results: Postoperatively the RPA could be identified to the expected anatomic location (Figure 1G,H). Inhaled NO started stopped on postoperative day 11. Postoperative course was stormy and weaning process from the ventilator remained unsuccessful for several times. Finally, the infant extubated on postoperative day 25 and discharged on day 40.
Conclusions: AORPA is a rare congenital heart malformation and needs surgical management. Pathophysiologically, a large left-to-right shunt from the aorta to the pulmonary circulation is created. The contralateral lung is therefore subjected to the entire right ventricular output. This anomaly must be corrected the soon as possible because can lead to irreversible results.

Keywords: anomalous pulmonary artery, ascending aorta, congenital heart disease, hemitruncus arteriosus, pulmonary hypertension

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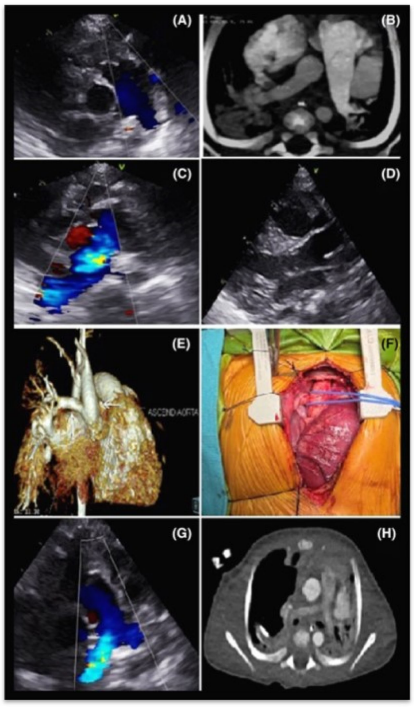
Figure 1



(A): Short axis echocardiographic view depicting the main pulmonary artery (MPA). Only the left pulmonary artery (LPA) originates from MPA. (B): Long axis view showed a vessel that arose lateral and posterior from the ascending aorta. (C): Short-axis view showed a flow in the location of the right pulmonary artery but no vessel connected to the trunk of the pulmonary artery. (D): CT angiography showed the connection ascending aorta and right pulmonary artery. (E): CT reconstruction showed the complete anatomy of malformation. (F): Intraoperative image. (G): Postoperative echocardiography showing the good flow pattern through the RPA. (H): Postoperative CT showing the good surgical result. RPA is connected to MPA.

PP-577

Fungal endocarditis in endocardial pacemaker secondary to post-surgical intermittent atrio-ventricular block. report of a case

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Background and Aim: The clinical case of a 5-year-old boy with congenital heart disease, with a diagnosis of ventricular septal defect and pulmonary subvalvular stenosis, who underwent total correction surgery and was discharged without complications, is described. Six months after surgery, he presented with syncope and cyanosis twice a week, for which he was analyzed with Holter monitoring, which concluded complete intermittent atrio-ventricular block.

A DDR mode endocardial pacemaker was placed, the patient did not experience syncope again

Method: One month after the pacemaker was placed, he presented fever with a diagnosis of tonsillitis and revived amoxicillin antibiotic treatment for a week, presenting with intermittent fever. Then, with a diagnosis of sinusitis, he received antibiotics with cefuroxime for 10 days without improvement, with a fever of 39 degrees persisting without focus, which is why he An echocardiogram was performed, revealing pacemaker key endocarditis at the level of the right atrium. He was hospitalized with intravenous antibiotics for 18 days without persistent clinical improvement in fever. The medical meeting indicates the removal of the pacemaker cables, placing a temporary epicardium pacemaker. Patient in septic shock, control echocardiogram shows vegetations at the level of the tricuspid valve, with antibiotic vancomycin, meropenen reduces fever to 38.5 degrees. Contaminated pacemaker cables cannot be cultivated. Patient continues to have a fever of 40 degrees that does not subside with paracetamol, fluconazole is started, fever persists, hyporexia. Blood cultures, stool cultures, and urine cultures were performed with negative results. culture patient on several occasions with negative cultures. Central venous catheter culture report indicates Candida glabrata, management was started with caspofungin, reducing fever with a control echocardiogram without the presence of vegetations.

Results: Caspofungin was administered for 40 days and then with oral treatment. Intermittent atrioventricular block was not evident on Holter, so the temporary pacemaker was removed.

Conclusions: currently the patient is in sinus rhythm, with functional class I/IV.

Discussion: in the literature review, fungal infection in non-immunosuppressed children is rare. We still do not know whether conservative or surgical management was ideal.

Keywords: pacemaker endocarditis. heart surgery. complete atrioventricular block, pediatric cardiology

PP-579

Challenges in the diagnosis of critical aortic coarctation: a multicenter retrospective study in the Lombardy region, Italy

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Background and Aim: Critical aortic coarctations pose a unique diagnostic challenge during prenatal ultrasound examinations due to the absence of reliable indicators. The study aimed to assess the efficacy of prenatal and postnatal screening methods for detecting critical aortic coarctations in Lombardy.

Method: A multicenter retrospective cohort study conducted between 2018 and 2021, focusing on newborns born at or beyond 34 weeks of gestational age who underwent surgical or hemodynamic procedures for critical aortic coarctation within the first 30 days of life. The study included isolated aortic coarctations and those with aortic arch hypoplasia and/or ventricular septum defects. Complex congenital heart diseases were instead excluded.

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Data from three pediatric cardiac surgery centers were collected and analyzed for demographics, diagnosis timing, perioperative factors, and post-procedural outcomes.
Results: Among the 68 neonates, 36 cases (52.9%) received a prenatal diagnosis, 22 (32.4%) were diagnosed postnatally but before hospital discharge, and 10 (14.7%) after hospital discharge. Diagnoses before hospital discharge resulted from clinical signs (63.6%), pulsoximetry and perfusion index screening tests (9.1%), and unrelated echocardiogram indications (27.3%). Conversely, 50% of diagnoses after discharge occurred in the emergency room due to acute heart failure and shock. The average age at diagnosis after discharge was 9 days. Shock rates were lower in those diagnosed earlier (9.1%) and absent in those with prenatal diagnosis. Isolated aortic coarctations were more likely to remain undetected in prenatal diagnosis (59.4%) and before hospital discharge (37.5%), compared to the more complex cases (36.1% and 2.8%, respectively). Neonates diagnosed postnatally had a higher need for preprocedural invasive ventilation (34.4% vs. 2.8%) and more frequently underwent emergency surgery (34.4% vs. 0%).
Conclusions: Diagnosis of critical aortic coarctation remains challenging, with approximately 50% of cases escaping prenatal detection, and around 14% remain undiagnosed postnatally. Clinical signs still play a pivotal role in identifying CoA not detected prenatally, especially during the first two weeks of life. Enhanced screening and diagnostic methodologies are essential to address these challenges, ultimately improving outcomes and the quality of life for affected neonates.

Keywords: Aortic Coarctation, Congenital Heart Disease, Diagnosis, Screening

PP-580

Quality assessment in pediatric cardiology using network technologies

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Background and Aim: Multicenter consortia, real-world databases, networks and voluntary registries have been established with the purpose of research in a larger scale and generalizability of findings. The Miracum (Medical Informatics in Research and Care in University Medicine) consortium has the goal to set up a nationwide infrastructure for the secondary use and sharing of health care data to improve medical care and research. With this new network technologies and data storage, we aim to implement a robust algorithm to process information on clinical outcome and quality assessment for pediatric cardiovascular patients treated in our hospital.
Method: Patients who had undergone cardiac surgery in our department in the years 2011 to 2020 were included in the analysis when aged <18 years. CHD was classified in four cardiac diagnosis groups (uncomplicated biventricular, complex biventricular, univentricular group I and II) defined by both OPS and ICD codes. We evaluated pre-operative, demographic (i.e. weight, age, presence of chromosomal abnormalities or concomitant malformations), peri-procedural (i.e. aortic cross clamp and bypass time) and post-surgical risk factors such as inflammatory and renal markers for the primary outcome mortality.
Results: A total of 1693 patients with 2146 hospitalization encounters were included. Heart disease groups comprised uncomplicated biventricular with 1254 (58.4%), complex biventricular with 603 (28.1%), univentricular stage I with 98 (4.6%) and univentricular stage II and III with 191 (8.9%) surgeries. Most significant risk factors for increased mortality were weight <2500g (HR 11.96; $p<0.001$), creatinine-ratio (HR 2.81;$p<0.001$) post-surgical leukocyte count <4000/µl (HR 3.28;$p<0.05$) and concomitant malformations as well as chromosomal abnormalities. Lower values of the ratio aortic clamp time/bypass time were
strongly associated with reduced mortality (HR 0.04, $p<0.01$).
Conclusions: Using data integration, we were able to get data assessment on a larger scale. Our analysis confirmed known non modifiable risk factors for increased mortality such as weight and chromosomal abnormalities. Interestingly, laboratory parameters such as leukocytopenia or creatinine-ratio showed a high predictive value. We understand our analysis as preparatory ground work for pooling data across different centres in a privacy–preserving manner by utilizing established infrastructure of the German Medical Informatics Initiative.

Keywords: Data integration, congenital heart surgery, quality assessment

PP-581

Preoperative assessment of left ventricular degeneration in late presenters with dextro-transposition of great arteries with intact ventricular septum

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Background and Aim: This study aims to investigate the pathogenesis of left ventricular degeneration in pediatric patients with dextro-transposition of the great arteries with intact ventricular septum (dTGA-IVS). Additionally, a comparative analysis of two treatment strategies for late-presenting cases of dTGA-IVS was conducted to assess the predictive value of preoperative echocardiography on left ventricular degradation.
Method: A retrospective analysis was performed on a cohort of 76 children with dTGA-IVS who underwent their first operation at an age greater than 28 days. The patients were divided into two groups: a two-stage aortic switch operation (ASO) group (n=38) and a one-stage ASO group (n=38). Baseline data, including left ventricular end-diastolic diameter (LVEDD), presence of patent ductus arteriosus (PDA), effective diameter of the atrial septal defect (ASD), and ventricular septal position, were collected and analyzed to compare the two groups.
Results: The study included 76 children, comprising 48 males (63.2%) and 28 females (36.8%), with a median age of 3.9 months (ranging from 31 days to 7 years). The average weight was 6.49 ±3.43 kg. Significant differences were observed between the groups in terms of intraoperative measurement of left ventricular systolic pressure/right ventricular systolic pressure (P=0.002) and effective shunt diameter of the atrial septal defect (P=0.001). Age, weight, and effective shunt diameter of the atrial septal defect at the time of surgical treatment were significantly negatively correlated with left ventricular systolic pressure/right ventricular systolic pressure.

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Conclusions: Preoperative evaluation plays a crucial role in selecting appropriate surgical strategies for children with delayed treatment for dTGA-IVS. Age, weight, and preoperative echocardiographic measurement of the effective shunt diameter of the atrial septal defect are significantly associated with the actual left ventricular functional status in children with dTGA-IVS.

Keywords: dTGA-IVS, arterial switch operation, late presenters, echocardiogram.

PP-582

The management and outcome of children with congenitally corrected transposition of the great arteries: a single center experience

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Background and Aim: To compare the outcomes in patients with congenitally corrected transposition of the great arteries (ccTGA) who were offered different surgical management strategies.
Method: Retrospectively, we included all patients in our centre from 2000 till 2020 diagnosed with ccTGA. The cohort was divided into 4 groups: (1) Patients with systemic right ventricle; (2) Patients with anatomic repair (in form of an atrial and arterial switch, or atrial switch and Rastelli operation); (3) Patients with one and a half ventricle repair (hemi-Mustard type) and bidirectional cavopulmonary connection and placement of right ventricle to pulmonary artery conduit; and (4) Patients with single ventricle who were not suitable for biventricular repair
Results: The study included 42 patients with ccTGA. In thirty-nine patients (93%) ccTGA was associated with other congenital heart defects. Group 1 (systemic right ventricle) had 16 patients (38%); seven of them underwent pulmonary artery banding (PA) with improvement in tricuspid valve regurgitation (TR) in four patients. Group 2 (anatomic group) and group 3 (one and half ventricle repair) had three patients (7%) each. Group 4 (Single ventricle palliation) consisted of 17 patients (40%); 15 of them (88%) completed total cavopulmonary connection (TCPC). Three early mortalities (7%) were recorded.
Conclusions: PA banding may improve the TR in patients with ccTGA. Patients who underwent anatomical repair (Rastelli type) or one and a half ventricle repair (hemi-Mustard type) showed good outcome with satisfactory biventricular function. Follow up of our managed patients by an individualized plan demonstrated satisfactory outcome.

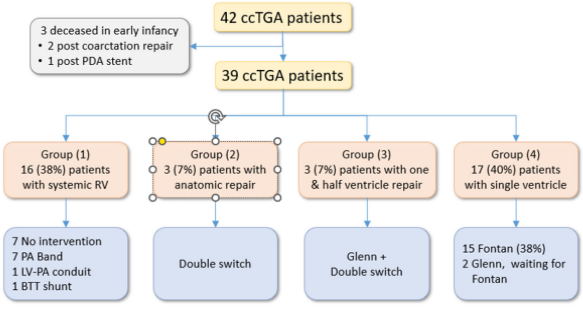

Figure 1: A flow chart demonstrating the overall management strategy of ccTGA BTT shunt: Blalock-Thomas-Taussig shunt, ccTGA: Congenitally corrected transposition of great arteries, LV: Left ventricle, PA: Pulmonary artery, PDA: Patent ductus arteriosus, RV: Right ventricle.

Keywords: Congenitally corrected transposition of great arteries (ccTGA), single ventricle palliation, pulmonary artery band.

PP-583

Factors affecting fontan-patients length of stay: Single-center study

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Background and Aim: Patients with functional single ventricle undergoing the Fontan procedure, in modification total cavopulmonary connection (TCPC) consume considerable resources. The purpose of this study is to evaluate preoperative risk factors for longer hospital stay (LOS) and to describe the perioperative course at a single institution over a 12-year period.
Method: Out of 137 patients, 134 (97.8%) were discharged, hospital mortality was 2.2% (n=3). The number of patients with the LOS for more than 30 days was 11 % (n =15). 126 (92%) patients with a follow-up period from 6 months to 12 years were evaluated, an average of 46.8±18.4 months (3.9±1.5 years). The most frequent late complication was recurrent pleural effusion (n=7, 5%). This complication caused an early readmition on 3.7 ±10.2 (from 1.0 to 6.0) months after discharge. LOS were associated with functional changes in TCPC circulation and/or nonoptimal hemodynamic characteristics.
Results: Complicated early postoperative course was observed in 63 (47%) patients. The most common complications were prolonged effusion (n=27, 20.2%), rhythm disturbances (n=19, 14.6%) and LOS (n=6, 4.6%). Lower levels of preoperative oxygen saturation, heterotaxy syndrome, right ventricular morphology, LCO requiring prolonged inotropic support (> 3 mcg/kg/min>3 days), as well as lower of total plasma protein level in the first days after TCPC (<60 g/l) were significant risk factors for prolonged effusion. The uncomplicated course of the late postoperative period was noted in 85.7% (n = 112) cases. Deaths in the long term after TCPC were not observed. The 12-year survival rate was 97.7%.
Conclusions: Preoperative hemodynamic characteristics and ventricle function was the major determinants of postoperative morbidity and mortality.

Keywords: Single ventricle, Fontan-procedure, total cavopulmonary connection, postoperative length of stay.

PP-584

Challenges of the surgical treatment of complicated melody valve endocarditis in a small national center

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Background and Aim: The Melody valve (Medtronic Inc., Minneapolis, USA) is one of the two currently available prosthetic valves for percutaneous pulmonic valve implantation. The incidence of Melody valve endocarditis (MVE) is 1.3%–9.1% per patient-year. There are no evidence-based guidelines for the management of MVE. The majority of MVEs that conservatively

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cannot be controlled undergo surgery. The surgical approach and choice of the conduit to replace infected pulmonary valve prosthesis still depends on surgeons' preference, because there are no clear recommendations regarding this issue. Another important issue the small national centers confront is the rapidly progressive disease necessitating the decision to transfer the patient to a more experienced foreign center or perform urgent surgery in their homeland. Our two case reports aim to show that challenging cases of MVE in the pediatric population may be successfully treated using homograft in a smaller national center.
Method: Patient 1 (17-year-old male) and Patient 2 (11-year-old male) underwent urgent surgery owing to the rapidly worsening MVE in the Croatian pediatric cardiac surgery center. Both patients underwent the total correction of Tetralogy of Fallot in early childhood. The Melody valve implantation was performed owing to pulmonary stenosis 13 months prior in Patient 1, and 17 months prior in Patient 2. Both patients had blood cultures positive for Staphylococcus aureus, were unsuccessfully treated with combined target antibiotic therapy and developed acute severe right heart failure. Patient 2 had a mediastinal abscess as well and was mechanically ventilated before surgery. Our heart team estimated the patients as candidates for urgent surgery that cannot be transferred to a more experienced foreign center.
Results: The Melody valve explantation and reconstruction of the RVOT using pulmonary homograft was performed in both patients. The postoperative course went uneventfully and both patients were discharged home.
Conclusions: High-risk surgical candidates with MVE may be successfully treated in smaller national centers. The scarcity of guidelines regarding surgical treatment and the shortage of homografts for RVOT reconstruction are ongoing challenges. Improvements in tissue engineering and valves of new generations might decrease the risk of endocarditis among patients with implanted pulmonary valve.

Keywords: Melody valve, endocarditis, surgery, challenges

Education/Training

PP-585
Electrocardiogram interpretation in the paediatric emergency department: Are we systematic?

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Background and Aim: Electrocardiogram is a basic and accessible test in clinical practice, since it is a cheap, quick, non-invasive and reproducible procedure. Its interpretation must be standardized and adapted to the pediatric patient. The aim of this study is to evaluate how interpretation of the electrocardiogram performed in a pediatric emergency department is done and how it is reflected in medical reports.
Method: Retrospective, descriptive, observational study including all eligible visits to the emergency department who attended for chest pain, palpitations, syncope, dizziness and seizures from 2019 to 2022. Cases in which an electrocardiogram was performed were selected. Clinical records were reviewed for demographic data, electrocardiogram description and the outcomes of the visit.
Results: 529 patients were included (266 male/263 women), with a median age of 11.2 y/o (SD 4). More than a half of the patients presented with chest pain (52.4%), syncope (24.8%) and seizures (11.2%). Most of the patients were discharged, 28 of them required admission to the hospital and 49 were referred to the outpatient cardiology clinic. 231 ECG (43.7%) were described by pediatric consultants while 298 (56.3%) were described by resident doctors. In more than 80% of medical reports it is reported: type or rythm, heart beat, QRS complex duration, PR interval duration, QTc duration and the presence (or not) of abnormalities in repolarisation. On the other hand, the presence (or not) of Q pathological waves, signs of hypertrophy, pre-excitation, signs of auricular growth and the presence (or not) of ectopic beats are reflected in less than 10% of the ECG (Table 1).
Resident doctors describe more frequently almost all items evaluated (Table 2), being these results statistically significant.
Conclusions: ECG are differently evaluated depending on who describes them. There is a statistical difference between resident doctors and consultants in almost all items. ECG description must be systematic and accounted for from the beginning of medical teaching, in order to correctly interpret the test and to benefit the medical approach of the patients.

Keywords: electrocardiogram, systematic description

	Present (%)	Absent (%)
Rhythm	96.8	3.2
Heart rate	94.3	5.7
QRS complex duration	90.2	9.8
PR interval duration	85.6	14.4
QTc	82.6	17.4
Abnormal repolarisation	81.3	18.7
QRS axis	79.8	20.2
T negative waves	30.8	69.2
Pathological Q waves	6	94
Signs of hypertrophy	6	94
Ectopic beats	5.7	94.3
Signs of pre-excitation	2.3	97.7
Signs of auricular growth	4.2	95.8

Table 1. ECG items interpretation

	Consultant (%)	Resident (%)	P value
Rhythm	40.8	56	0.000
Heart rate	38.8	55.6	0.000
QRS complex duration	37.2	52.9	0.000
PR interval duration	36.9	48.8	0.5
QTc	38.2	43.1	0.012
Abnormal repolarisation	32.3	49	0.000
QRS axis	30.4	49.3	0.000
T negative waves	10.2	20.6	0.001
Pathological Q waves	0.6	5.5	0.000
Signs of hypertrophy	3.6	2.5	0.069
Ectopic beats	4.7	0.9	0.000
Signs of pre-excitation	1.9	0.4	0.007
Signs of auricular growth	2.6	1.5	0.077

Table 2. Differences on items description between consultants and resident doctors

Table 1. ECG items interpretation Table 2. Differences on items description between consultants and resident doctors

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PP-587

Management of paediatric heart disease in low-income countries; sudan as the example: the challenges and the way forward, the role of visiting teams

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Background and Aim: Heart disease in children is a big burden on health care systems globally but it is an even bigger challenge in Low income countries (LICs) as acquired heart disease such as Rheumatic heart disease and cardiomyopathy are more prevalent in LICs
Method: We have reviewed the children cardiac cases in Sudan in 2021 that needed to have intervention locally or abroad. The finding of the review was shared with the senior paediatric cardiologists and paediatric cardiac surgeons using the Nominal Group Technique to reach a consensus about the state of the cardiac services for children.
Results: During the study period, 150 children had interventional cardiac catheterisation procedures and 104 children had paediatric cardiac surgery; The waiting list time for cardiac surgery was 14 months and 155 children had sent abroad for further management. Of those children who underwent procedures in Sudan, 72 procedures were done by visiting teams.
Conclusions: Expert group Finding: Challenges: • Economic and financial resources • Health care system challenges - Availability - Accessibility - Affordability • Training and brain drain • No reliable data The way forward • Influencing decision makers and stake holders • Innovative way of financing • Partnership model -Private sector -Non-profit organisation and International organisations • Establishing Centres of excellence and making use of visiting teams Visiting teams can help enormously in providing training for the local team and reduce the need for the patients to travel for treatment and for the medical staff to travel for training. Centres for excellence can start the services with visiting teams and this could develop to embedded team. The model of the visiting team should be a win-win situation for all the stakeholders, namely the patients, the local staff and the visiting team. Capacity building toward the ultimate goal of self-sufficient LICs programs will require a paradigm shift in the recognition by leadership, greater collaboration among stakeholders, encouragement of data sharing, and research development. In conclusion Providing paediatric cardiac services is a challenge to LICs, visiting teams can fill the gap in the short-term and help with the long-term development of the services by capacity building.

Keywords: cardiac, services, Low income countries, visiting teams

PP-588

CICU nurse shadowing programme: exploring new ways of learning in paediatric cardiac intensive care (A Pilot Study)

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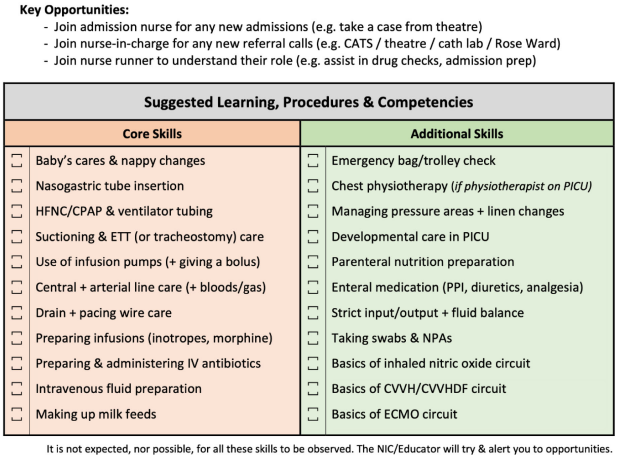
Background and Aim: Holistic and multi-disciplinary care within paediatrics & intensive care is widely valued, and the complex environment of the cardiac intensive care unit (CICU) is no exception. The role of the CICU bedside nurse is vast and crucial to a child's journey. The primary aim of this pilot study was to launch a shadowing programme involving cardiac intensive care doctors, adding a new dimension to their education, improve confidence in troubleshooting bedside problems, enhance team cohesion and eventually improve patient care.
Method: During a 6-month rotation, CICU senior house officers (SHO), registrars and consultants had an opportunity pre-allocated for a day of 'Nurse Shadowing'. A specific timetable, devised in combination with CICU Nurse Educators, allowed supervised experience in caring for patients, assisting in drug checks, and attending nursing meetings, with an optional list of skills to observe (e.g. priming an ECMO circuit). The impact of the pilot programme was analysed using anonymised questionnaires with Likert-scale questions (1 = not confident, 5 = very confident), assessing their confidence in suggested skills before and after – a mean improvement for each skill was then calculated. Supervising nurses also completed a questionnaire to evaluate their experience of the programme.
Results: 12 doctors were enrolled in the programme (5 SHOs, 6 registrars, 1 consultant). 12/12 (100%) of doctors would recommend the programme to future trainees, and 11/12 (91.6%) felt it improved their team-working and relationship with the nursing team. The three most valuable skills for trainees were preparing inotropic and intravenous infusions (+2.5), using infusion pumps (+2.3) and ventilator set-up (+1.9). Certain skills however, such CVVH and iNO set-up were less frequently observed. Thirteen CICU nurses were involved, and 13/13 (100%) agreed or strongly agreed that participation was enjoyable, improved their rapport with the doctors and that the programme would improve overall CICU patient care. There was no feedback to suggest that the programme was detrimental to either team member or patients.
Conclusions: This small-scale pilot programme has shown that nurse shadowing on a Paediatric Cardiac ICU can have promising impacts on paediatric medical education, as well as team cohesion and patient care, based on our staff's experiences.

Keywords: Paediatric cardiac intensive care, CICU, nursing, shadowing, education, teamwork

Figure 1



Key Opportunities:
- Join admission nurse for any new admissions (e.g. take a case from theatre)
- Join nurse-in-charge for any new referral calls (e.g. CATS / theatre / cath lab / Rose Ward)
- Join nurse runner to understand their role (e.g. assist in drug checks, admission prep)

Suggested Learning, Procedures & Competencies

Core Skills	Additional Skills
☐ Baby's cares & nappy changes	☐ Emergency bag/trolley check
☐ Nasogastric tube insertion	☐ Chest physiotherapy (*if physiotherapist on PICU*)
☐ HFNC/CPAP & ventilator tubing	☐ Managing pressure areas + linen changes
☐ Suctioning & ETT (or tracheostomy) care	☐ Developmental care in PICU
☐ Use of infusion pumps (+ giving a bolus)	☐ Parenteral nutrition preparation
☐ Central + arterial line care (+ bloods/gas)	☐ Enteral medication (PPI, diuretics, analgesia)
☐ Drain + pacing wire care	☐ Strict input/output + fluid balance
☐ Preparing infusions (inotropes, morphine)	☐ Taking swabs & NPAs
☐ Preparing & administering IV antibiotics	☐ Basics of inhaled nitric oxide circuit
☐ Intravenous fluid preparation	☐ Basics of CVVH/CVVHDF circuit
☐ Making up milk feeds	☐ Basics of ECMO circuit

It is not expected, nor possible, for all these skills to be observed. The NIC/Educator will try & alert you to opportunities.

An example of suggested skills and competencies for CICU doctors to observe and perform whilst shadowing

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PP-589

The pediatric aortopathy connection: A provincial patient and family networking and educational event for inherited aortopathies

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Background and Aim: Inherited aortopathies include syndromic (e.g. Marfans, Loeys-Dietz, vascular Ehlers Danlos) and nonsyndromic heritable conditions that affect the aorta with an increased risk for aortic root dilation and dissection. These conditions often require specialized care addressing medical and surgical management, emergency preparedness, family planning, genetic testing, and lifestyle. Multiple subspecialists may additionally be required in syndromic diagnoses.
Such chronic medical conditions can be isolating, especially when there are heritable implications or a risk of sudden events. Families frequently disclosed feeling isolated, wanting opportunities to meet others and learn about their condition, inspiring this project. Our goal was to create a province-wide patient and family networking and educational event for families impacted by inherited aortopathies.
Method: 1) Event organization: Our organization committee was comprised of patient/family representatives, physicians (adult and pediatric cardiology, genetics, radiology), allied health (nursing, psychology, physiotherapy), and researchers. We held meetings to determine goals and objectives, educational topics, fundraising, and event logistics.
2) Needs assessment: Participants completed pre-conference questionnaires with demographic and qualitative questions, and nine Likert scale questions (5=strongly agree) regarding medical understanding, desire to meet others, and sense of empowerment and connection.
3) Quality improvement: Attendees completed post-questionnaires of the same Likert questions, and qualitative questions for event improvement.
Results: We created the Pediatric Aortopathy Connection (PAC), a single day, province-wide patient and family education and networking event for people impacted by inherited aortopathies.
1) Needs assessment: Pre-conference responses from 30 families (N=44 adults, N=37 children) represented a breadth of conditions: Marfans (N=40), Loeys-Diez (N=28), and other (N=13). Results outlined below.
2) Content: PAC consisted of sessions for education (3D aortic models, genetics, radiology, ophthalmology), mental health (tailored by age), networking, healthy living, patient testimonials, and medical Q&A panels.
3) Quality improvement: 44 participants attended. Comparing pre- and post-conference responses there was increase in understanding (3.5+/-1.0 to 4.5+/-0.5), empowerment (3.5+/-0.8 to 4.6+/-0.2), and sense of connection (2.0+/-1.6 to 4.5 +/-0.5).
Conclusions: PAC, our inaugural province-wide patient/family networking and education event for inherited aortopathies, successfully enhanced knowledge, sense of connection, and empowerment for our families. We hope to hold this event biennially, expanding interprovincially and virtually to increase accessibility.

Keywords: Aortopathy, aorta, congenital heart disease, Marfans, pediatric cardiology

PP-590

Twinning international paediatric cardiology fellowship programs: A transformative educational experience with potential for global adoption

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Background and Aim: Over the last decade, education and training in paediatric cardiology has undergone a disruptive transformation. Trainees experience considerable stress achieving all the competencies required to become a competent paediatric cardiologist. A twin program of education between Texas Children's Hospital (TCH), Houston, Texas and Children's Health Ireland (CHI) at Crumlin Dublin was developed in 2020. It centres around co-chaired educational sessions conducted on a two monthly basis in webinar format. It is intended to; provide informative content, highlight challenges in practice and training internationally, and foster collaborative research. The aim of this study was to evaluate the utility of the program as perceived by trainees.
Method: Individuals were eligible to participate in the study if they were in fellowship or higher specialist training in paediatric cardiology in either institution and, had attended at least one session. In total 6 participants from CHI and 20 from TCH were invited to participate. A questionnaire was administered online using closed and open-ended questions related to demographic information, utility of joint sessions, practice variation and clinical uncertainty, challenges and limitations of the program, and educational experiences of fellows. Quantitative and exploratory qualitative analyses were used on the closed and open-ended questions respectively.
Results: Fourteen fellows (n=10 from TCH, n=4 from CHI) participated in the study yielding a participation rate of 54%. Mean attended sessions was 5.25 (SD 3.2). 93% found the sessions educationally useful. The highest rated sessions highlighted complex management decisions, clinical uncertainty, and practice variation between the two institutions. Free text commentary revealed that working in a field with limited randomized control trial evidence was a major source of uncertainty for fellows. Highlighted areas for improvement were allocating time for sessions (50% found it difficult to attend due to time conflicts); encouraging collaborative research between the institutions and utilisation of technology-enhanced learning solutions. 86% agreed that this model of learning could be extrapolated internationally.

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Conclusions: This pedagogical model could be replicated across multiple international paediatric cardiology units and facilitate "collaborative learning" among centres across the globe. Furthermore, this novel educational model could also be adopted by other medical specialities.

Keywords: education, twinning, online, technology-enhanced learning, global, collaboration

Fetal Cardiology

PP-592
Examining the ascending aorta-ivc relationship during fetal echocardiography to enhance prenatal diagnosis of cardiac conotruncal anomalies

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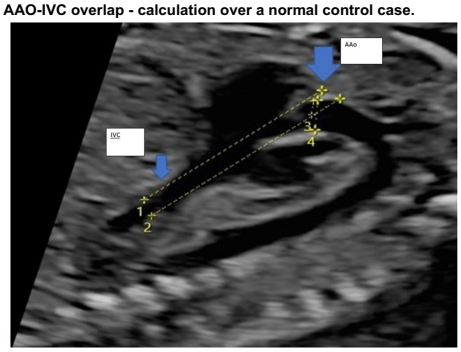
Background and Aim: The diagnosis of conotruncal cardiac anomalies (CTA) still presents challenges during fetal echocardiography. This report describes a novel methodology attempting to enhance CTA detection that examines the relationship between the Inferior Vena Cava (IVC) and the Ascending Aorta (AAo) on the routinely obtained parasagittal aortic arch view.
Method: This retrospective case-control study was carried out on pregnant women referred for fetal echocardiography for the common indications. A parasagittal view of the aortic arch which includes the upper portion of the IVC was routinely obtained and the images were archived. For the present study, we extracted this archived view and reevaluated it in 20 fetuses with CTA in comparison with 40 normal controls at a similar gestational age (GA). The calculation was performed by extending straight lines upward in continuation with the IVC walls towards the AAo upon the parasagittal aortic arch view. Once those lines were situated in between the lateral walls of the AAo at the level of the aortic root, the percentage of overlap between the inner extended IVC line to the AAo lateral wall divided by the AAo diameter, just above the aortic valve, was calculated. In case those lines were not situated between the AAo lateral walls we defined the AAo-IVC overlap as zero.
Results: The CTA group included 4 cases of transposition of great arteries (TGA), 5 cases of Tetralogy of Fallot (TOF), 9 cases of double outlet right ventricle (DORV), 1 case of malposition of the great arteries and 1 case of truncus arteriosus (TA). While in 19 out of the 20 cases in the CTA group there was no AAo-IVC overlap, all cases in the control group exhibited a pronounced degree of overlap (median 73 (IQR 60.5-87.25)). It should be noted that, in the control group, the degree of overlap exhibited a moderate negative correlation with GA ($r = -0.46$, $p = 0.003$).
Conclusions: CTA may be associated with lack of overlap of the IVC continuation and the AAo as demonstrated during fetal echocardiography when obtaining a standard parasagittal aortic arch view. This initial observation deserves further detailed evaluation.

Keywords: Fetal cardiology, Conotruncal cardiac malformations, Fetal echocardiography

Degree of AAO-IVC overlap - calculation over a normal control case.



Legend: AAo- ascending aorta IVC- inferior vena cava 3-IVC to AAo lateral wall distance 4- AAo diameter IVC-AAo overlap (percentages) 3/4x100.

PP-593
Utilizing chatgpt to facilitate referrals for fetal echocardiography

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Background and Aim: OpenAI's GPT-4, an AI, is being extensively explored as a potential decision support tool in medicine. This study evaluates its accuracy in streamlining referrals for fetal echocardiography (FE), aiming to enhance early diagnosis and outcomes for congenital heart defects, a major contributor to infant morbidity and mortality.
Method: We collected retrospective data from FE at our institution, reviewed independently by pediatric cardiologist and gynecologist (human experts (HEX), and GPT-4, following established guidelines. We assessed agreement on referral necessity between HEX and AI, with HEX reviewing disagreements.
Results: Among 59 fetal echocardiograms (61 fetuses), 50.8% were deemed normal, 27.9% minor congenital heart defects (CHD), and 18.0% major CHD by cardiologist. Recommendations for fetal echocardiography were made by a cardiologist in 47.5%, gynecologist in 49.2%, and AI in 59.0% of cases. Table 1 compares AI recommendations to actual FE results, revealing accuracies of 81.0% for cardiologist and 80.0% for gynecologist in AI versus human recommendations ($p < 0.001$). For minor CHD cases, HEX and AI recommended FE in 47.1% and 64.7%, respectively. For major CHD cases, HEX and AI referred all cases and 90.9%, respectively.
Conclusions: Assessing AI's accuracy in fetal echocardiography (FE) indications, we observed moderate agreement with medical professionals and high overall accuracy, despite a persistent 20% gap in recommendations between AI and human experts

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(HEX). Notable discrepancies stemmed from context misunderstandings, data misprocessing, and AI's lack of specialized medical knowledge, leading to ambiguity in input interpretation. While AI excels in natural language processing, it struggles with medical nuances, prompting the need for guidance from clinical guidelines. To address this, specific guidance for searching and referencing medical concepts is essential. Despite missing one major CHD case (9%), AI identified 65% of minor CHD cases compared to HEX's 47%. The study suggests AI as a valuable decision aid tool for clinicians, but recommends careful review by clinicians and anticipates improvements in AI usability and safety.

Keywords: Fetal echocardiography, Artificial intelligence, Fetal cardiology.

PP-594

Fetal 4D flow cmr improves prediction for a restricted inter-atrial septum in transposition of the great arteries

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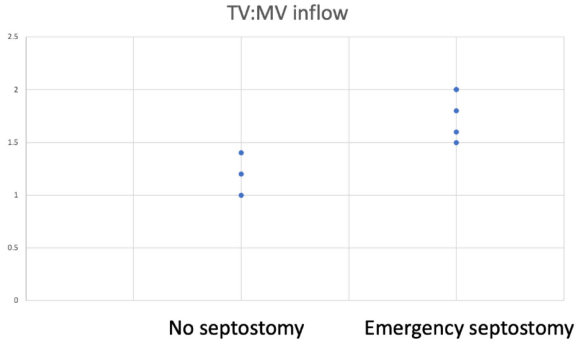
Background and Aim: Developments in the acceleration of both acquisition (compressed sensing(CS)) and post-processing, as well as in Doppler ultrasound (DUS) based fetal cardiac gating (Smartsync, Northh Medical, Hamburg, Germany), have recently improved the practical feasibility for performing 4D Flow Cardiovascular Magnetic Resonance (CMR) prenatally. Especially in later gestation, echocardiographic views can be limited and fetal CMR can add valuable information to aid perinatal care.
This pilot study aims to assess the added diagnostic value of fetal CMR in predicting a restricted intra-atrial septum (IAS) in transposition of the great arteries (TGA).
Method: 8 pregnant women were prospectively recruited from fetal cardiology clinics and underwent fetal CMR at a median gestation 35+2 weeks (range 34+2 – 36+1 weeks) on a 1.5T or 3T clinical scanner (MAGNETOM Soa and Prisma, Siemens Healthineers AG., Erlangen, Germany). DUS was used for cardiac gating. The CS 4D Flow CMR (acquired resolution 1.5mm) research sequence used for flow assessment has been extensively validated in neonates. Analysis was completed using commercially available analysis software (PIE medical imaging software, CASS, The Netherlands). Ratio of Tricuspid:Mitral valve inflow (TV:MV) was used as a surrogate for IAS flow. Direct flow assessment of the IAS was also attempted.
Results: The CMR scan was well tolerated by all 8 women. 4D Flow CMR assessment was successful in 7/8 fetuses. In the remaining one, 2D flow was used for analysis. Quantification of the direct flow across the IAC was only possible in 5/8 cases.
5/8 cases required emergency septostomy. Echocardiographic assessment only raised concerns with a restricted IAS in 2/5 cases. TV:MV inflow was >1.5:1 in all cases that needed emergency septostomy and <1.5:1 in all cases not requiring septostomy. Furthermore, the 2 cases with the most significant desaturations of <20% after delivery correlated with the highest TV:MV ratio of over 1.8:1.
Conclusions: This pilot study suggests that TV:MV inflow ratio of >1.5:1 is indicative of IAS restriction with >1.8:1 likely severe restriction.

Keywords: Transposition of the great arteries, fetal MRI, 4D flow MRI

Figure 1: Tricuspid: Mitral valve inflow as surrogate for IAS flow.



PP-595

Management and outcome of hydrops fetalis in the setting of fetal atrial flutter a 4 cases study

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Background and Aim: Fetal atrial flutter is a rare condition. It can be life-threatening as it can lead to fetal heart dysfunction, hydrops fetalis and even fetal demise. This study aims to highlight the difficulties in the management of fetal atrial flutter complicated with hydrops fetalis
Method: It's a retrospective study of all cases of documented hydrops fetalis complicating a fetal atrial flutter hospitalized in the neonatal intensive care unit of Sfax between January 2004 and November 2023.
Results: We registered 4 fetuses diagnosed prenatally with atrial flutter complicated with hydrops fetalis. A male predominance was observed with a sex ratio of 3. Prenatal diagnosis was made in all cases by fetal echocardiography. It was indicated because of hydrops fetalis in all cases. Intrauterine treatment was administered in all cases. It was based on amiodarone in one case, digoxin in two cases and digoxin associated with sotalol then relayed by flecainide in one case. After birth, all newborns showed signs of heart failure. The diagnosis was confirmed in all cases by postnatal electrocardiogram. Echocardiography was performed for all patients. It showed severe persistent pulmonary hypertension and biventricular dysfunction in one case. It was normal in the other three cases. The first line treatment was amiodarone. An external electric conversion was required in 2 cases. The mean age of conversion to sinus rhythm was 4 days [1 to 9 days]. The outcome was favorable in 3 newborns. One newborn died on day 4 of life, despite conversion to sinus rhythm after an external

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electric conversion on day one of life. He was born at 30 gestational weeks. He presented with severe hydrops fetalis, heart failure and persistent pulmonary hypertension. He required immediate mechanical ventilation, administration of vasoactive drugs and iterative ascites drainage. No recurrence of the flutter was observed in the surviving infants with a mean follow-up of 2 years.
Conclusions: Despite the possibility of intrauterine therapy, the management of fetal atrial flutter remains challenging; especially when it is complicated with hydrops featlis. The outcome depends on the prompt of diagnosis and management, which must be multidisciplinary.

Keywords: fetal, flutter, tachycardia, cardioversion

PP-596
Placental vascular malperfusion abnormalities in fetuses with congenital heart diseases
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Background and Aim: Placental Anomalies are a common occurrence during pregnancy and can significantly affect the health of the fetus. Congenital heart defects (CHD) are among the most prevalent congenital anomalies that can impact fetal cardiac development. Recent studies have suggested a potential correlation between placental anomalies and the development of CHD, highlighting the possibility of an interaction between these two conditions. Understanding this relationship could provide valuable insights for early diagnosis, monitoring, and management of CHD. The objective of our study is to assess the presence of macroscopic and microscopic placental anomalies in a cohort of pregnancies complicated by fetuses with CHD.
Method: This is a retrospective single-center study conducted from March 2021 to October 2023. Ninety-five placentas from fetuses affected by CHD were collected and analyzed. These samples were examined by a single pathologist following the criteria outlined in the Amsterdam Placental Workshop Group Consensus Statement. When the delivery occurred in other centers, the placentas were transported in order to be analyzed by the same operator. The considered pathologies include maternal vascular malperfusion (MVM), fetal vascular malperfusion (FVM), and combined maternal-fetal vascular malperfusion (MFVM).
Results: The most represented CHD pathologies are Transposition of the Great Arteries (IVS n=24 – 25.3%; Complex-type n=14, 14.7%), Tetralogy of Fallot (n= 9, 9.5%), Pulmonary Atresia with Ventricular Septal Defect (n=6, 6.3%), Pulmonary Atresia with Intact Ventricular Septum (n=11, 11.6%). Other group include Left Heart Obstruction, Cardiomyopathies. The average weight of the placentas was 537.3 ± 168.5 g. Thirty-nine placentas (41%) were Large for Gestational Age and 23 (24.2%) were Small for Gestational Age. Additionally, the mean neonatal weight/placental weight ratio was calculated as 6 ± 0.23. MVM lesions were observed in 40 placentas (42.1%), while MFVM were identified in 22 placentas (22.1%). Three samples exhibited signs of exclusive FVM.
Conclusions: More than half of the analyzed placentas showed anomalies in terms of maternal and fetal malperfusion. The absence of a control group prevents us from comparing our samples with placentas from normal pregnancies, which would provide insight into the actual impact of cardiac abnormalities on placental structure and functionality during fetal life.

Keywords: Placenta, Congenital Heart Disease, Vascular Malperfusion

PP-597
Placental lesions in pregnancies complicated by transpositions of great arteries
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Background and Aim: Placental anomalies during pregnancy can greatly affect fetal health, including congenital heart defects (CHD) like Transpositions of Great Arteries (TGA). TGA has distinct fetal pathophysiology. Recent studies indicate a possible link between placental anomalies and CHD, suggesting an interaction. Examining placental features in TGA pregnancies offers insights into the placenta-heart development relationship.
Method: We conducted a retrospective, single-center study spanning from March 2021 to October 2023. A total of 38 placentas from fetuses affected by (TGA) were collected for analysis. A single pathologist meticulously examined these samples, employing the criteria outlined in the Amsterdam Placental Workshop Group Consensus Statement. The study encompassed various pathologies, including maternal vascular malperfusion (MVM), fetal vascular malperfusion (FVM), and combined maternal-fetal vascular malperfusion (MFVM). Subsequently, the data were categorized into two groups: Intact Ventricular Septum TGA (IVS-TGA) and TGA complex-type (cTGA).
Results: We analyzed 24 placentas from pregnancies complicated by Intact IVS-TGA and 14 placentas from pregnancies complicated by cTGA. Mean neonates birth weight was 3094 ± 322 g. Mean Gestational Age at birth: 38.2. ± 0.68 weeks. The placental weight of the IVS TGA group was slightly higher, although not statistically significant, being 599 g in IVS TGA group and 592.4 g in cTGA group. Of 38 placentas, 19 (50%) were LGA, 5 (13%) were SGA. Additionally the birthweight/placenta ratio resulted to be similar in the two groups (IVS TGA: 5.68; cTGA: 5.31), corresponding to that of a fetus of 34-35 weeks of gestational age. The difference in the prevalence of maternal vascular malperfusion (MVM) in IVS TGA group was 33% (n=8), compared to 42% (n=6) in the cTGA group.
Conclusions: From the results obtained, it appears evident that in patients with TGA, both simple and complex, there is a defect in placentation.It is indicated by the finding of a reduced neonate/placenta weight ratio, corresponding to a preterm newborn. This finding, with an absolutely adequate average weight of the newborns, may indeed highlight an attempt by the placenta to compensate for a perfusion deficit by increasing its size. Moreover, it is also evident that more than 50% exhibit alterations in maternal or combined vascular malperfusion.

Keywords: Placenta, Transposition of Great Artreies, Malperfusion

PP-598

Evaluation of fetal cardiac function in gestational diabetes mellitus by two-dimensional speckle-tracking technology

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Background and Aim: Subclinical fetal cardiac dysfunction is recently described as an effect of gestacional diabetes (GD). Two-dimensional speckle-tracking technology for fetal heart evaluation is currently available. Feasibility, reproducibility and normal range values have already been reported. This study aimed to assess the effect of GD on fetal cardiac function by two-dimensional speckle-tracking technology.
Method: We performed a prospective observational study that included 89 pregnant women, 42 with GD and 47 healthy. A four-chamber 3s cine-loop was recorded and analyzed with Fetal Heart Quantification (FetalHQ® from GE®). Global longitudinal strain (GLS) for both ventricles was calculated. Demographic data shows no 3rd trimester scans or uncontroled GD. Demographic and cardiac differences between the two groups were analyzed.
Results: Gestacional age (GA) was 23 (SD 3.02) weeks. GLS of left ventricle (LV): –21.88% (SD 5.81%), right ventricle (RV) –17.16% (SD 6.89%), left atrium (LA) 22.32% (SD 8.10%). T test and linear regression analysis show statistic correlation between LV and RV GLS (beta 0.423; p-value 0.0006, Multiple R-squared: 0.13). No significant correlation was found between GD and GLS values nor between gestational age and GLS values.
Conclusions: GLS was a feasible and reproductible technique. LV and RV strain have a significant correlation as expected. No significant correlation was found between LV and LA strain. No evidence in this study of cardiac dysfunction in GD patients at 2nd trimester in a well controlled cohort. Larger studies are needed.

Keywords: Fetal, Strain, PrenatalDiagnosis

PP-599

Congenital long QT síndrome and fetal bradycardia: the importance of the diagnosis

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Background and Aim: Long QT Syndrome (LQTS) is one of the causes of cardiac sudden death in young population. The diagnosis during the fetal period is challenging because routine fetal electrocardiogram (ECG) or magnetocardiography for QT interval measurement is not readily available. The detection of fetal bradycardia has been proposed as one of the criteria for diagnosis.
Method: We present a case of a LQTS whose suspicion was the detection of sustained fetal bradycardia.
Results: A full-term newborn girl was delivered /37+3 weeks, 2250 grams) via uneventful vaginal delivery after induction due to type 1 intrauterine growth restriction detected since week 28. She had received two courses of lung maturation therapy due to threatened preterm birth. Prenatal ultrasounds revealed sustained fetal bradycardia (100-110 beats per minute) from week 28, 1:1 conduction and no hemodynamic impact which persisted throughout the entire gestation. Fetal suffering and maternal endocrine-metabolic abnormalities were ruled out (including negative anti-SSA and anti-SSB). The mother was not receiving any medications, and neither parent had a significant family history. Both parents had repeated normal ECGs (QTc intervals < 440 ms).
The newborn was admitted and monitoring, confirming sinus bradycardia without hemodynamic impact. Echocardiogram showed a structurally and functionally normal heart. Serial ECGs revealed repolarization abnormalities with prolonged QTc intervals up to 480-490 ms. A 24-hour Holter monitoring did not provide new findings. She was discharged with a home monitor. Due to suspicion of congenital LQTS, genetic testing was performed on the patient, confirming type 1 LQTS with a heterozigous pathogenic variant c.727 C>T, p.Arg243Cys in the KCNQ1 gene. After genetic confirmation, oral nadolol treatment was initiated and the monitor was removed. The mother was found to be homozygous for the same pathogenic variant in KCNQ1, exhibiting QTc intervals of up to 470 ms in subsequent ECGs and nadolol was prescribed. Genetic testing was proposed to maternal grandparents and sister, who have currently declined.
Conclusions: LQTS is often diagnosed postnatally. It's important to be aware of their prenatal clinical features in order to establish an early diagnosis and treatment, study relatives and minimize the risk of sudden cardiac death.

Keywords: Long QT syndrome, fetal bradicardya, sudden cardiac death

PP-600

The efficacy of extended fetal echocardiograms at earlier gestational age performed by pediatric cardiologists

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Background and Aim: The fetal screening should be performed between 18- and 22-weeks' gestational age (GA) according to the guidelines. However, some obstetricians prefer the screening after 22 weeks GA for several reasons in Japan. The aim of this study is to evaluate the efficacy of fetal echocardiograms by pediatric cardiologists at earlier than 19 weeks in assessment of cardiac structures.
Method: Extended fetal echocardiogram was performed in pregnant women with high risk for congenital heart disease (CHD). Cardiac structures and function were assessed by pediatric cardiologists using Voluson E8, E10 or Expert 22(GE Healthcare). The diagnosis of CHD, maternal information and pregnancy/postnatal outcomes were obtained from the medical records. The severity of CHD was classified into four categories as below: (IV) critical, who were suspected to have poor prognosis, or to require the urgent postnatal intervention, (III) complex, who would require the surgery before the discharge from NICU or Fontan candidates, (II) moderate, who would require the surgical repair in childhood, (I) mild, who would not require any surgeries but follow-up visits or normal.
Results: Fetal echocardiograms at less than 19 weeks GA were performed in 65 cases between 2015/3 and 2023/10. The median GA at the time of exam was 17 weeks. The termination of pregnancy (TOP) was chosen in 23 cases (35.4%) and one intrauterine fetal demise was occurred. The incidence of CHD was 43% and the severity of postnatal condition was prenatally predicted as (IV) in 12 cases, (III) in 11 cases, and (II) in 4 cases. Among the liveborn follow-up group including normal, the consistency of diagnosis was perfect except the case with small ventricular septal defect,

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and the Fontan candidate born without suspected heterotaxy. In cases with any major extracardiac anomaly or trisomy 21 were detected, the TOP was made even in (II) and (I) groups, otherwise most TOP were seen in (III) or (IV) groups as expected.
Conclusions: With the high detection rates of CHD by experienced pediatric cardiologists, the advanced fetal echocardiogram before 19 weeks was enough to diagnose cardiac conditions with a great agreement with postnatal outcome.

Keywords: Fetal echocardiogram, congenital heart disease, prenatal diagnosis

PP-601
Unraveling the pandemic puzzle: pediatric pots diagnoses in the COVID-19 ERA
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Background and Aim: Postural Orthostatic Tachycardia Syndrome (POTS) is a common cause of orthostatic intolerance characterized by heart rate increase upon standing without orthostatic hypotension. This study aimed to compare POTS diagnosis rates, clinical presentations, and laboratory findings in pediatric patients before and during the COVID-19 pandemic.
Method: A retrospective analysis included 1420 head-up tilt tests, with 750 conducted before and 670 during the pandemic. Data on demographic profiles, complaints, heart rate variability, hemoglobin, B12 levels, and electrocardiographic findings were assessed.
Results: Pre-pandemic POTS diagnoses accounted for 6.1% (46/750) compared to 10.5% (71/670) during the pandemic. The rate of diagnosing POTS was significantly higher during the pandemic ($p < 0.05$).Female predominance persisted (76% vs. 69%), and syncope was the commonest complaint (84% vs. 80%). Hemoglobin levels showed no significant difference, while B12 levels were higher during the pandemic (p: 0.043). Electrocardiographic findings remained inconclusive. Heart rate variability was similar between periods.
Conclusions: COVID-19 coincided with a surge in pediatric POTS diagnoses, hinting at a potential link between viral exposure and increased incidence. Despite higher B12 levels during the pandemic, COVID-19-related mechanisms might exacerbate POTS symptoms. This emphasizes the need for further exploration into viral interactions with POTS and better management strategies for affected pediatric patients

Keywords: Pediatric, Postural Orthostatic Tachycardia Syndrome (POTS), COVID-19, Viral Exposure,

PP-602
Postnatal diagnosis of congenital heart disease in Ireland 2020-2023
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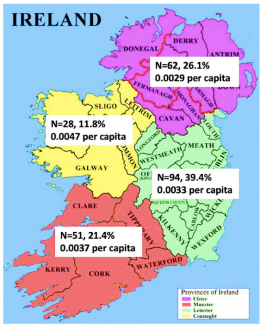
Background and Aim: This is a national review across the Island of Ireland of all patients with a postnatal diagnosis of congenital heart disease (CHD) that required surgical or catheter intervention in the first year of life. Objectives included identification of the structural lesions most likely to be missed and classification of the rate of postnatal detection by geographical region.
Method: We performed a retrospective cohort study reviewing the departmental databases to identify all patients with postnatal diagnosis of CHD on the Island of Ireland between January 2020 and July 2023 who required either surgical or cardiac catheter intervention in the first year of life.
Results: 775 patients had a surgical or catheter intervention, at less than 1 year of life, between January 2020 and July 2023. 94 of these patients underwent patent ductus arteriosus (PDA) closure and were excluded. 241/681(35.4%) of patients had a postnatal diagnosis of CHD. Of the postnatal diagnosis patients 76/241(31.5%) had Ventricular septal defects (VSD), 48/241(19.9%) Isolated Coarctation of aorta, 3/241(1.2%) Interrupted arch, 33/241(13.7%) Atrioventricular septal defects (AVSD), 24/241(9.9%) Conotruncal defects, 20/241(8.3%) Left heart obstruction, 14/241(5.8%) Anomalous pulmonary veins, 8/241(3.3%) Right heart obstruction, 7/241(2.9%) Transposition of the great arteries (TGA), 3/241(1.2%) Atrial septal defects and 5/241(2.1%) Other diagnoses. 75/241 (31.1%) of postnatal diagnoses were duct-dependent congenital heart lesions (CCHD) and of these 54/75 (72%) were critical aortic arch anomalies. Across four main regions of Ireland 39.4% of postnatal diagnoses were from Leinster (0.0033 per capita), 26.05% from Ulster (0.0028 per capita), 21.4% from Munster (0.0037 per capita) and 11.76% from Connacht (0.0047 per capita).
Conclusions: The greatest evidence for prenatal screening exists for the prenatal detection of CHD. This is particularly true of the duct dependent critical congenital heart disease group who comprised 31.1% of this cohort. The CCHD lesion most represented in our study was critical aortic arch obstruction in 72%. Despite advances in antenatal care and imaging quality some congenital heart lesions remain difficult to detect prenatally. Our regional analysis, with identification of hospital of booking, will help us identify where within the country to target funding and education to improve prenatal care and detection of congenital heart disease.

Keywords: postnatal, diagnosis, congenital heart, duct-dependent, aortic coarctation,

Postnatal Diagnosis of CHD in Ireland requiring surgery or intervention <1 year of life. January 2020 –July 2023



Congenital Heart Disease	Postnatal diagnosis patients	%
Atrial septal defect	3	1.20%
Coarctation of Aorta	48	19.90%
Interrupted Aortic Arch	3	1.20%
Ventricular septal defect	76	31.50%
Right heart Obstruction	5 + 3 (single ventricle)	3.30%
Left Heart Obstruction	15 + 5 (single ventricle)	8.30%
Anomalous pulmonary venous return	14	5.80%
Transposition of the Great Arteries	7	2.90%
Conotruncal defects	24	9.90%
Atrioventricular septal defects	30 + 3 AVSD & Coarctation of Aorta	13.70%
Other	5	2.10%
Total	241	

Image of regional CHD postnatal diagnoses across Ireland and table illustration with sub-classification of congenital heart disease that was diagnosed postnatally.

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PP-603

Biventricular remodelling and subclinical dysfunction in fetuses with tetralogy of fallot. A speckle tracking echocardiography study

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Background and Aim: Recent studies on fetal cardiovascular remodeling describe the presence of mild right ventricle (RV) hypertrophy from fetal life in tetralogy of Fallot (ToF). We aim to define the pattern of cardiovascular remodeling and function in a large cohort of fetuses with ToF by conventional 2D and speckle tracking echocardiography (STE).

Method: A multicentric prospective study (2011-2023) was conducted in two referral centers for congenital heart disease (BCNatal in Spain and university hospital of Gießen and Marburg in Germany). Sixty-tree fetuses with isolated ToF and 66 healthy fetuses were included. A 2D comprehensive echocardiography and speckle tracking analysis from 4-chamber view clips were performed at a mean gestational age of 31.6 weeks. Morphometric parameters (ventricular size and wall thickness) were evaluated by 2D echocardiography. Biventricular global longitudinal strain (GLS), tricuspid and mitral annular plane systolic excursion (TAPSE, MAPSE), RV fractional area change (FAC) and left ventricle (LV) ejection fraction (EF) were assessed by STE.

Results: Among the 63 ToF fetuses, 5 cases presented criteria for pulmonary atresia. There were no significant differences in maternal characteristics. Preeclampsia (6.35%) and fetal growth restriction (19.1%) were more frequent in the ToF group. Neonates with ToF showed lower birthweight (3075 vs 3430 grams; p=0.001) at a similar gestational age at birth (39.0 (38.0-39.6) vs 40.0 weeks (39.0-40.4); p=0.154). Compared to controls, ToF fetuses showed signs of mild biventricular concentric hypertrophy, with increased relative wall thickness in both ventricles [RV= 0.66 (0.54-0.87) vs 0.50 (0.46-0.57); p=0.001and LV= 0.68 (0.52-0.92) vs 0.51 (0.44-0.82); p=0.001]. These changes were associated with subclinical systolic dysfunction in the ToF group. RV (-17.25±3.76% vs -19.33±3.09%; p= 0.001) and LV (-17.96±3.78% vs -20.87 ±3.45%; p<0.001) GLS were significantly decreased as well as TAPSE (4.77±1.68mm vs 5.90±1.71mm; p=0.032) and MAPSE (4.09±1.79mm vs 5.45±1.60mm; p=<0.001). While RV FAC remained within normal ranges, LV EF was significantly decreased compared to controls (41.01±11.90% vs 49.44±10.30%; p=0.001).

Conclusions: Fetuses with ToF present signs of mild biventricular concentric hypertrophy with subclinical systolic dysfunction and lower myocardial deformation. Further studies are needed to correlate these findings with postnatal data to improve prognostic evaluation and management from fetal life.

Keywords: Fetal cardiology, Tetralogy of Fallot, speckle tracking echocardiography, fetal echocardiography

PP-604

Predictive factors and outcomes in patients with congenital complete atrioventricular block: two uk centre study

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Background and Aim: Congenital, complete heart block (CCHB) is the one of the most serious fetal cardiac complications of passively transferred maternal anti-Ro/anti-La antibodies. It is associated with high risk of intrauterine and perinatal mortality. Up to 70% of CCHB survivors will require permanent pacemaker implantation. Prenatal detection and management may improve the outcomes in CCHB. This study aimed to evaluate potential predictive risk factors of pacemaker placement in antibody-mediated complete heart block. The secondary purpose of this study was to determine the outcomes in patients with antenatal diagnosis of immune mediated CCHB.

Method: This was retrospective study from two UK centres (Great Ormond Street hospital, London and Royal hospital for Children, Glasgow) from January 2007 to January 2023. Neonates with documented anti Ro/La maternal antibodies who were diagnosed with heart block prenatally or within 30 days of life were included. Cohort subjects had normal cardiac structure or simple cardiac structural lesions (atrial and ventricular septal defect, patent ductus arteriosus). Observational data were collected from electronical patients records and medical databases.

Results: Forty-nine cases were identified with immune mediated CCHB. Prenatal diagnosis of CCHB occurred in 46/49 (93.9%) at mean gestational age (GA) of 25 ± 4 days. In ten cases, there was known history of maternal anti Ro/La antibodies. Regarding perinatal outcomes, there were 43 livebirths with mean GA at delivery 36 ± 4 days (30% were born <37 weeks), five intrauterine demise, five neonatal deaths and one termination of pregnancy. Maternal therapy with dexamethasone was given in 16 cases in presence of fetal hydrops, reduced ventricular function or endomyocardial fibroelastosis. While three neonates received postnatal beta-agonist infusion at birth, no patients required temporary pacing. Twenty-five patients (51%) required permanent pacemaker implantation; 12/25 patients (48%) required a neonatal pacemaker (≤30 days of life) at a mean age of 5±2 days. Mean heart

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rate (HR) during third trimester of 44 and 53 was significantly related with perinatal mortality and early pacing respectively.
Conclusions: In this study late fetal HR was the most significant predictor for postnatal outcome. This finding can be useful in prenatal counselling and delivery planning.

Keywords: Congenital complete heart block, prenatal diagnosis, outcomes

PP-605

Assisted reproductive techniques and severity of associated congenital heart disease: experience of a single center

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Background and Aim: There is no consensus in current practice guidelines on whether conception by assisted reproductive techniques (ART) is an indication for performing a fetal echocardiogram. Studies showed a slightly higher risk of congenital heart disease (CHD) in ART as compared with spontaneous conception. Little is know about the severity of CHD associated to ART. We aimed to compare type of cardiac diseases according to the modality of conception.
Method: We evaluated our population of fetus with CHD diagnosis referred to our insitution between 2018-2023. The fetuses were divided in two groups: GROUP 1 with major cardiac anomaly (TOF, DORV, HLHS, CAV, TGA) and GROUP 2 with minor cardiac anomalies (Coarctation, DIV, right ventricle predominance, double arch, right arch). We analyzed for each group the different modality of conception (spontaneous, FIVET, ICSI, ovodonation, IUI). A T student test was used to compare these 2 groups ($p<0.05$)
Results: The population was of 287 fetus with cardiac involvement; 10 were excluded from the analysis as the diagnosis was an arrhythmia and 20 without available details about the conception.Group 1 was of 140 fetuses: 133 spontaneous and 7 assisted. Group 2 was of 117 fetus: 110 spontaneous and 6 assisted. No statistical differences was found ($p= 0.22$)
Conclusions: We did not notice a significant different in severity of CHD according to the type of the conception. We are aware of the need of a bigger multicentric study to confirm our conclusions in a more representative population.

Keywords: assisted reproductive techniques, congenital cardiac risk

PP-606

Incomplete prenatal diagnosis of complex congenital heart disease

Piedad Sánchez Gil Romero[1], Ángela Uceda Galiano[1], Viviana Arreo Del Val[1], Lucía Deiros Bronte[1], Marta Inmaculada Gambra Arzoz[1], Montserrat Bret Zurita[2], Roberto Rodríguez González[3], Paloma López Ortego[4], Raúl Sánchez Pérez[5], Alejandro Martín Asenal Schaffi[1], Luis García Guereta Silva[1] and Federico Gutiérrez Larraya Aguado[1]
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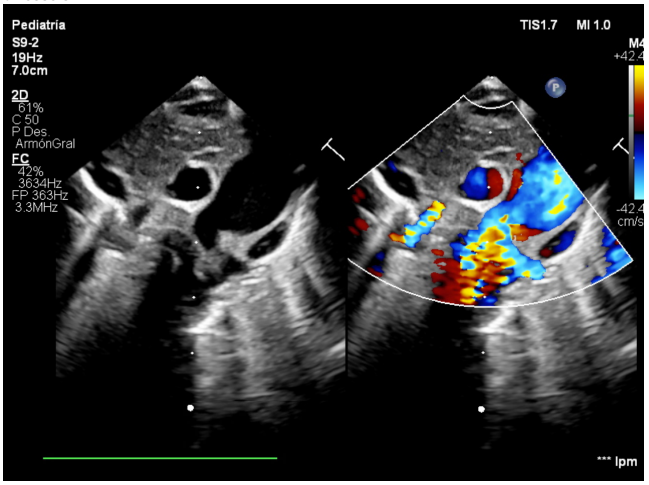
Background and Aim: Anomalous origin of the pulmonary arteries is a rare but serious congenital heart disease that requires early diagnosis for proper treatment to avoid the development of heart failure and early pulmonary hypertension.
The three-vessel tracheal plane is highly useful in fetal echocardiography to diagnose anomalies in the disposition of the great vessels, although its prenatal diagnosis can sometimes be challenging, especially if they have a trajectory that is anterior to the trachea.
Method: We present the case of a 25-year-old primigest mother with no relevant medical history, diagnosed with a complex fetal heart condition at 20 weeks of gestation. She presented to our center at 36 weeks and prenatal ultrasound revealed Shone's complex with mitro-aortic hypoplasia and coarctation of the aorta with persistent left vena cava draining into the coronary sinus.
Results: At birth, an echocardiogram and thoracic CT scan were performed, confirming the prenatal findings. Additionally, an anomalous origin of the right pulmonary branch was observed, arising from a right ductus originating in the right brachiocephalic trunk, and a left pulmonary branch originating in the pulmonary trunk. Furthermore, these complementary tests revealed an anomalous pulmonary venous drainage from the right lung (scimitar syndrome) with a single pulmonary vein draining into the inferior vena cava. As extracardiac findings, right lung hypoplasia, complex lumbosacral abnormality and anal atresia were diagnosed, prompting a genetic study.
Although the family was informed prenatally of the complexity of the heart disease, the findings detected in the postnatal study limited the treatment options for our patient, leading to a joint decision with the family on the adequacy of the therapeutic effort.
Conclusions: The right pulmonary branch may have an anomalous origin from the ascending aorta, from a right ductus or from systemic-pulmonary collaterals. Its association with other malformations is common; however, we have not found in the literature any case associating Shone's complex, scimitar syndrome and anomaly of pulmonary branch origin.
Intrauterine visualization of pulmonary venous drainage anomalies and the origin of pulmonary branches is challenging, and the diagnosis may sometimes limit the surgical correction of patients, thereby influencing their prognosis.

Keywords: Congenital heart disease, pulmonary arteries, Shone's complex, scymitar syndrome, fetal echocardiography

6 vessels



In this image, from right to left, we can see in a cross section: the persistent left superior vena cava, the pulmonary trunk with the left pulmonary branch with proximal course to the right, the ascending aorta, the left pulmonary branch and the right superior vena cava.

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PP-607

Ultrasound imaging of lymphatic inflow to fetal venous angles

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Background and Aim: In ultrasound examination lymph flowing from thoracic duct (TD) to venous angles (VA) is visible as hyperechogenic spontaneous contrast due to high protein content of lymph. It can also be seen on fetal ultrasound. Information about site of termination of TD is important, because of risk of lymphatic flow impairment following venous thrombosis after central vein cannulation. Lymphatic flow disorders can also complicate clinical course in patients with univentricular physiology or patients with genetic anomalies such as RASopathies, Turner syndrome and many others. The study aimed to assess the site of lymphatic drainage in fetuses during fetal echocardiographic scans.
Method: During several fetal echocardiography investigations lymphatic inflow to venous angles was visualized and recorded. Then the findings were retrospectively assessed and analyzed. The following data was selected: lymphatic inflow to VA, fetus gestational age, congenital heart disease (CHD) of fetus.
Results: Between 2020 and 2023 lymphatic inflow to venous angles was recorded in 57 exams of fetuses. Average fetus gestational age was 26 weeks (range 17-37 weeks). From this group of patients: 38 presented with no defects, 16 presented with CHD, 3 fetuses had arrhythmia (premature atrial complexes). Among CHD the most common were right aortic arch (3) and aberrant right subclavian artery (3). In all 57 fetuses the lymphatic inflow was seen to left VA in 29 (51%), right VA 13 (23%), both VAs in 15 (26%). In 38 fetuses with no defects the lymphatic inflow was seen to left VA in 21 (57%), right VA 6 (16%), both VAs in 10 (27%). In 16 fetuses with CHD the lymphatic inflow was seen to left VA in 5 (31%), right VA 7 (44%), both VAs in 4 (25%). In one fetus in whom lymphatic drainage to both venous angles was seen there has been alternating flow to right and left sides.
Conclusions: Examination during fetal period can provide valuable information about lymphatic inflow to VAs corresponding to TD drainage site. This can help to minimalize risk of complication of central vein cannulation in newborns and contribute to better understanding of lymphatic flow disorders in patients with CHD.

Keywords: fetal ultrasound, lymphatic flow disorder, lymphatic ultrasound imaging

PP-608

A rare fetal cardiac anomaly: Antenatal diagnosis of aortopulmonary (A-P) window

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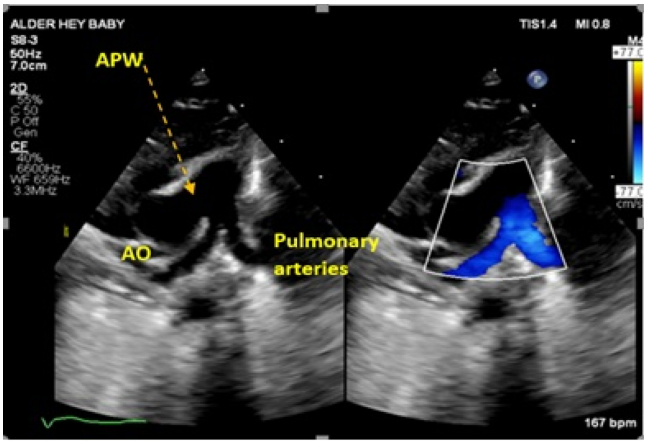
Background and Aim: Aortopulmonary window (A-P) is a rare congenital cardiac anomaly that occurs between 0.1% and 0.6% of congenital cardiac defects. It is even rarer to diagnose it on antenatal fetal echocardiogram. It is often diagnosed in early infancy with continuous murmur and congestive heart failure. A-P window can be isolated or in about one third cases, associated with intracardiac-defects. Embryologically this is due to failure of closure of cono-truncal ridge by neural crest cells from dorsal aortic sac between fourth and sixth aortic arches. A careful sweep between 3 vessel view (3VV) and 3 vessel tracheal view (3VT) can pick-up this defect which allows antenatal counselling for early surgery.
Method: A 32-year-old gravida 5 and Para 4, previous healthy normal deliveries, was referred to our specialist fetal cardiology unit for suspected cardiac anomaly after her 20 weeks anomaly scan. She had low risk 1st trimester screening. Specialist fetal echocardiogram at 21-week gestation, showed balanced ventricles with good function, a small 2-3mm perimembranous ventricular septal defect, two separate 4mm semilunar valves with normal doppler flow. As we panned cranially from 3VV to 3VT, there was a moderate 3-4mm A-P window was picked out between main pulmonary artery (MPA) and ascending aorta (AA). There were normal confluent branch pulmonary arteries and the ductus arteriosus was smaller. Patient was counselled against heart failure management, surgical closure and risk of pulmonary hypertension. She declined genetic amniocentesis.
Results: Baby was born at term with 3.2kg birth weight by normal vaginal delivery. Tertiary cardiology echocardiogram confirmed a large 6-7mm A-P window between MPA and AA. There were two separate semilunar valves and confluent pulmonary arteries. There was normal left sided aortic arch identified. Parents were counselled for surgery and baby was listed for open heart surgery between second and third month of life.
Conclusions: Antenatal diagnosis of A-P window is very rare, this can be picked up by performing a careful sweep to visualise A-P septum between 3VV and 3VT view. Early diagnosis allows antenatal counselling and reduces post-natal delay in diagnosis and complications like congestive heart failure and pulmonary hypertension.

Keywords: Fetal echocardiogram, aortopulmonary window, cardiac anomaly

Fetal and postnatal echocardiogram images 1-4



1. Fetal echo 3VV to cranial sweep showing A-P window (Top right) 2. Fetal echo 3VV to 3VT sweep showing A-P window (Top left) 3. Postnatal echo showing separate semilunar valves (Bottom right) 4. Postnatal echo showing A-P window and confluent branch pulmonary arteries (Bottom left)

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PP-609

A rare fetal cardiac anomaly: Antenatal diagnosis of aortopulmonary (A-P) window

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Background and Aim: Aortopulmonary window (A-P) is a rare congenital cardiac anomaly that occurs between 0.1% and 0.6% of congenital cardiac defects. It is even rarer to diagnose it on antenatal fetal echocardiogram. It is often diagnosed in early infancy with continuous murmur and congestive heart failure. A-P window can be isolated or in about one third cases, associated with intracardiac-defects. Embryologically this is due to failure of closure of cono-truncal ridge by neural crest cells from dorsal aortic sac between fourth and sixth aortic arches. A careful sweep between 3 vessel view (3VV) and 3 vessel tracheal view (3VT) can pick-up this defect which allows antenatal counselling for early surgery.
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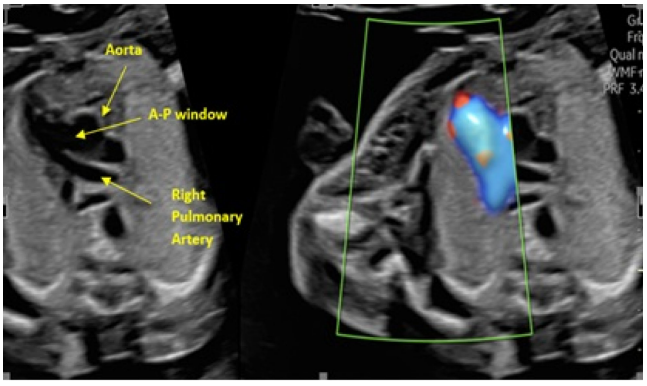
Conclusions: Antenatal diagnosis of A-P window is very rare, this can be picked up by performing a careful sweep to visualise A-P septum between 3VV and 3VT view. Early diagnosis allows antenatal counselling and reduces post-natal delay in diagnosis and complications like congestive heart failure and pulmonary hypertension.

Keywords: Fetal echocardiogram, aortopulmonary window, cardiac anomaly

Fetal echocardiogram image



1. Fetal echo 3VV to cranial sweep showing A-P window

PP-610

The evolving genetic aetiology of cardiac outflow tract anomalies

Ronel Talker[1], Adalina Sacco[2], Lyndall Sarkies[3], Tazeen Ashraf[1], Pranav Pandya[2], Victoria Jowett[1] and Sara Hillman[2]
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Background and Aim: Conotruncal (cardiac outflow tract) anomalies represent approximately 20% of all antenatally-detected cardiac defects, yet their association with genetic abnormalities varies significantly according to the specific cardiac malformation. This study assessed the diagnostic yield of antenatal and postnatal genetic testing in antenatally-detected conotruncal defects since the introduction of microarray and exome sequencing (ES).
Method: This retrospective study included all cases of conotruncal anomalies detected antenatally between 1 January 2018 and 31 December 2021. Cardiac diagnoses were made using cardiologist-performed fetal echocardiography and all patients were offered QF-PCR and microarray as prenatal invasive genetic testing. As part of a research program after 1 January 2021, if negative then additional targeted exome sequencing (ES) was also offered. Postnatally, some patients were offered genetic testing on a case-by-case basis.
Results: There were 301 cases of conotruncal anomalies included during the 4-year study period. Genetic testing was performed in 192 cases (63.8%), of which 66 were only performed postnatally. Clinically significant genetic abnormalities were detected in 53/180 cases (27.6%), most commonly 22q11.2 microdeletion (10.4%), followed by Trisomy 21 (3.1%). Trisomy 13/18 was found in 11/192 (5.7%). Amongst the cardiac diagnoses, Tetralogy of Fallot (ToF) had a pathogenic finding in 25.3% (25/99) and a similar incidence whether left or right aortic arch. Transposition of the great arteries (TGA) had a positive result in 5% (1/20). Double outlet right ventricle (DORV) had a positive result in 28.9% (13/45, with 5 cases of Trisomy 13). 66.6% (4/6) of interrupted aortic arch (IAA) cases had a positive result (all 22q11.2 microdeletion) (Table 1). In 249 cases of isolated conotruncal anomalies, 5 patients underwent ES - 4 were negative and 1 had a variant of unknown significance. In 52 of cases

Table 1
Genetic testing and diagnoses according to specific cardiac malformation

Cardiac Diagnosis	Cases	Genetic testing	Genetic abnormality	Most common abnormality
Tetralogy of Fallot (ToF) - Left aortic arch - Right aortic arch - Unknown	**124** 71 30 23	**99/124 (79.8%)** 55/71 (77.5%) 28/30 (93.3%) -	**25/99 (25.3%)** 14/55 (25.5%) 8/28 (28.6%) -	 22q11.2 del 13/99 (13.1%)
Transposition of the great arteries	71	20/71 (28.2%)	1/20 (5%)	Trisomy 18 13/99 (13.1%)
Double outlet right ventricle - Fallot-type - TGA-type - Unknown	**66** 22 12 32	**45/66 (68.2%)** 16/22 (72.7%) 6/12 (50%) -	**13/45 (28.9%)** 4/16 (25%) 0/6 (0%) -	 Trisomy 13 5/45 (11.1%)
Pulmonary atresia with VSD	17	11/17 (64.7%)	3/11 (27.3%)	22q11.2 del (1) 47, +18 (1) 46/47+2 mosaic (1)
Common arterial trunk (CAT)	16	11/16 (68.8%)	1/11 (9.1%)	8q23.3 deletion
Interrupted aortic arch (IAA)	7	6/7 (85.7%)	4/6 (66.7%)	22q11.2 del 4/6 (66.7%)
Total	**301**	**192 (63.8%)**	**53 (27.6%)**	-

Genetic testing and diagnoses according to specific cardiac malformation

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of non-isolated conotruncal anomalies with 43 of these receiving genetic testing, 11 underwent ES. Five had a clinically-significant result, increasing the yield of clinically-significant genetic diagnoses from 32.6% (14/43) to 44.2% (19/43) these patients with ES. *Conclusions:* Genetic abnormalities are present in approximately one-quarter of antenatally-detected conotruncal anomalies, particularly in IAA, ToF and DORV. Targeted exome sequencing leads to a significant increase in genetic diagnosis in non-isolated cases.

Keywords: Conotruncal, outflow tract, fetal, genetics, microarray, exome sequencing

PP-611

Two-dimensional speckle tracking echocardiography used to measure global longitudinal strain and strain rate in fetuses with congenital heart disease

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Background and Aim: Two-dimensional speckle tracking echocardiography (2D-STE) is a novel echocardiographic technique, that can provide information on fetal heart function. Global longitudinal strain (GLS) and global longitudinal strain rate (GLSR) are speckle tracking features that provide information on the longitudinal deformity of the fetal cardiac wall. Research on speckle tracking has been performed on healthy fetuses, fetuses with twin-to-twin transfusion syndrome, fetuses with growth restriction and fetuses with other fetal and/or maternal conditions, like hypertensive pregnancy disorders, but research on fetuses with congenital heart disease (CHD) remains limited. The aim of this review is to investigate and provide an overview of the literature available on 2D-STE (GLS/GLSR) in fetuses with CHD.
Method: A systematic literature search was conducted on EMBASE, Medline ALL Ovid, Web of Science Core Collection and Cochrane Central Register of Controlled Trials. Articles with data on the left ventricular (LV) and right ventricular (RV) GLS and/or GLSR in fetuses with CHD were included. GLS and/or GLSR of fetuses with CHD was compared to the strain values of healthy fetuses.
Results: Out of 388 articles, a total of 31 studies were included in this systematic review. The studies varied in multiple study characteristics, including gestational age of the fetus, study type, size of the study population, fetal heart rate, frame rate, and echocardiographic and/or speckle tracking software. Moreover, different CHD types were investigated. Some studies reported a significant decrease in GLS and/or GLSR in fetuses with CHD compared to healthy controls, while other studies reported no difference in these strain values. A meta-analysis could be performed on most of the subgroups (based on CHD type). Overall, considerable heterogeneity was seen due to differences in study design.
Conclusions: Due to heterogeneity between the studies included, no overall conclusion could be drawn from the primary outcomes. Several factors might have had an influence on the heterogeneity reported, amongst which differences in study design, CHD, gestational age, frame rate, echocardiographic devices and speckle tracking software. A prospective longitudinal study on LV and RV GLS and GLSR in fetuses with CHD is needed to provide detailed information on the function of the structural abnormal fetal heart.

Keywords: Prenatal ultrasonography, congenital heart disease, global longitudinal strain, systematic review

PP-613

Fetal bradycardia and long Qt syndrome

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Background and Aim: Fetal bradycardia may be defined as a ventricular rate more than two standard deviations below the gestation specific mean. Important causes include complete heart block, 2:1 AV block (2:1AVB), multiple blocked atrial ectopics and sinus bradycardia. Sinus bradycardia and/or 2:1AVB are recognised to be a potential presentation of fetal long-QT syndrome (LQTS). Measurement of normalised left ventricular isovolumetric relaxation time (N-LVIRT) has been proposed as a helpful measurement as a surrogate for the QT interval in fetal life. This study aimed to describe the associations of fetuses presenting with either sinus bradycardia or 2:1AVB.
Method: Retrospective review of local fetal cardiology database between 01/01/2018 to 05/09/2023 for cases presenting with bradycardia (defined as z-score >2SD below the mean for gestational age (GA)). Fetuses with anti-Ro/La antibodies, major congenital heart disease and ventricular tachycardia were excluded. Data were collected on patient demographics, fetal echocardiography findings, clinical genetics investigations, family screening and fetal outcome.
Results: Eighteen fetuses fulfilled the search criteria. Gestational age at presentation was 24.4(21.7-29.1) [Median(IQR)] weeks. Presenting rhythm was sinus bradycardia in 15, 2:1AVB in two and both sinus bradycardia and 2:1AVB in one. Genetic testing was performed in 11, antenatal in nine. Eight had a positive LQTS genotype (six KCNQ1, one KCNE1, and one CALM2). Diagnosis in the fetus led to a new diagnosis in a family member in six cases. Presenting fetal heart rate (FHR) in sinus rhythm/bradycardia z-score was -3.06 (-3.80 to -2.71) [Median(IQR)]. Across all clinical reviews FHR z-score was -2.98 (-3.32 to -2.60) [Median(IQR)]. The N-LVIRT was calculated in 13 cases (35 total measurements) and was abnormal for 2/35(3%) measurements in two different patients. One patient had a subsequent diagnosis of LQTS.
Conclusions: Sinus bradycardia is an important presentation of fetal LQTS. Identification in fetal life is significant because it can have implications for the fetus and family members who are unaware of their diagnosis. Echocardiogaphy parameters in our initial cohort did not correlate with genetically confirmed LQTS. Further work is needed to look at the role of echocardiography in diagnosis of this patient population.

Keywords: fetal, bradycardia, long QT syndrome

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Table: Presenting rhythm and echocardiography parameters for individual cases in the series.

Case	Gestation at presentation	Presenting rhythm	Fetal heart rate in sinus rhythm (presentation)	Fetal heart rate z-score in sinus rhythm Median (range) [presentation]	Number of reviews by fetal cardiology team	Fetal heart rate z-score in sinus rhythm Median (range) [across all clinical reviews]	N-LVIRT (%) [range across reviews]
1	23	2:1AVB	Not sinus	NA	5	Not sinus	
2	27	Sinus brady	114-126	-3.00 (-3.70 to -2.26)	1	-2.9785 (-3.70 to -2.26)	11.0
3	29	Sinus brady	115-150	-1.33 (-3.36 to 0.69)	2	-2.1735 (-3.36 to 0.69)	10.0-14.5
4	14	Sinus brady	136	-2.24	5	-2.983 (-4.54 to -1.79)	
5	22	Sinus brady	120-128	-2.94 (-3.45 to -2.42)	4	-3.457 (-3.85 to -2.42)	10.0-10.2
6	30	Sinus brady	93-106	-4.98 (-5.71 to -4.24)	5	-4.241 (-5.71 to -2.30)	
7	21	Sinus brady and 2:1AVB	127	-2.73	4	-3.3215 (-4.19 to -2.73)	11.6-14.8
8	27	Sinus brady	119-120	-3.04 (-3.10 to -3.00)	3	-2.523 (-3.10 to -1.71)	11.0-12.3
9	21	Sinus brady	127-139	-1.87 (-2.66 to -1.08)	3	-1.753 (-2.65 to -1.08)	9.6
10	33	Sinus brady	108-127	-2.69 (-3.72 to -1.66)	3	-2.82 (-3.72 to 0.48)	10.0
11	18	Sinus brady	122-124	-3.65 (-3.51 to -3.80)	5	-3.2695 (-3.79 to -2.11)	6.0-12.1
12	29	Sinus brady	104-107	-4.44 (-4.27 to -4.62)	3	-4.616 (-4.89 to -4.27)	7.8-13.5
13	29	Sinus brady	112-122	-3.09 (-2.51 to -3.66)	3	-2.6005 (-3.66 to -1.70)	11.4
14	21	Sinus brady	120	-3.66	6	-2.944 (-3.69 to -2.08)	8.1-10.8
15	34	Sinus brady	100-110	-3.94 (-3.40 to -4.47)	2	-3.203 (-4.47 to -1.66)	9.0-12.4
16	21	2:1AVB	Not sinus	NA	6	-3.055 (-4.18 to -2.18)	3.6-9.5
17	22	Sinus brady	108-110	-4.85 (-4.72 to -4.98)	3	-4.815 (-5.52 to -1.80)	
18	25	Sinus brady	113-128	-3.13 (-2.20 to -4.06)	3	-2.069 (-4.06 to -0.92)	

Table 1: Presenting rhythm and echocardiography parameters for individual cases in the series.
2:1AVB = 2:1 atrioventricular block, sinus brady = sinus bradycardia, AV interval = atrioventricular interval, N-LVIRT = normalised left ventricular isovolumetric relaxation time

PP-614

Unravelling the complex web of factors influencing critical congenital heart disease outcomes in new zealand: an exploration of ethnicity and beyond

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Background and Aim: Critical congenital heart disease (CCHD) is a significant contributor to infant mortality. While ethnicity has been known to influence the outcome of certain CCHD types, the contemporary factors contributing to this association remain unclear. Therefore, we aimed to identify factors associated with CCHD outcomes and examine their interaction with ethnicity within a national New Zealand (NZ) cohort between 2006 and 2019.
Method: We conducted a retrospective, population-based cohort study of CCHD cases reported in the national NZ Fetal, Cardiology and Cardiac Surgical, Maternity, Perinatal, and Infant Mortality datasets where the mothers chose to continue with the pregnancy. The primary endpoint was all-cause mortality within the first postnatal year or stillbirth. Primary outcomes in relation to various risk factors were analysed, including maternal factors (age at delivery, ethnicity, deprivation score, residential location), infant factors (co-morbidities, CCHD subtype, gestation at birth, birthweight z-score), and clinical care factors (diagnostic timing, management pathway – palliative or surgical, year of birth). Survival analysis was employed to scrutinise the data, and multiple risk factors were assessed in a Cox regression model.
Results: Between 2006-2019, there were 864,950 live births, stillbirths, and terminations from 20 weeks' gestation in NZ, including 1278 CCHD cases. Of the 1039 CCHD cases included, 241 (33.2%) met the primary endpoint. Ethnicity was an indicator of infant mortality risk with Indigenous Māori, Pasifika, and Asian unadjusted hazard ratios (HR) higher than referent Europeans (HR 1.5 [1.0-1.8], 1.6 [1.1-2.4], and 1.8 [1.2-2.7], respectively). Further univariate risk factors included: deprivation, infant co-morbidities, cardiac subtype, gestation at birth, birthweight z-score, and the management pathway. Multivariable analysis demonstrated that birthweight, deprivation, cardiac subtype, and management pathway were independently associated with the primary endpoint when adjusting for the observed risk factors.
Conclusions: Ethnicity was a significant factor associated with disparate CCHD infant outcomes, which was explained, at least in part, by modifiable and unmodifiable factors. Thus, this study

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contributes evidence-based modifiable targets for policymakers and healthcare leaders planning to advance health equity in CCHD outcomes by ethnicity in NZ.

Keywords: Indigenous, congenital heart disease, ethnicity, equity

PP-615

Antenatal diagnosis of pulmonary atresia with intact ventricular septum – a single centre experience

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Background and Aim: Pulmonary atresia with intact ventricular septum is a rare congenital heart disease, occurring in 1 in 22,000 live births in the United Kingdom and Ireland. The heterogenous and variable morphological spectrum of the condition adds to the complexity for postnatal assessment to determine whether a biventricular repair is amenable, or whether univentricular circulation is needed. Antenatal diagnosis allows for a safe plan for delivery, including guiding postnatal care and management plan. The objective of this audit is to review our institutional data and outcome of patients with pulmonary atresia with intact ventricular septum. *Method:* From 2015 to 2022, a retrospective analysis of our institutional data was performed using information from our fetal database and cross-referencing with confirmed postnatal diagnosis. Demographic features, anatomical features, management pathway and subsequent clinical outcomes were examined.
Results: 42 antenatal diagnoses of pulmonary atresia with intact ventricular septum were made in our institution in the period analysed, which resulted in 22 terminations of pregnancy (52%). There were 2 intra-uterine fetal deaths and 18 live births (43%). Of the live births, 6 families chose compassionate care in view of the complex cardiac history with counselling provided antenatally; information for one patient was not available as their care was in a different centre. The remainder of the patients (n=11) underwent cardiac investigations with echocardiography, and either cardiac catheter or computed tomography with subsequent discussion at our multidisciplinary meeting. 36% (n=4) of the patients were palliated with a single ventricle approach, whilst one patient is planned for 1.5 ventricles. One patient underwent a biventricular repair (9%). There were 3 deaths during the period (27.2%); one prior to intervention, one during and one separate to the cardiac procedure (due to respiratory illness). The care of 2 patients were taken over by a different centre following request for second opinion.
Conclusions: Pulmonary atresia with intact ventricular septum remains a rare and complex diagnosis with more than half resulting in termination of pregnancy and approximately 1/3 of the live births opting for compassionate care. The management postnatally remains challenging and the use of multiple imaging modalities should be employed to tailor the approach for patients.

Keywords: Congenital heart disease, fetal cardiology

PP-616

Prenatal diagnosis of critical and non-critical congenital heart disease in british columbia canada

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Background and Aim: British Columbia is a Province in Canada that spans over 944,735 km2, with parts of the Province only accessible by ferry. In addition, there is only one tertiary care hospital in the Province that services the needs of families who are expecting a child with congenital heart disease (CHD). The Province's geography and hospital service model necessitates accurate and timely prenatal diagnosis of CHD by fetal echocardiography. This allows for prenatal planning, reduced perinatal morbidity and mortality. We sought to evaluate our recent experience with prenatal diagnosis of CHD in our Province.
Method: This was a quality improvement (QI)/quality assurance (QA) review. Using our clinical and echocardiography imaging databases, we reviewed all CHD cases that presented between January 2015 and December 2022. Cases were classified as critical (requiring intervention in the first month of life) and non-critical but significant (requiring intervention in the first year of life). The following data was collected: diagnosis, gestational age at diagnosis, birth plan, and need for relocation. Frequency (%) tables were generated and medians (interquartile ranges) are presented.
Results: The total number of mother's who had a fetal echocardiogram performed was 3797, with 735 fetuses identified as having a prenatal diagnosis of critical (n=482) and non-critical but significant CHD (n=253). The rate of diagnosis of critical and non-critical but significant CHD in those referred for a fetal echocardiogram was 19%. Median gestational age at time of diagnosis was 22 (21-26) weeks and 3 (1-5) days. The termination rate was 23% in the critical and 19% in the non-critical diagnoses. Intrauterine fetal demise occurred in 2% of critical and 2% of non-critical diagnoses. After diagnosis, 96% of critical diagnoses were triaged to deliver at the tertiary care centre, of which 40% needed to relocate. 74 % of non-critical diagnoses planned to deliver at the tertiary care centre, of which 30% needed to relocate.
Conclusions: The fetal cardiology program in our province is identifying critical and non critical but significant CHD and triaging appropriately to higher level care.

Keywords: Fetal Cardiology, Quality Improvement/Quality Assurance, Congenital Heart Disease

PP-617

Risk factors and presentations of fetal echocardiography: What we found in a tertiary care centre ?

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Background and Aim: Fetal Echocardiography is already established screening tool for the detection of congenital cardiac anomalies. But its use and awareness remains less in many under developed and developing countries of the world. So limited data is available about the usage and outcome of fetal echocardiography in theses settings To study risk factors and presentations of fetal echocardiography.
Method: All pregnant women referred for fetal echocardiography in pediatric cardiology department of Bangabandhu Sheikh Mujib medical university (BSMMU) from June 2022 to July 2023 were included in the study.
Results: A total of 121 pregnant women underwent fetal echocardiography. The mean age of patients was 30± 4.8 years. Study population presented with gestational age of <16weeks to > 37weeks (mean of 25.7 ±4.4 weeks). The various risk factor includes

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Diabetes Meletus35(16.35%), Hypertension11(5.14%), Hypothyroidism 15(7%), Epilepsy 1(0.46%), Thalassemia 1(0.46%), Maternal CHD 3(1.4%), Maternal RHD 3(1.4%), Polyhydramnios 2(0.93%), Bad obstetric history 7(3.2%), previous child with CHD 1(0.46%), Fetal hydrops on USG 2(0.93%), Twin pregnancy 1(0.46%) and others 4(1.85%). Total 86 (40.18%) echo reports were normal and 128 (59.8%) were abnormal. Different abnormalities detected includes VSD 7(3.27%), AVSD 1(0.46%), Truncus arteriosus 4(1.86%), Tricuspid atresia 6(2.8%), single ventricle 2(0.93%), Bradycardia 4 (1.86%), cardiomegaly 3(1.4%), Fetal hydrops 3(1.4%), pericardial effusion and pleural effusion.
Conclusions: Fetal Echocardiography has provided a wide benefit of diagnosing cardiac defect even before the baby born. This helps to resuscitate the baby at most effective care level and allows parents to be prepared for that. But in our country mothers are coming in late gestational age rather than 22-28 weeks.
Proper awareness and referral can improve the scenario.

Keywords: Risk factors, presentation, fetal echocardiography

PP-618

Fetal bradycardia and neonatal tachycardia due to an ectopic atrial focus

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Background and Aim: Fetal bradycardia is defined as a sustained heart rate less than 100 beats per minute (bpm) and represents a potentially life-threatening condition for the fetus with various causes. One of the most important cardiac causes is AV block. A usually benign differential diagnosis to 2:1 AV block is a constant atrial bigeminus with blocked premature atrial beats
Method: At 21 weeks of gestation fetal bradycardia was first observed in our case. Fetal echocardiography showed a structural normal heart with a constant blocked atrial bigeminus with an ventricular rate of 80 bpm. The patient was monitored without therapy once weekly. He showed regular intrauterine growth without signs of cardiac decompensation until 36 weeks of gestation with sustained blocked atrial bigeminus and resulting bradycardia. Because of the lacking possibilty of an adaequate differentiation between the blocked atrial bigeminus and other, potentially life-threatining causes of fetal bradycardia during a sponatneous birth like perinatal asphyxia, we decided to perform cesarean section at 36 weeks of gestation.
Results: Postnatal the patient showed a transient tachypnoea of the newborn with a need for non-invasive CPAP for 2 days. The ECG showed the blocked atrial bigeminus with ventricular rates of 80 bpm as seen intrauterine. After a few days of life, episodes of ectopic atrial tachycardia were observed increasingly. A therapy with propafenone with a maximum dose of 250 mg/m2 was started, but discontinued due to the development of broad complex tachycardias under therapy. Under Sotalol (3,5 mg/kg) and Digoxin (0,1 mg/m2) we were able to achieve stable sinus rhythm.
Conclusions: Blocked atrial bigeminus is a generally benign cause of fetal bradycardia and an important differential diagnosis to 2:1 AV block. The differentiation of this benign situation to other, potentially life-threatening situations is crusial as usually no therapy is needed. Ectopic atrial tachycardia can develop due to the pathological focal autaomaticity in the fetus as well as in the newborn, therfore close monitoring is manditory in these patients

Keywords: Blocked atrial bigeminus, fetal bradycardia, ectopic atrial tachycardia

PP-619

Tailoring fetal echocardiography indications to the limited resources in Saskatchewan, Canada

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Background and Aim: Congenital heart disease (CHD) is the leading cause of infant mortality due to birth defects. Prenatal diagnosis of CHD using fetal echocardiography has many benefits including assessment of the optimal site for delivery and reduced costs to the healthcare system regarding treatment and transportation. The indications for fetal echocardiography screening for congenital heart disease (CHD) in Saskatchewan, Canada, were previously based on the 2014 American Heart Association recommendations. Due to limited resources, these indications were changed in 2017, to introduce an intermediate level where fetal echocardiography was only indicated if initial screening by obstetric ultrasound was abnormal. The aim of this study was to assess the clinical impact of these changes on the prenatal diagnostic rates of CHD in Saskatchewan.
Method: All women undergoing fetal echocardiography screening for CHD in Saskatchewan for 3-years before (1Jan2014-31Dec2016) and 3-years after (1Jan2017-31Dec2019) these changes were introduced, were identified from our fetal echocardiography database. Data was abstracted retrospectively including: fetal echocardiography date and location; maternal age, location of residence and obstetrical history; and fetal echocardiography indication, gestational age, and cardiac and extra-cardiac diagnoses.
Results: Women undergoing fetal echocardiography screening before (N=567) compared with after (N=597) these changes were introduced were of similar age (30.3±7.4 vs. 30.2±6.4 years, p=0.858) and at similar gestation (28.2±8.6 vs. 28.2±7.4 weeks, p=0.988). Maternal and fetal factors also appeared similar, other than maternal diabetes as the indication for fetal echocardiography (14% vs. 10%). This is likely due to maternal pre-existing diabetes with good control (HbA1c <7% at conception) being changed to an intermediate risk indication. The percentage of CHD detected at fetal echocardiography remained similar (50% vs. 49%) before and after changing the indications for fetal echocardiography in Saskatchewan.
Conclusions: Introduction of an intermediate level of indications for fetal ECHO does not appear to have significantly affected prenatal diagnosis rates of CHD in Saskatchewan. This has important resource implications for our limited provincial availability of fetal echocardiography. This data is being used to inform healthcare resource planning in the development of a provincial Fetal Echocardiography Program to ensure all women in Saskatchewan receive optimum care independent of geographical location or socioeconomic demographics.

Keywords: Fetal echocardiography, congenital heart disease, screening, indications, resource limitations

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PP-621

Perinatal management pathways for transposition of the great arteries with intact ventricular septum in cardiac units in the UK and Ireland

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Background and Aim: Transposition of the Great Arteries with Intact Ventricular Septum (TGAIVS) is increasingly diagnosed antenatally. Outcomes for definitive surgery are excellent but there remains an important cause of pre surgical morbidity and mortality resulting from neonatal hypoxia due to poor mixing through the atrial communication and arterial duct. Predicting this risk antenatally is imperfect. The pathways developed by units in the UK to mitigate this risk vary depending on local infrastructure and resources. Our aim was to examine how individual units in the UK have sought to minimise the preoperative risk of patients with an antenatal diagnosis of TGAIVS, understand the constraints of geography and staffing, identifying areas for future study to improve outcome in this condition.

Method: Questionnaire to all cardiac surgical units in the UK to obtain details of antenatal and perinatal pathway for patients with an antenatal diagnosis of TGAIVS.

Results: All 11 cardiac units in the UK and Ireland that offer surgery for TGAIVS responded. 5/11 (45%) of units have maternity and cardiac services collocated. The distance for non-collocated units ranged from 0.7-3.6 miles from maternity unit to cardiac centre. Final assessment in fetal cardiology was 32-34 weeks in 2/11 (18%), 34-36 weeks in 7/11 (63%) and over 37 weeks in 2/11 (18%). The usual planned mode of delivery, for patients without antenatal concerns, was awaiting spontaneous labour in 3/11 (27%), induction of labour in 6/11 (55%), and elective caesarian section in 2 units (18%) both of which were non collocated units. The median planned week of delivery was 39 weeks. Two units are able to accommodate geographically distant patients. Units had 2-4 interventional cardiologists. No unit has an interventional cardiologist present at all deliveries. 5/11 (45%) performed septostomy at the bedside, 6/11 (55%) in the cardiac catheterisation laboratory.

Conclusions: The increase in antenatal diagnosis of TGAIVS provides an opportunity to improve outcome as there are variations in current pathways. Further research is needed to compare different models of care to establish the impact of colocation and optimal mode and timing of delivery for this high-risk group.

Keywords: TGA, fetus, outcome, transposition, CHD

PP-622

Hemodynamic and histological changes in a fetal animal model for left ventricular hypoplasia – a feasibility pilot studY

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Background and Aim: Brain growth and development is altered in fetuses with hypoplastic left heart syndrome due to retrograde aortic flow pattern and impaired cerebral perfusion. Little is known about histological changes of the affected fetal brains. We aimed on determining blood flow patterns by fetal magnetic resonance imaging (MRI) to be associated with histological findings in a chronic animal model for left ventricular (LV) hypoplasia.

Method: Eight pregnant ewes (four with twins) with a mean (SD) body weight 69.5+/-10.8 kg were anesthetized and intubated at two time points: (A) at mid of gestation (0.5) for percutaneous left atrial (LA) coil implantation guided by fetal ultrasound to induce a mitral valve stenosis with reduced filling of the LV, and (B) at late gestation before birth (0.9) for cardiac and cerebral MRI. Successful development of LV hypoplasia was determined by functional cardiac MRI on a 3T scanner and 2D quantitative flow MRI at the ductus arteriosus and the proximal ascending aorta. Macroscopic evaluation of the fetal brain was performed. Histological findings were assessed by hematoxylin and eosin (HE), Luxol fast blue and by immunohistochemistry (IH) with neuronal and glial markers.

Results: Three animals developed LV hypoplasia and were compared to five control (sham) animals. The left ventricular end-diastolic volume in the LV hypoplasia group was decreased with 0.32 mL/kg (+/-0.05) vs. 0.46 mL/kg (+/-0.05) in controls. The flow in the proximal ascending aorta was reduced with 26.7 mL/kg/min (+/-.2) vs. 184.1 mL/kg/min (+/- 45.0) in controls. This was also associated with increased flow in the ductus arteriosus 215.6 (+/- 43.0) vs 164.0 (+/- 54.1). Cerebral findings were analysed in post mortem T2 weighted fast spin echo anatomical imaging as well as DTI. Cerebral pathology indicates a less mature cortical brain development (panel).

Conclusions: Our preliminary results demonstrate the impact of LV hypoplasia on brain perfusion determined by fetal MRI in the context of cellular changes shown by brain histology. In future studies, these cellular alterations have to be further determined in the altered brain development within this high risk population.

Keywords: Hypoplastic left heart syndrome, brain development, fetal

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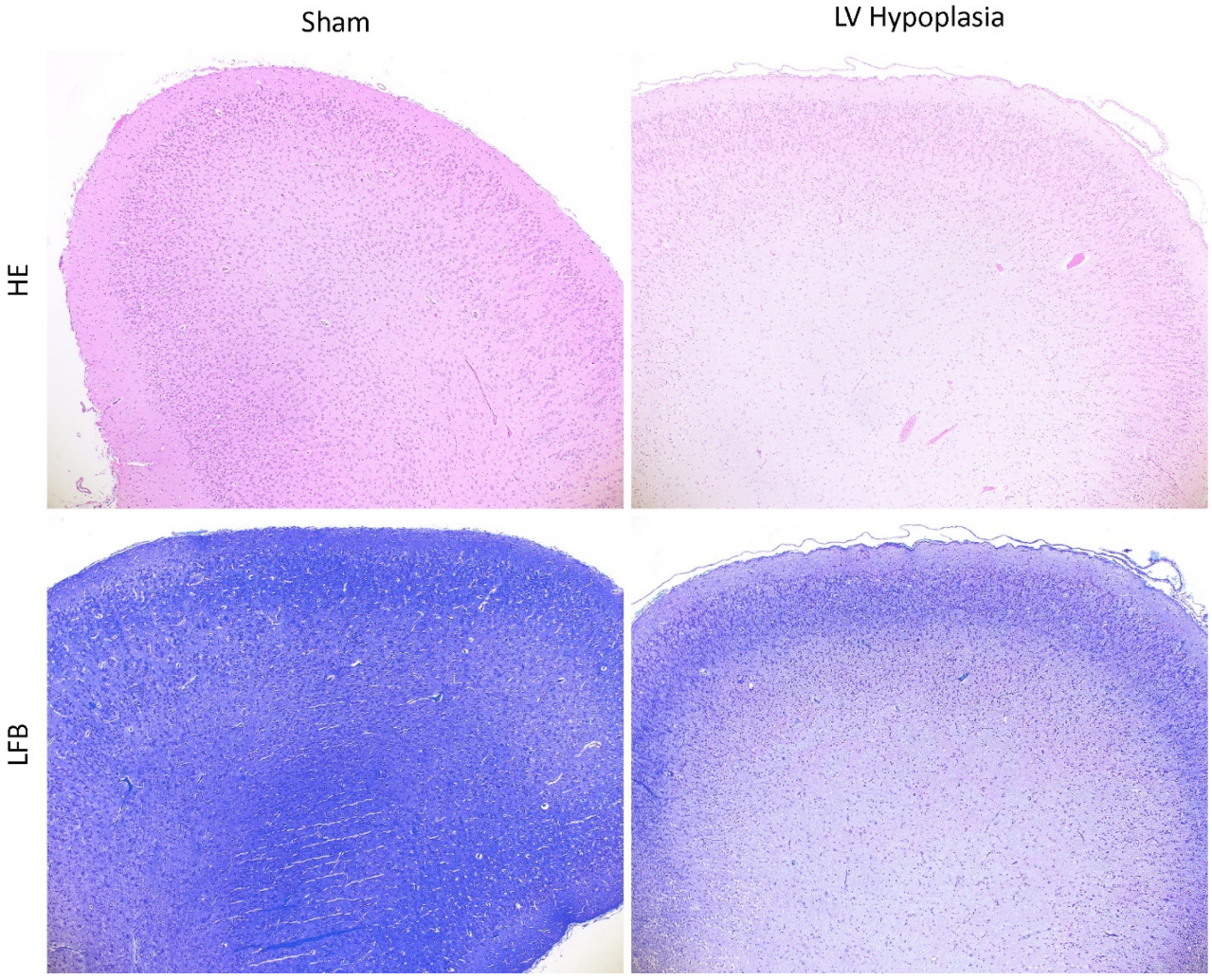
Panel



General Cardiology

PP-625

A systematic review of clinical practice guidelines on the management of malnutrition in children with congenital heart disease

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Background and Aim: Congenital heart diseases (CHD) are one of the most common inborn disorders, with a prevalence of 0.8-1.2%. Affected children are often malnourished due to increased requirements. This may lead to severe long-term complications, such as growth impairment, limitation of a child's development, or increased mortality. Several authoritative organizations published guidelines addressing nutritional intervention in children with CHD. We aimed to systematically assess the consistency of recommendations, the methodological quality of guidelines, and the quality of evidence supporting each recommendation.
Method: PubMed, Embase, Cochrane Database, World Health Organization Global Index Medicus, and 16 scientific societies websites were searched until September 2023. Any clinical guidelines or practice recommendations focusing on the management of malnutrition in children with CHD were included. The guideline quality was assessed using the Appraisal of Guidelines for Research and Evaluation 2nd edition (AGREE II), tool. An online tool, My AGREE PLUS has been used by 2 reviewers to assess the included guidelines.
Results: After screening 765 records, two guidelines published in 2013 and 2022 were included in our review. The quality scores were very high in three domains (Scope and Purpose 100%, Clarity of Presentation 94%, and Editorial Independence 100%). In other domains scores were lower (Stakeholder Involvement 31%, Rigour of Development 67%, Applicability 17%). Main issues concerned the lack of implementation advice or tools and the lack of criteria to measure the application of guideline recommendations. The score for the Overall Assessment was 83%. Due to the low number of included guidelines, we did not calculate the median scores for individual domains.
Conclusions: Despite an abundance of literature on CHD-related malnutrition, only two guidelines met our inclusion criteria. The main reason for exclusion was the absence of any system of rating the evidence. Included guidelines were of good quality, within specific recommendations, both publications were largely in agreement. There is a pressing need for comprehensive,

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multi-threaded guidelines, incorporating implementation strategies and methods for performance assessment.

Keywords: malnutrition, guidelines, CHD

PP-626

Syndromes and malformations in live-born children with double outlet right ventricle in norway; A population-based study

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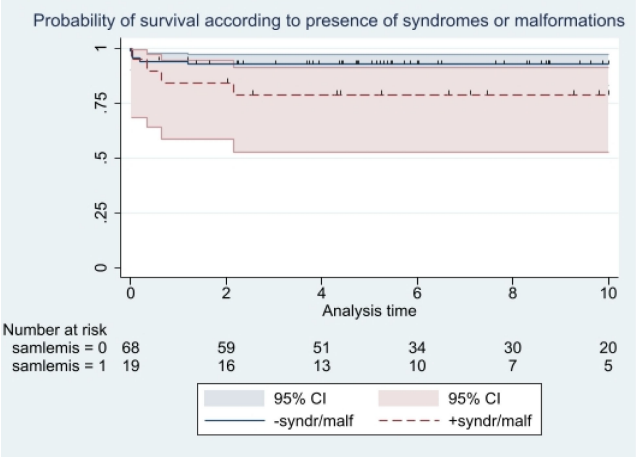
Background and Aim: Population-based studies of the prevalence of syndromes and malformations among live born children with double outlet right ventricle (DORV) are lacking. We therefore surveyed the prevalence of syndromes and malformations among Norwegian children born with DORV and to describe treatment courses, interventions, complications and mortality.
Method: All children born with DORV between 2003 and 2017 were identified in the Oslo University Hospital registry. Patients' characteristics, presence of syndromes or malformations, interventions, complications, and deaths were recorded. Echocardiographic data were reviewed for classification according to current standards. We investigated time-dependent surgical reintervention and mortality using Kaplan–Meier analyses and determinants of treatment complications, reintervention, and death using regression analyses.
Results: 93 children born with DORV between 2003 and 2017 represented an annual mean prevalence of 1.07 per 10,000 births in Norway. 25 of these children were born with a wide range of syndromes or other non-cardiac malformations and 6 of these children received palliative care. With an intention to treat, a surgical route with primary biventricular repair was followed for 12 children, staged biventricular repair for 5, and univentricular repair for 2 children. Major complications occurred in 0% and 10.5% children following catheter or surgical intervention, respectively. No significant determinants of surgical complications were identified. Overall survival following treatment was 84.2%, 84.2%, 78.6%, and 78.6% and corresponding freedom from surgical reintervention was 93.8%, 87.5%, 80.2%, and 80.2% at 1, 2, 5, and 10 years, respectively. Kaplan-Meier plot revealed a non-significant trend for increased risk of mortality with presence of syndromes or non-cardiac malformations among children with DORV. No increased risk of surgical re-intervention was found for children with syndromes or malformations.
Conclusions: We found an annual incidence of DORV of 1.07 per 100.000 live births in Norway. Approximately 1:4 children with DORV were born with additional non-cardiac malformations or syndromes,which in 1:3 cases were of a severity leading to comfort care. However, most children with syndromes or malformations accompanying DORV were offered tailored treatment encompassing primary biventricular, staged biventricular or univentricular repair with favourable outcomes although risk of mortality tended to be increased compared to children with DORV without additional syndromes or malformations.

Keywords: DORV, mortality, syndrome, malformation, double outlet right ventricle

Survival among children with DORV according to presence of syndromes or malformations



PP-627

Effect of kidney transplant on ventricular dysfunction in a patient with end-stage renal disease

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Background and Aim: Cardiovascular disease is a major cause of morbidity and mortality in end-stage renal disease (ESRD) in adult patients. In children it is usually due to associated cardiomyopathies. It is important to perform a study aimed at the origin of the heart failure in order to offer a targeted and effective treatment, keeping the patient on the waiting list for renal transplantation.
Method: We present the case of a 13-year-old patient with ESRD secondary to focal segmental glomerulosclerosis (TRCP6 mutation) corticoresistant with debut in November 2020. She´s been on peritoneal dialysis sinnce October 2021.
Results: Cardiological evaluation in June 2021 was normal. Four months after the start of peritoneal dialysis, the patient began to show signs of congestive heart failure (CHF), progressive dyspnea, orthopnea, peripheral edema, with New York Heat Association (NYHA) functional class II-III. A new cardiological evaluation was performed in February 2022 with ventricular dysfunction, with ejection fraction (LVEF)35%, severe left ventricular dilatation (VITD 60.4mm) and moderate mitral insufficiency and moderate pericardial effusion (15mm). Complete study with MRI without gadolinium in March 2022 (left ventricular dysfunction with LVEF 35%), genetic study of cardiomyopathies (Negative) and cardiac catheterization without data of pulmonary hypertension (PCP 19 mmHg, IRVP 4 UWm2, CI 1.95l/min/m2). In March 2022 she starts treatment with sacubitril/Valsartan with clinical stabilization, so she remains on the waiting list for renal transplantation. She receives monthly cycles of levosimendan. She maintains treatment with Carvedilol, Amlodipine and Darbepoetin alfa. In June 2022 kidney transplant was perfomed. She was a good evolution, blood perssue controlled with sacubitril/valsartan, carvedilol, amlodipine. Post-transplant controls

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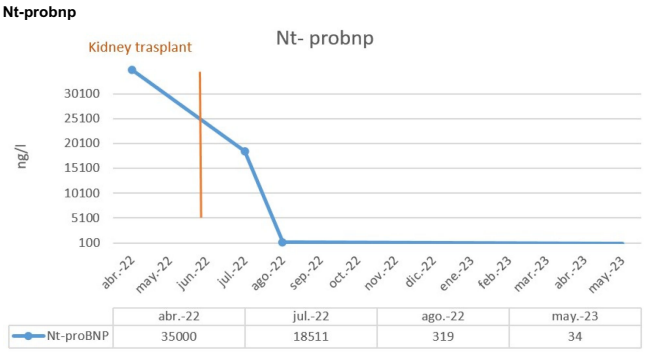
showed normalization of cardiac function, disappearance mitral insufficienccy; as well as improvement of analytical parameters (NtproBNP 34 ng/L).
Currently she is in NHYA functional class I, under treament with sacubitril/valsartan and amlodipine.
Conclusions: Kidney transplantation in ESRD patients with advanced systolic heart failure results in an increase in LVEF, improves functional status of CHF, and increases survival. To abrogate the adverse effects of prolonged dialysis on myocardial function, ESRD patients should be counseled for kidney transplantation as soon as the diagnosis of systolic heart failure is established.

Keywords: Heart failure, kidney transplant, end-stage renal disease,



PP-628
Atypical cases of kawasaki disease. pitfalls in diagnosis and cardiac outcomeS

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Background and Aim: Atypical Kawasaki disease (KD), usually leads to delayed diagnosis and appropriate treatment, increasing the risk of cardiac complications. We report the leading clinical symptoms and/or laboratory features at the time of diagnosis in a group of children with atypical KD and correlate cardiac complications with the time of diagnosis.
Method: From November 2022 to October 2023, six children with atypical KD were diagnosed in a tertiary care hospital.
Results: There were 3 boys/3 girls, aged between 14 to 24 months (5/6) and 1/6 was 15 years-old. The time of diagnosis ranged from 2-12 days (5/6), and only in the 15-year-old patient was 22 days. All patients had fever and 5/6 skin rash. Other leading symptoms were sacroiliitis or hip joint effusion (2/6), balanoposthitis (1/6), limping with normal hip ultrasound (1/6), while 3/6 patients had prominent gastrointestinal symptoms: hyperamylasemia with high ALT and γ-GT (1/6), abdominal distention with stool discolouration, jaundice and high total and direct bilirubin (1/6) and hepatosplenomegaly with elevated AST, ALT and γ-GT (1/6). From six laboratory findings for diagnosis of incomplete KD, total WBC count >15,000/mm3 had 2/6 patients, anemia:6/6, platelet count >450,000:3/6, serum albumin <3g/dL:1/6, elevated ALT levels:1/6, and pyuria:2/6 children. As for cardiovascular complications, all patients had coronary arteries involvement, with the most serious lesions (coronary dilatation with z-score>5) in 2/6, who had the longest time since diagnosis (22 and 12 days respectively). All patients treated with intravenous immunoglobulin: 2/6 had single infusion and 2/6 two infusions. As for additional therapy: low-dose aspirin 3/6, clopidogrel (due to G6PD deficiency) 2/6, and aspirin with clopidogrel in 1/6. The dilatation regressed gradually within 4 to 6 weeks in 4/6, but in two patients with z-score>5 persisted for 3 months. Larger coronary dilatation and delayed regression were associated with delayed diagnosis.
Conclusions: Early diagnosis of atypical KD is a challenge for the pediatrician, because cardiovascular sequelae appear the same with typical cases. As a normal echocardiogram in the first week of illness does not rule out the diagnosis of atypical KD, we suggest that serial echocardiograms should be performed in all patients with suspicious symptoms and uncertain diagnosis.

Keywords: coronary dilatation, atypical Kawasaki, children

PP-629
Implementation of value-based healthcare principles through the use of patient-reported outcome measures within a paediatric cardiology unit

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Background and Aim: This study explores the implementation of Value-Based Health Care (VBHC) principles through the use of Patient-Reported Outcome Measures (PROMs) within a Pediatric Cardiology Unit of a tertiary hospital. It aims to explore the effectiveness and challenges in implementing patient-centric outcomes, cost efficiency, interdisciplinary collaboration, patient participation, technological advancements in data collection, aligned incentives, and continuous improvement and standardization.
Method: The methodology involved adapting the Outpatients Questionnaire 2011 (Picker Institute European -NHS) to evaluate patient experiences in the Pediatric Cardiology Unit. A comprehensive survey was conducted, encompassing various aspects of patient care, including waiting times, clarity of signage, cleanliness, communication regarding tests and interventions, professional conduct, privacy, decision-making, trust, medication information, and overall respect and dignity. The responses were quantitatively analyzed to derive insights into the effectiveness of VBHC implementation.
Results: The results indicated high levels of patient satisfaction and trust in the healthcare professionals. Notable findings included efficient waiting times, clear communication regarding medical procedures and results, and a strong sense of respect and dignity. The data suggested a successful integration of VBHC principles, particularly in areas of patient involvement in decision-making, clarity of medication information, and overall satisfaction with the care received.
Conclusions: The study highlights the effectiveness of implementing VBHC in a Pediatric Cardiology Unit, demonstrating significant positive impacts on patient experiences through the use of PROMs. Key strengths identified include interdisciplinary collaboration, patient-centric approaches, and effective communication strategies. The findings underscore the importance of continuous measurement and improvement in healthcare services,

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aligning with the overarching goals of VBHC to enhance patient outcomes and value. The study suggests a need for ongoing evaluation and adaptation of VBHC strategies to address identified gaps and enhance overall healthcare value. The methodology, while comprehensive, may have limitations in capturing the full scope of patient experiences and the nuanced impacts of VBHC implementation. Future directions could include further exploration of cost-related aspects and technology integration for data collection and analysis.

Keywords: Value-Based Healthcare principles, VBHC, Patient-Centered Care, Patient-Reported Outcome Measures, PROMs.

PP-630

Kawasaki disease: description of a 25 year cases serie from a referral hospital in northern spain

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Background and Aim: Kawasaki disease (KD) is a self-limited systemic vasculitis that generally occurs in children. Children with KD are at risk for cardiovascular (CV) sequelae, particularly coronary artery abnormalities (CAAs). The risk is higher without proper treatment with intravenous immunoglobulin (IVIG).
Method: We describe a cohort of patients (observational study) with KD diagnosis over 25 years in a referral university hospital in northern Spain, before the SARS-CoV-2 pandemic. Classic diagnosis criteria were used. CAAs were identified by echocardiography and/or coronary CT.
Results: 61 patients (without gender differences) were diagnosed with KD. Mean age at diagnosis: 3,6 years. Due to seasonality: 65% summer, 9% spring. 78,2 % were classified as complete KD. There was an increase in acute phase reactants, leucocytosis and thrombocitosis. All patients presented a monophasic disease course with mean duration of symptoms of 12,8+/-4.9 days. 10 patients (6 %) developed CAAs. Only 3 maintained CAAs beyond the first year after the acute illness. 96,6% received adequated treatment with IVIG (2g/kg before tenth day of the onset of the symptoms), 98% received acetylsalicylic acid at anti-inflammatory doses and subsequently at antiplatelet doses. 4 received intravenous corticosteroids (due to refractory or high-risk KD according to Kobayashi score and/or National consensus on diagnosis, traetment and cardiological follow-up of KD score from the Spanish Paediatric Associacion -AEP-),3 received infliximab due to refractary illness. No other inmunosupressants were required. A 16-year-old patient died because of myocardial ischemia resulting from previously undiagnosed CAAs. He presented unexplained untreated fever (probably incomplete KD) when he was 7 months old. The incidence rate of KD in our country was 0,37 cases/100.000/year but it is much higher in the last 12 years.
Conclusions: Our incidence is much lower than reported in other series but it has tripled in recent years. Different factors can explain it: an increase in real incidence or underdiagnosis/underregistration in the first years of study. However, clinical features are similar than reported. Only 1% of cases maintained CAAs beyond de first year. The vast majority of patients were treated early and most of them responded to usual IVIG treatment.

Keywords: Kawasaki disease, children, coronary artery abnormalities

PP-631

Correlation of electrical and mechanical response after cardiac resynchronization therapy

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Background and Aim: Cardiac resynchronization therapy (CRT) constitutes a cornerstone in advanced heart failure treatment. Although structural remodeling correlates with improved long-term outcomes, the role of electrical remodeling is poorly understood. In practice, the degree of CRT response is assessed 6 months after CRT implantation using echocardiographic change (Δ) of left ventricular volume and ejection fraction (EF).
We undertook this study to determine the potential relationship between ventricular and electrical remodeling.
Method: In this study, all patients underwent baseline 12-lead ECG and transthoracic echocardiography (TTE) prior to CRT implantation and after. Response to CRT was defined by ≥ 5% absolute improvement in EF on TTE and electrical remodeling as a decrease in QRS width.
Results: Twenty patients were included (age, 54 [13] years; 80% men); only 35% of them had ischiemic cardiomyopathy, and the mean EF was 27%. The mean QRS duration prior to CRT was 160 ms. The mean EF after CRT was 42%, and EF improvement (delta EF) was 63%, from 12% to 230% ($P < .05$) after implantation. Ninety-five (95%) patients were considered responders as they had a LVEF ≥ 5% absolute improvement. The mean QRSd reduced from 160 to 127 ms ($P < .05$), whether in biventricular pacing or LV pacing. These patients showed a significant decrease in QRS duration on follow-up in addition to a significant improvement in ejection fraction. A noteworthy finding is the correlation between QRS duration and LVEF after CRT ($P = 0.048$). traducing the relationship between the presence of ventricular and electrical reverse remodeling achieved with CRT.
Conclusions: Our findings support the importance of programming to obtain the narrowest-paced QRS possible. The inherent limitations of sample size and the descriptive nature of the study.

Keywords: remodeling, resynchronisation, QRS

PP-633

Heart pseudoaneurysm developed in the course of multisystem inflammatory syndrome in children

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Background and Aim: Multisystem Inflammatory Syndrome in Children (MIS-C) is a rare condition associated with SARS-CoV-2 infection. It is a novel disease, and its pathophysiology, as well as the complete clinical picture, are still unknown.
Method: We present the case of a 22-month-old boy exhibiting signs of MIS-C and testing positive for SARS-CoV-2 IgG antibodies. During routine echocardiography, a hypokinetic and thin-walled aneurysm of the left ventricle apex was identified. Subsequent heart magnetic resonance imaging revealed a dyskinetic false aneurysm of the left ventricle apex with a full-wall ischemic scar. Aortography confirmed normal coronary artery course, with sufficient perfusion of essential branches and no stenosis or aneurysms. The heart team consulted the boy and determined his eligibility for surgery. The pseudoaneurysm was surgically removed up to the margin of healthy tissues, and both the surgery and periprocedural period were uneventful.
Results: Determining the origin of the aneurysm proves challenging. The most likely etiology appears to be an ischemic lesion arising from a disorder in coronary circulation due to the complex course of MIS-C. It is possible that the disorder resolved spontaneously before aortography was conducted. Although other etiologies, such as perinatal, intrauterine, or congenital lesions or complications of pericarditis, should be considered, we find these reasons less probable.
Conclusions: The etiology of the presented left ventricle pseudoaneurysm cannot be definitively confirmed. However, the MRI results, coupled with histopathological examination, suggest reversible ischemic changes in the coronary arteries in MIS-C as the most likely cause.

Keywords: SARS-CoV-2 virus, COVID-19, Multisystem Inflammatory Syndrome in Children, heart aneurysm, cardiac surgery

PP-634
Precordial pain, sincope and heart failure in a 14 yo boy, demasking an aortic dissection

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Background and Aim: To present a 14 yo boy, admitted in our clinic after a severe precordial pain followed by syncope, during an amical discussion with his mother. He was transported alone with ambulance in the ED, where he was stable, with normal ECG, no precordial pain, normal O2Sat and blood pressure and cardio-pulmonary X ray with enlarged cardiac silhouette and aortic button that did not panic the on-duty doctors. He was monitored in the pediatric department.
Method: Next day he was cardiological evaluated, clinical, ECG and Echocardiography. No pain was found, but the patient was obnubilated, not able to walk on his legs, presenting light dyspnea, tachycardia and hypotension.
Results: Grade IV/6 diastolic murmur in the aortic area, absent pulse at the right radial artery and hepatomegaly were found. ECG showed inverted T waves in left precordial leads. Echocardiography revealed large aneurysm of the ascending aorta, Zscore 9.9, aortic dissection with a flap starting from the supra bulbar position to the aortic arch, involving the brachiocephalic trunk and severe aortic insufficiency. Angio CT was performed showing large aortic dissection, Stanford A, DeBakey II, with obstruction of the commom right carotid artery and a Kommerell diverticulum. Cardiac biomarkers were increased. The patient worked in constructions, lifting heavy weights. He was addressed to the pediatric cardiovascular surgery, where Bentall-de Bono procedure was performed using a 26 mm Dacron Graft Conduit with St Jude 23 mm prosthesis and coronary artery reimplantation. Anticoagulation started after surgery. Fever appeared after 10 days, and endocarditis treatment was started despite any positive hemoculture, because the patient was on antibiotics. Vancomycine, Rifampicine and Amikacine was started, with a good evolution. Angio CT was repeated one month later, showing the increase of the Kommerell diverticulum and residual dissection flap.
Conclusions: Aortic dissection is very rare in children, especially in the absence of Marfan features. Severe precordial pain with syncope must be evaluated by echocardiography and angio CT if necessary. A second intervention is required for the residual flap and Kommerell diverticulum dilatation. No effort, regular follow-up and chronic anticoagulation with INR control is recommended. Genetic evaluation is mandatory.

Keywords: severe precordial pain, syncope, aortic dissection, children

PP-635
Assessment of right ventricular function in children after surgical repair of tetralogy of fallot

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Background and Aim: To examine the relationship between plasma levels of N-terminal proB type natriuretic peptide (NTproBNP) value in patients after surgical correction of Tetralogy Fallot with the dimensions of the right heart chambers, echocardiographic parameters of systole and diastolic function of the right heart.
Method: The research include 39 patients aged 1-17 years, 1±17 years after the primary surgical correction of the tetralogy Fallot, divided into two groups: Group I- patients who underwent surgical correction less than 10 years ago beginning of the study and Group II-patients who underwent operative correction in a period longer than 10 years before beginning of the study. Patient histories, measured NT-proBNP levels and transthoracic echocardiography parameters were analysed.
Results: 39 patients participated in the study, with an average age of 9.1±4.63 years, and a range of 1 to 17 years. 20 patients (51.3%) underwent surgery less than 10 years ago, on average 4.85 years ago, while 19 (48.7%) respondents underwent surgery more than 10 years ago, on average 11.74 years ago. It was found that there is a significant statistical difference in gender representation between the two investigated groups (x2=5.267; p=0.022). No significant statistical difference was found in the frequency of therapy consumption (x2=3.288; p=0.070). Dysrhythmias were not found in any patient. In relation to the elapsed time since the operation, significantly higher values of PVR were found in subjects who had the operation performed ten or more years ago (p=0.022). Significantly higher values of FS-RVOT (%) (p=0.009) and significantly higher values of RV-FAC (%) were also determined; p=0.012. A significant correlation between the value of NTproBNP and the E"/A" index was determined, r=0.468; p=0.037. The relationship was evaluated as a medium-strong relationship, with a positive sign.

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Conclusions: Quantification of right heart function is possible and should be routinely performed using a combination of different parameters. Increased plasma NT-proBNP levels in patients after surgical repair of ToF are related to RV systolic and dyastolic dysfunction, evaluated by 2D-, M-mode and TDI.

Keywords: tetralogy Fallot, right heart, NTproBNP, echocardiography

PP-636

A rare case of cardiac rhabdomyomas and long qt syndrome

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Background and Aim: We present a rare case of 3 day old neonate born premature at 34 weeks gestation age. The child presented to our centre with respiratory distress syndrome and bradycardia with heart rate varying between 70-80 beats per minute.
Method: Surface Electrocardiograms and transthoracic echocardiography was done. The echocardiography was done with GE-Vivid E95 machine.
Results: The surface EKG revealed sinus rhythm with long QTc of 624 milliseconds by Fridericia formula and 659msec by Bazett's formula. There was functional 2:1 Atrioventricular block as the length of time that repolarization was taking was too long and hence precluding 1:1 conduction all the time. Echocardiography revealed a large echogenic mass likely as rhabdomyoma in upper ventricular septum measuring about 2*3 cms and 2 additional small masses in left ventricular lateral wall. The child was hemodynamically stable and was managed for respiratory distress syndrome. The child was discharged home on oral propranolol and mexiletine. Child was scanned every for 4 months over last 2 years and the rhabdomyomas have largely regressed We suspected child to have Tuberous sclerosis,although there are no other systemic manifestation at 2 years of age in this case. The genetic tests are awaited and the Qtc has shortened with latest EKG showing QTc of 388 msec (Bazett) with 1:1 conduction. We reviewed the literature and found this is the only reported case of concurrently occurring cardiac rhabdomyomas and long QT syndrome.
Conclusions: Cardiac rhabdomyomas are known to be associated with Tuberous Sclerosis complex and long Qt syndrome with functional 2:1 AV block can occur in type iii long QT.

Keywords: Cardiac Rhabdomyoma, long QT

PP-637

Acute myocardial infarction in a pre-teen as an initial presentation of glomerulopathy with nephrotic range proteinuria. A case report

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Background and Aim: Acute myocardial infarction (AMI) in children is rare; AMI can occur in patients with congenital heart disease, specifically with coronary artery anomalies or post coronary aneurysm and thrombosis with Kawasaki disease. Infrequently, AMI presents in healthy children. We present an uncommon presentation of AMI in a healthy pre-teen with glomerulonephritis and nephrotic range proteinuria.
Method: An 11-year-old male presented to the emergency department with chest pain, severe diffuse headache and brief episodes of blurry vision. He was tachycardic and had a significantly elevated blood pressure with measurements up to 200 mmHg systolic. Laboratory investigations revealed hypoalbuminemia, hyperlipidemia, proteinuria and low C3 values. Troponin was significantly elevated, and ECG showed changes suggestive of ischemia. He progressively developed migrated shoulder pain and ST dynamic changes were observed (figure 1). Echocardiography revealed regional wall abnormalities and cardiac MRI showed ischemic late gadolinium enhancement distribution and significant microvascular occlusion in the LAD territory and vasculitis in the RCA (figure 2). Urgent catheterization confirmed the LAD occlusion (figure 3) and balloon angioplasty was performed successfully. Antithrombotic therapy, a statin, betablockers and ACE inhibitor were commenced post AMI, in addition to steroids for glomerulonephritis with nephrotic range proteinuria. PET-CT was unremarkable for large vessel vasculitis.
Results: Pediatric AMI is rare with an incidence of 6.6 events per 1 million patient-years in teenagers between the ages of 13-18 years. AMI etiologies are diverse and often multi-factorial leading to delayed diagnosis and significant morbidity. While venous thrombosis is a well–known complication of nephrotic syndrome, arterial thrombosis is rare, occurring in 1–5.5% of cases, secondary to the combination of hyperlipidemic and a hypercoagulability state.
Conclusions: Coronary occlusion is a rare complication of nephrotic range proteinuria. A comprehensive cardiac evaluation including coronary angiogram may be useful in cases of underlying nephropathy accompanied with cardiac symptoms.

Keywords: Acute Myocardial Infarction, proteinuria, glomerulonephritis

Figure 1-3

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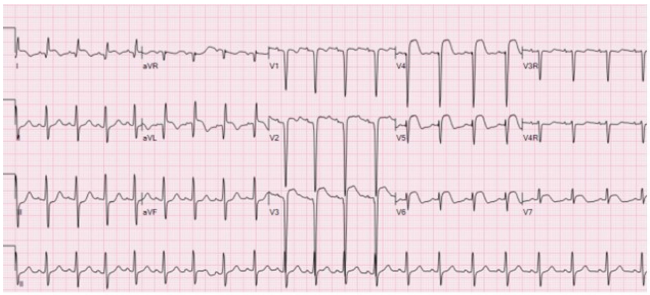

Figure 1 – 15 leads ECG showing Sinus Tachycardia, ST elevation (upsloping) in anterolateral leads, T wave inversion in V2-V6, lead I and aVL. No reciprocal changes, poor R wave progression.

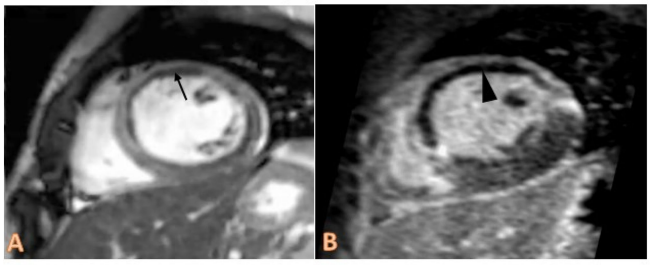

Figure 2 - A set of 2 corresponding short axis images. **(A)**Early (after contrast injection) contrast-enhanced cardiovascular magnetic resonance imaging (MRI) showing a central hypo enhanced area corresponding to microvascular obstruction (MVO) in the anteroseptal region (**arrow**). **(B)**Late gadolinium-enhanced cardiovascular showing hyperenhancement indicating an anteroseptal myocardial infarction **(arrowhead)**.

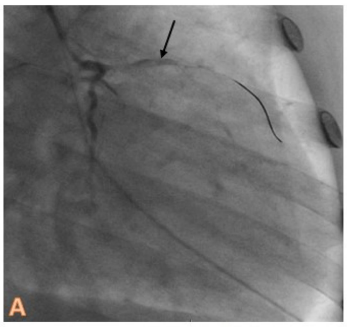

Figure 3A – Selective angiography in the LMCA revealed flow in left circumflex with severely reduced flow in LAD territory (**arrow**).

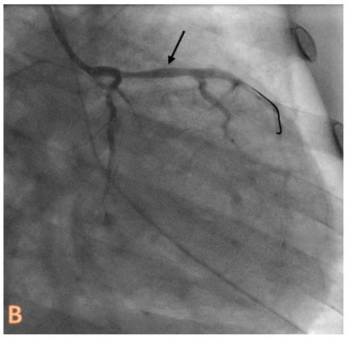

Figure 3B – Selective angiography in the LMCA after balloon angioplasty show significantly improved flow in the LAD territory (**arrow**).

PP-638

Effectiveness of medium-dose intravenous immunoglobulin (1 G/KG) vs high-dose intravenous immunoglobulin (2 G/KG) in the treatment of kawasaki disease

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Background and Aim: Kawasaki disease (KD), the leading cause of acquired heart disease in children, is an acute childhood systemic vasculitis. It is associated with coronary artery aneurysms (CAA), that could be prevented by intravenous immunoglobulin (IVIG) administration. High-dose IVIG (2 g/kg) is usually given in the treatment of Kawasaki disease (KD). However, medium-dose IVIG (1 g/kg) is a low-cost treatment and may have the same efficacy. We aim to determine whether the treatment with IVIG at an initial dose of 1 g/kg is effective for preventing CAA.
Method: A multicenter retrospective cohort study was conducted. A total of 507 patients with complete KD who were treated with high-dose and medium-dose immunoglobulin at Cipto Mangunkusumo Hospital and Kawasaki Center, Indonesia from January 2012 to January 2022 were enrolled. Patients treated with a single infusion of medium-dose IVIG (1 g/kg) were defined as group A, and patients treated with high-dose IVIG (2 g/kg) were defined as group B. Patient characteristics were compared between the two groups; demographic features, laboratory findings, mucocutaneous involvement, day of fever, duration of fever after treatment, length of stay, and rates of CAA from echocardiography during the follow-up period.
Results: Medium-dose IVIG was given in 24 patients (group A). High-dose IVIG was given in 483 patients (group B). Age and gender distributions, white blood cell and platelet counts, day of fever when IVIG was administered, duration of fever after IVIG treatment, and length of stay did not differ significantly between the two groups ($p > 0.05$). All patients had mucocutaneous involvement. Median of weight for age (WAZ) was higher in group A (+0,35 vs -0,26; $p < 0.05$). Median of concentrations of Creactive protein was higher in group B (59,5 mg/L vs 81 mg/L; $p < 0.05$). Coronary artery aneurysms were not found in group A and in 9 patients (1.9%) in group B during the follow-up period ($p > 0.05$).
Conclusions: Treatment of KD with IVIG at an initial dose of 1 g/kg could show the same effectiveness as the high-dose IVIG (2 g/kg) and might be an option for low- and middle-income country.

Keywords: Intravenous immunoglobulin, Kawasaki disease

Clinical outcomes of Kawasaki disease to IVIG doses

Clinical outcome	IVIG dose		P value
	1 g/kg (n=24)	2 g/kg (n=483)	
Coronary artery aneurysms (CAA)			
Yes	0 (0%)	9 (1,9%)	1,000 *
No	24 (100,0%)	474 (98,1%)	
Duration of fever (after IVIG treatment)			
<24 hours	22 (91,7 %)	473 (97,9%)	0,289
<48 hours	1 (4,2%)	5 (1%)	Ref
No fever	1 (4,2%)	5 (1%)	0,096
Length of stay			
Length of stay (days)	4 (2-10)	5 (1-16)	0,198

*Fisher's exact test

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PP-639

Unmasking macrotroponin: A diagnostic challenge in pediatric myocarditis evaluation

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Background and Aim: Troponin positivity in myocarditis diagnosis is commonly assessed using highly sensitive cardiac troponin T (hs-cTnT) tests. However, discrepancies between clinical, electrocardiographic, and echocardiographic evaluations alongside positive troponin results prompt investigation into potential causes of false troponin positivity.
Method: A seven-year-old male presented with respiratory distress and chest pain. Elevated hs-cTnT levels (0.069 ng/ml) and cardiac MRI revealed apical oedema and T2 signal enhancement, leading to a myocarditis diagnosis. Upon clinical improvement and negative cardiac troponin T, follow-up displayed persistently borderline troponin T levels alongside normal electrocardiographic and echocardiographic findings. Further assessment revealed persistent troponin T positivity despite negative troponin I, suggestive of macrotroponin presence confirmed by recovery rates after the study.
Results: Hs-cTnT serves as a crucial diagnostic tool in suspected myocarditis cases; however, discordance among evaluations requires investigation into potential false-positive causes. Excluding anatomical pathology through cardiac MRI, and discerning macrotroponin presence becomes pivotal when cardiac troponin T is positive while troponin I remains negative. Recognizing macrotroponin's influence in cases with high troponin levels incongruent with clinical symptoms is imperative to prevent unnecessary hospitalizations and treatment expenses
Conclusions: In cases exhibiting discrepancies between clinical evaluations and cardiac troponin levels, considering the presence of macrotroponin is pivotal for accurate diagnosis and management decisions, thus avoiding unnecessary healthcare burdens.

Keywords: Pediatric Myocarditis, Cardiac Troponin, Macrotroponin, Diagnostic Challenges, Troponin Positivity,

PP-640

Atypical clinical manifestations in kawasaki disease: Implications for diagnosis and treatment delays

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Background and Aim: The presence of symptoms not included in the diagnostic criteria of Kawasaki Disease (KD) may pose challenges in both diagnosing the condition and initiating treatment. Our study aims to evaluate the incidence, clinical characteristics, and impact on the diagnostic and treatment process of different clinical findings not included in the diagnostic criteria.
Method: Between 2005 and 2019, cases diagnosed with KD were retrospectively examined. Both complete and incomplete cases were encompassed in the study. Symptoms and manifestations beyond the classical features were categorized according to organ systems. Furthermore, the diagnosis dates, initiation of treatment, coronary involvement, and rates of treatment resistance among patients were also evaluated.
Results: A total of 161 patients were included. The mean age was 42.69 months (3 months to 14 years). Patients included 48.1% (n:78) with incomplete KD and 51.2% (n:83) with complete KD. Coronary artery involvement was present in 28% of the patients. The rate of resistance to IVIG treatment was 17.3%. Among the total of 161 patients, 42 (26.1%) exhibited clinical findings different from the classic symptoms. Gastrointestinal involvement was the most common at 40% (n:17), followed by joint involvement at 17% (n:7), pulmonary involvement at 12% (n:5), and an equal number of genitourinary and neurological involvements at 9.5% (n:4). Hematological system involvement was observed in 5% (n:2), and Kawasaki Shock Syndrome in 7% (n:3). It was found that cases with different clinical findings received a statistically significant delayed diagnosis compared to those without such findings ($p=0.000$). Additionally, these cases also had lower albumin levels compared to those without different clinical findings ($p=0.018$). However, no statistical difference was observed between the presence of different clinical findings and the development of coronary artery aneurysms ($p>0.05$), nor between the presence of different clinical findings and IVIG resistance ($p>0.05$).
Conclusions: Patients with different clinical findings experienced a significant delay in receiving a diagnosis and consequently initiating treatment. However, there was no identified association between delayed diagnosis and the development of coronary artery abnormalities. This suggests that despite delays in diagnosis potentially postponing the start of treatment, administering IVIG might still mitigate coronary artery involvement.

Keywords: atypical findings, kawasaki disease, diagnosis

PP-641

Simultaneous assessment of renal angina index and n-KDIGO staging for detecting neonatal acute kidney injury post-contrast exposure

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Background and Aim: Newborns are at higher risk of developing acute kidney injury. As a major risk factor for acute kidney injury, the increasing use of contrast agents for diagnostic and interventional procedures has made contrast-induced acute kidney injury the third most common etiology of hospital-acquired acute kidney injury.
Method: Renal Angina Index (RAI) evaluation was performed in all patients before cardiac catheterization angiography with contrast material was performed in the neonatal intensive care unit. The lowest creatinine value before the procedure and the highest value after the procedure were used in the staging of Neonatal Kidney Disease: Improving Global Outcomes (n-KDIGO). Acute kidney injury was defined as an RAI of 8 or higher and n-KDIGO stages 2 and 3.
Results: Based on early data (day 1), positive RAI values were 12.1% (n=8), while they were 10.4% (n=5) based on late data

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(day 7). However, none of our patients exhibited acute kidney injury according to the n-KDIGO staging system.
Conclusions: Our findings suggest that RAI scoring may have detected acute kidney injury that n-KDIGO staging failed to detect, and that n-KDIGO or RAI alone may not detect all neonatal infants with early-stage kidney injury. Therefore, the simultaneous evaluation of both RAI and n-KDIGO staging in neonates exposed to contrast agents during cardiac catheterization angiography or other procedures may enable the detection of acute kidney injury that could otherwise be overlooked.

Keywords: Congenital heart defect, acut kidney injury, newborn, contrast agent

PP-642

The assessment of the impact of physical exercise level on the quantity of endothelial cell precursor progenitor cells in peripheral blood in patients

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Background and Aim: At the present stage, there are discussions about the predictive value of the level of progenitor cells of endothelial cell precursors (PCE) in peripheral blood for cardiovascular complications in high-risk patients, particularly after acute myocardial infarction. Since endothelial dysfunction is one of the initiating mechanisms of myocardial infarction, it is important to assess the risks of recurrent cardiovascular events in patients during cardiac rehabilitation with increased physical exercise.
OBJECTIVE: To identify the correlation between the restoration of physical exercise tolerance (PET) level and the quantity of PCE in peripheral blood of patients after acute coronary syndrome (ACS).
Method: Ninety male patients were examined, including 79 (87.8 ±6.8%) with a diagnosis of myocardial infarction, of which 65 (82.3±8.4%) had STEMI and 14 (17.7±8.4%) had NSTEMI, and 11 (12±6.8%) with unstable angina. The average age of patients was (56.5±10.5) years. Coronary angiography revealed atherosclerotic involvement of one vessel in 23 patients (25.6 ±9.0%), two vessels in 26 (28.9±9.4%), and three vessels in 41 (45.6±10.3%). Hemodynamically significant three-vessel involvement was found in 24 patients (26.7±9.1%), two-vessel involvement in 31 (34.4±9.8%), and one-vessel involvement in 35 (38.9±10.1%). All patients underwent exercise testing using the graded exercise method, assessment of intracardiac hemodynamics using two-dimensional and Doppler echocardiography, coronary angiography, biochemical and general clinical blood tests, and quantification of PCE levels in peripheral blood.
Results: The obtained data indicate that there is an associative relationship between better restoration of PET levels after acute coronary syndrome and the restoration of endothelial function in patients with a favorable course of the post-infarction period during repeated examination, compared to patients whose PET levels did not exceed 50 watts (low PET). This is evidenced by a statistically significant higher number of PCE after physical exercise in the group with a favorable course and high PET (Group A) compared to the group with an unfavorable course and low PET (Group B).
Conclusions: Studying the response in terms of changes in PCE quantity in response to physical exercise can help determine the progression of systemic endothelial dysfunction, which affects the patient's prognosis. Decreased PCE during physical exercise.

Keywords: physical exercise, progenitor cells, acute coronary syndrome, cardiac rehabilitation

PP-643

No recent improvement in the first-year survival of infants with congenital heart defect: A population-based french registry study from 1994 to 2021

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Background and Aim: Survival in congenital heart defect (CHD) depends on medical and surgical advances and the organization of the healthcare system. In order to identify potential interventions to improve outcomes, it is important to have current data on the management of these children. To evaluate trends in 1-year survival in CHD, using a contemporary birth registry study.
Method: The study population consisted of 3,781 live births with CHD. We retrieved CHD livebirths spanning from 1994 to 2020 from the Paris Registry of Congenital Malformations (Remapar), for women living and delivered in Paris. We estimated and compared survival probabilities at 1-year of age, overall and by CHD subgroups subtype, in successive 4 periods using the Kaplan-Meier method and log-rank tests.
Results: The overall survival of infants with CHD at 1-year of age was 91.4% (95%CI: 90.4-92.2). Survival was higher for isolated CHD defined as those without any genetic anomalies/malformations in other systems 94.6% (95% CI: 93.7-95.3), compared to CHD with malformations of other systems 83.6% (95% CI: 79.5-87.0) and those with genetic anomalies 73.0% (95%CI 68.2-77.3). Survival has increased between periods 1994-2000 and 2001-2007, overall and for isolated CHD, respectively from 87.4% to 91.7% ($p<0.01$) and from 90.8% to 95.3% ($p<0.01$), and then remained unchanged (Figure). Improved survival is mainly driven by isolated neonatally operated CHD. For both infants with CHD and other malformations and those associated with genetic anomalies, survival did not increase over the study period.
Conclusions: After an increase until the beginning of the 21st century, first-year survival has not changed in the last 15 years, either overall or for isolated CHD.

Keywords: congenital heart defect, survival, registry, population-based, infant mortality

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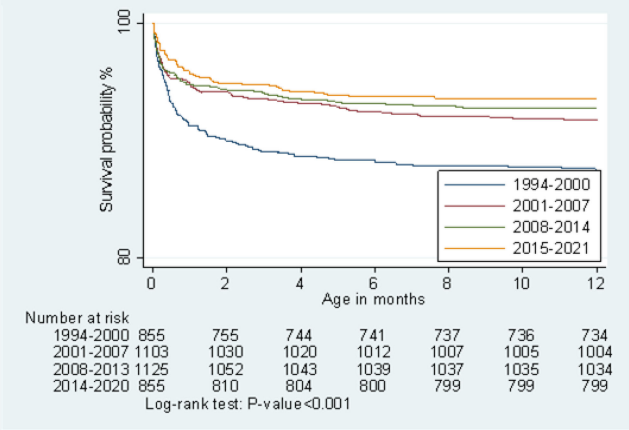
Figure: Survival curves at 1-year of age for infants with CHD according to time period



PP-644

Double aortic arch presenting as neonatal respiratory failure and stridor: A case report

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Background and Aim: DAA malformation is rare congenital heart disease that may cause tracheal and esophageal compression. It's the most common type of vascular ring malformation. However, respiratory failure owing to critical airway obstruction in the neonatal period is rare with few reported cases in the literature

Method: We report a case of a newborn patient with a double aortic arch who presented with stridor and Respiratory distress since birth.

Results: A female newborn with no family history, was born via spontaneous vaginal delivery at 40 week's gestation, to a 31-year-old-mother with a poorly monitored pregnancy. At birth, she was admitted to the Neonatal intensive care unit for immediate severe respiratory distress with biphasic stridor. On physical examination, the newborn was eutrophic with a birth weight of 3600g. She was cyanotic, polypneic with supra-sternal and sub-costal recessions. Inspiratory and expiratory stridor was noted which did not improve with positioning. A cardiac auscultation revealed normal heart sounds. Cardiac ultrasound revealed a double aortic arch and a severe pulmonary hypertension. The computed tomography angiography of the chest confirmed the diagnosis of DAA with circumferential compression of the trachea and esophagus. The infant required emergent intubation within the first few hours of life. But due to severe hypoxemia, the newborn succumbed before the surgical intervention.

Conclusions: We report on a case of double aortic arch with neonatal revelation, the clinical severity of the anomaly. So, Clinicians should consider the possibility of a congenital vascular ring.

Keywords: double aortic arcch, neonatal, vascular ring

PP-647

Cardiac fibromas in small children: 6-year experience of management

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Background and Aim: Cardiac fibromas are rare tumors, mainly in young children. There is no consensus to indications for surgical resection of fibroma. We summarize the single center experience to compare the results of surgical treatment and natural course of cardiac fibromas in pediatric population.

Method: A retrospective study of 12 children with cardiac fibromas diagnosed in our center from 2014 to 2019 was analyzed. Patients were divided into 2 groups: the group I (n=6), who underwent surgical tumor resection, and the group II (n=6), who did not receive surgical treatment.

Results: The mean age of patients at the time of initial diagnosis was 2.7 years (range 0-8.6 years) and did not differ between groups. The localization of fibromas was as follows: 5 (41.7%) cases in the left ventricle, 5 (41.7%) – in the right ventricle and 2 (16.7%) – in the right atrium. In the group I, the mean age at the time of surgery was 2.5 years (4 days - 5.8 years). In all cases, surgical resection was performed within 3 months of diagnosis. All patients survived. In one case (16.7%), a small ventricular septal defect was formed during complete resection of a giant fibroma. During the postoperative follow-up, which was 4.1 years (2.8-5.3 years), tumor recurrence was documented in two cases (33.3%). In particular, one of them required re-excision of the right atrial tumor with a good long-term result. In the group II, the follow-up period was 5.1 years (3.1-8.9 years). None of the patients had worsening of clinical symptoms. In 3 patients (50.0%), regression of the relative tumor size was observed. The mean indexed area of the tumor during the observation period did not undergo significant changes from the first examination (0.933±0.837 cm2/m2) to the last (0.976±0.681 cm2/m2).

Conclusions: Surgical resection of cardiac fibromas has an increased risk of recurrence and complications, which requires a balanced approach to determine the indications for resection. Observation of the clinical course and fibroma growth can be useful in clarifying the need for surgical treatment.

Keywords: Cardiac fibroma, surgical resection, natural course

PP-648

Umbilical venous catheter placement in isomerism – the importance of considering the venous anatomy

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Background and Aim: Isomerism is associated with complex cardiac anatomy including abnormal systemic venous connections. Although insertion of umbilical vein catheter (UVC) is a standard of care in neonatal period, its use can cause serious complications. We present a rare case of intraperitoneal extravasation of total parental nutrition (TPN) in a neonate with left atrial isomerism secondary to UVC malposition in left portal vein.

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Method: Retrospective review of electronic medical records was performed.
Results: A 35-year-old woman, gravida 1 was referred to our fetal cardiology clinic at 25+6 weeks of gestation due to suspected heart defect. Fetal echocardiogram showed left atrial isomerism with complete balanced atrioventricular septal defect, aorta arising anteriorly from the right ventricle and pulmonary atresia with right aortic arch. There was interruption of inferior caval vein with azygous continuation to left-sided superior caval vein draining to left-sided atrium. Additional right-sided caval vein and hepatic veins drained to right-sided atrium while pulmonary veins to left-sided atrium. Throughout pregnancy, these findings were unchanged with normal rhythm. A female infant was born at 39 weeks of gestation via emergency c-section due to failed induction weighing 3.5 kg. She was started on Prostaglandin infusion and an umbilical venous line was placed to commence TPN as per our policy for ductus dependent lesions. Although UVC was not in the ideal position, patient was thought to be at higher risk to alternate central venous access under general anaesthesia. A postnatal echocardiogram and computed tomography confirmed prenatal diagnosis. Due to the tortuosity of ductus arteriosus, a surgical placement of a restrictive right to pulmonary artery conduit was decided as the most appropriate and long-lasting initial palliation. On day 6 of life, patient presented with abdominal distention and low-grade temperature with mildly increased inflammation markers and transaminases. An ultrasound confirmed our suspicion of TPN extravasation. UVC was replaced by right femoral venous line and peritoneal drain was placed for 3 days to relieve 200 mls of TPN. Patient was successfully recovered and fit for surgery after 6 days.
Conclusions: UVC should not be electively inserted in neonates with left isomerism and interrupted IVC due to risk of malposition.

Keywords: isomerism, abnormal systemic venous connection, umbilical venous catheter malposition

PP-649
The effect of transcatheter asd closure on the frequency of headache in the childhood
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Background and Aim: A close relationship has been found between congenital heart disease and migraine headaches. We planned to determine the relationship between headache and migraine clinics in patients with atrial septal defect (ASD) in the childhood group and to evaluate the change after transcatheter closure.
Method: 82 patients who underwent transcatheter closure with the diagnosis of secundum ASD in our clinic were included, aged 9.2 ±3 (5-16) years, 47 F, defect diameter 15.8±5.3 (6-29) mm, device diameter 17.5±6 (7-30) mm. The devices used were Occlutech (35 pts, 42.7%), Amplatzer (30 pts, 36.6%), Lifetech (17 pts, 20.7%) atrial septal occluder. The study was conducted prospectively by filling out a survey form prepared according to ICHD-3 (International Classification of Headache Disorders) criteria. The survey was conducted by contacting the patient and their relatives before and at the 3rd, 6th, and 9th months after the repair procedure. 20 patients with headache before closure were compared with 62 patients without.
Results: In patients with pain, no significant difference was found between the age of onset of pain, gender, weight, defect diameter, mean pulmonary artery pressure, shunt rate and device diameter ($p>0.05$). While 20 (24.3%) of the patients had recurrent headaches before the procedure, 7 patients (8.5%), one of whom had a new onset in the 3rd month, 4 (4.8%) in the 6th month, and 5 (6%) patients in the 9th month, had headaches. A significant difference was found between the initial headache and the presence of pain in the 3rd month ($p=0.002$). There was no statistically significant difference between the device brand used and the presence of headache in the 3rd month ($p>0.05$). There was a decrease in the frequency of headaches that continued in the 6th and 9th months.
Conclusions: The decrease in the frequency of headache and migraine attacks after transcatheter closure in patients with left-to-right shunt is important as it indicates microemboli. However, the presence of opposing study findings shows that disease with left-right shunt, migraine pathogenesis and device technical features have a multifactorial interaction and that dual antiplatelet therapy in children may be required in selected cases.

Keywords: transcatheter ASD closure, migraine, childhood

PP-651
A single centre experience of late presentations of unrepaired cyanotic congenital heart disease
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Alder Hey Children's Hospital, Liverpool, United Kingdom

Background and Aim: It is rare to see unoperated cyanotic congenital heart disease (CHD) in older children born in the UK. There are now increasing cases of refugee and migrant children in the UK presenting with cyanotic CHD who have not previously undergone any intervention or surgery in earlier life. The management of these children is challenging with lack of published evidence for late surgical options hence it can be difficult to prognosticate (Table 1). This study describes our centres experience of presentation and management of older children with unrepaired cyanotic CHD.
Method: We reviewed MDT discussions over a 2 year period to identify cases of older children with cyanotic CHD who had not undergone any surgical or interventional treatment.
Results: Over the 2 years period 3 cases were identified. These included 2 children, aged 5 and 3 years, with unoperated Tetralogy Of Fallot (TOF) and a 14 year old with Single ventricle pathology (Double Outlet Right Ventricle, Transposition of the Great Arteries and Pulmonary Stenosis (DORV, TGA, PS)). The children with TOF both presented with profound spells within days of arriving in the UK. Clinical signs of cyanosis and clubbing were evident. They were both polycythaemia and one had had a previous cerebro-vascular accident (CVA). The patient with DORV, TGA, PS presented with cyanosis and extremely poor exercise tolerance. He was wheelchair bound, with clubbing and hypertrophic osteopathy of his knee joints. In these cases, MDT discussions and planning lead to staged surgeries to achieve complete repair for the children with TOF and high-risk late Fontan completion for the child with DORV, TGA, PS. After completion of the Fontan the child's exercise tolerance improved markedly and the hypertrophic osteopathy resolved.

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Conclusions: With increasing numbers of migrant children in the UK late presentations of cyanotic CHD may become more common. Collating information on management strategies and outcomes after late interventions will allow for evidence-based decision making for future cases.

Keywords: fontan, migrant, unrepaired, interventions, cyanotic

Table 1

Author	Study	Main conclusion
Fuchigami T et Al	**Long-term follow-up of Fontan completion in adults and adolescents** Retrospective cohort study of 79 patients undergoing Fontan ≥15 years old.	In patients who were ≥15 years old, the surgical results of the Fontan operation were acceptable. Approximately half of the late deaths were sudden deaths, mainly occurring 10-20 years postoperatively.
Parvathi U Iyer et Al	**Management of late presentation congenital heart disease.** Literature review	Surgery associated with adverse long-term outcomes. Recommends conservative approach when in the presence of pulmonary hypertension. Severe low cardiac output state and Pulmonary hypertension complicate post op. Many practices will be validated only by midterm or long-term follow-up of survivors.

Table with the limited published evidence of late surgical outcomes

PP-652
Cardiovascular health and vitamin d in healthy, preterm born young adults

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Background and Aim: Prematurity has been associated with higher risk for cardiovascular diseases (CVD) later in life. The mechanisms to cause CVD in adulthood after preterm birth seems to be multifactorial. In 1990's Vitamin D (Vit D) levels were not routinely monitored. Also low Vit D levels has been associated with higher risk of CVD and mortality. Our aim is to study the cardiovascular health after preterm birth in young adults.
Method: We studied preterm born population (PT) originated from infants born in 1994-1997 Oulu University Hospital. Inclusion criteria (PT) were: 1) Healthy young adults born before 34 gestation weeks with very low birth weight (<1500g) and 2) all subjects born under 32 gestation weeks. 38 controls born full-term (FT) were age and gender matched. Their medical records were monitored, but also subjects filled wide health enquiry. All subjects participated for cardio-respiratory and laboratory tests.
Results: Out of 38 participants 22 were females and 16 males in both groups. The PT participants had higher mean SBP (p=0.01) values in the beginning of the study. At baseline 76% of PT and 74 % FT had appropriate Vit D level (>50 nmol/l). Echo parameters in right or left ventricular (LV) systolic function were normal without differences. LV diastolic E/A ratios differentiated only between malegroups (1.60 vs 1.90, p = < 0.001). LV diastolic values were in normal range in all participants in the study. Still statistical difference was seen at baseline between groups (p=0.006 and 0.015). After a year PT participants got higher mean values for E' lat (from 0.16 to 0.17 m/s, p = 0.011) and E'med (from 0.14 to 0.15 m/s, p = 0.002), respectively. Also, E/E'lat ratios were smaller in control group (p = 0.040), and after a year E/E' ratio decreased in PT participants (p = 0.029).
Conclusions: Our study showed that vitamin D defiency is still found from remarkable part of population (24/26%) of young healthy adults in Finland. Also, we saw early hints of diastolic and blood pressure mechanisms in PT group behind the higher risk for cardiovascular diseases later in life compared to FT born adults.

Keywords: Preterm born adult, Cardiovascular health, Vitamin D

PP-654
Elevation of troponin i among children reporting to the pediatric emergency department with chest pain

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Background and Aim: Chest pain is a frequent cause for admission to the pediatric emergency department (ED). Although the causes are primarily benign, they may be associated with more severe conditions, including cardiac causes. Therefore, myocardial enzyme testing is used to evaluate the likelihood of cardiac involvement. In this study, we examined the proportion of positive troponin I tests among patients reporting to the pediatric ED.
Method: A retrospective analysis of patients aged <18 reporting to the pediatric ED of the Children's Hospital with chest pain from 26.10.2022 to 26.10.2023 was performed. The database was searched for ICD-10 codes: R07.2 (Precordial pain), R07.3 (Other chest pain), R07.4 (Chest pain, unspecified), I26 (Pulmonary embolism), I40 (Active myocarditis), I51.4 (Myocarditis, unspecified).
Results: 255 patients aged 2 to 17,93 (average 12,4, median 13,3) were analyzed. There were 131 males (51.4%) and 124 females (48.6%) with a total of 262 visits. Among these patients, troponin I assay was performed in 191 (72,9%) cases. 71 patients without troponin assessment were excluded from the study. Troponin I levels were elevated (>26.2 ng/l) in 14 (7,33%) cases. Acute myocarditis was identified as the cause of elevated troponin in 9 cases, 1 case was attributed to an event of supraventricular tachycardia, 1 was related to an episode of supraventricular tachycardia in a patient with concomitant coronary artery fistula, 1 assay was performed in a patient after cardiac transplantation and in 2 cases the cause of chest pain was unknown.
Conclusions: The serum level of troponin I remains a valuable diagnostic tool in the management of children presenting to the pediatric ED with chest pain. An elevated troponin may indicate serious cardiac causes of chest pain that require further cardiological investigation. Inflammatory heart diseases are the main cause of elevated troponin among children reporting to the ED.

Keywords: chest pain, troponin, pediatric emergency department

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PP-655

Electrocardiographic characteristics in children with cerebral palsy

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Background and Aim: Cerebral palsy (CP) is a motor impairment resulting from a brain injury that occurs in the perinatal period and causes activity limitations. Only a few studies have been performed to compare electrocardiograph (ECG) patterns between children with CP and healthy controls. Until a recent trial, there is no evidence of a relationship between the ECG patterns and the impairment level among children with CP. This study aimed to investigate the association between ECG characteristics and functional impairment in CP population.
Method: Forty-one patients with CP aged 1,5-15,5 years were included and graded in five levels of Gross Motor Function Classification (GMFCS) described by Palisano et al. (1997). The following ECG variables were analyzed: heart rate (HR), QRS duration, P duration, PR interval, QT interval, mean corrected QT intervals (QTc), P-wave axis, and QRS-axis.
Results: The pooled ECG parameters for each GMFCS level and between-group post hoc analysis of variance results are presented in Table. Additionally, data of children classified as GMFCS level I-III and children classified as GMFCS level IV-V were analyzed. They did not differ according to age, gestational age, and weight. GMFCS I-III patients compared to GMFCS IV-V presented significantly lower HR (96 ± 11 bpm vs. 122 ± 8 bpm; $p<0.001$), shorter duration of P-wave (82,1 ± 9,4 ms vs. 94,2 ±16,3 ms; $p<0.01$), leftward P axis (50 ± 27° vs 69 ± 12°, $p<0.05$).
Conclusions: In children with CP, HR, P-wave axis, and duration alter regarding the severity of the disease. The clinical value of these findings needs further evaluation.

Keywords: cerebral palcy, ECG, P-wave.

Table

	All (n=41)	GMFCS I (n=6)	GMFCS II (n=15)	GMFCS III (n=8)	GMFCS IV (n=4)	GMFCS V (n=8)	p
Age [years, months]	5,4 (3,3)	8,6 (2,4)	5,4 (2,3)	3,4 (1,2)	3,4 (1,2)	6,4 (5,2)	
Pattern h/d/q	10/19/12	1/3/2	8/6/1	1/6/1	0/2/2	0/2/6	
HR [beats per min]	104 (15)	82 (7)	96 (7) †	106 (5) ††††	124 (3) ††††††^	121 (9) ††††††^^	<0.001 *
QRS duration [ms]	82 (8)	88 (5)	85 (9)	79 (4)	77 (10)	79 (8)	NS *
PR interval [ms]	123 (20)	142 (18)	122 (15)	115 (16)	138 (33)	113(14)	<0.05 *
P duration [ms]	86 (13)	89 (5)	82 (9)	76 (10)	105 (24) ‡^^	89 (9)	<0.01 *
QT interval [ms]	333 (23)	357 (18)	342 (18)	330 (14)	321 (19) †	308 (14) †††††	<0.001 *
QTc interval [ms]	434 (24)	445 (15)	429 (22)	428 (26)	461 (32)	429 (20)	NS #
P axis [°]	55 (25)	66 (9)	50 (22)	37 (37)	71 (16)	67 (10)	<0.05 #
QRS axis [°]	75 (27)	68 (42)	72 (20)	77 (36)	82 (13)	79 (27)	NS #

Mean (SD); * - ANOVA; # - Kruskal-Wallis; post hoc test: † vs. GMFCS I; ‡ vs. GMFCS II; ^ vs. GMFCS III; ǂ vs. GMFCS IV; h – hemiplegia, d – diplegia, q - quadriplegia

ECG parameters analysis in children with CP for each GMFCS level

PP-656

Significance of colonization by antimicrobial resistant organisms prior to congenital heart disease surgery in children from low-income countries

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Background and Aim: Children with congenital heart disease (CHD) from low-to-middle income countries (LMIC) are suspected to have a high prevalence of antimicrobial resistance microorganisms (ARMOs) carriage but data are currently lacking. Carriage of ARMOs could impact the post-operative course in pediatric intensive care unit (PICU). The aAim of the study was to assess the prevalence and the impact on post-operative outcomes of ARMOs carriage in children with CHD from LMIC with a CHDand its impact on post-operative outcomes.
Method: Retrospective monocentric cohort study from 01/2019 to 12/2022. Included patients were children (0-18 years) admitted from a LMIC admitted after a CHD surgery and anwith AMRO screening performed the week before. Infectious episodes and post-operative evolution were compared based on ARMOs carriage status.
Results: Among 224 surgeries (222 patients), with a median age of 38.5 months (IQR 22-85.5) at admission, ARMOs carriage was evidenced in 95 episodes (42.4%). Main organisms isolated were Extended Spectrum Beta-Lactamase (ESBL) producing E. coli (75 (33.5%) and ESBL-K. pneumoniae (30 (13.4%)). Median mechanical ventilation duration was 1 day (IQR 0-1), PICU stay 3 days (IQR 2-4) and hospital stay 6.5 days (IQR 5-10). A total of 17 infectious episodes occurred in 15 patients, mostly consisting in hospital-acquired pneumonia (HAP) (12/17). Only two infections were caused by a colonizing ARMO. Occurrence of infection was similar between carriers and non-carriers. Carrier status was not associated with a prolonged duration of mechanical ventilation, PICU or hospital stay. Use of vancomycin and carbapenems was higher in case of ARMOs carriage. Negative swab screening may lead to sparing most of empirical vancomycin therapy (11/12) for HAP based on current guidelines.
Conclusions: Prevalence of AMROs carriage is high in children from LMIC and has a limited impact on patients' outcome. However, ARMOs carriage leads to higher consumption of antimicrobial agents. Screening may help saving usage of broad-spectrum antibiotic in case of negative swab.

Keywords: antimicrobial resistant organisms, congenital heart disease, cardiac surgery, children

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PP-657

Clinical presentation and outcome of pediatric infective endocarditis

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Background and Aim: Infective endocarditis (IE) is an infrequent disease in childhood and most often affects children with congenital heart defects (CHD); in rare cases, the disease can also develop in patients with structurally normal hearts. Our study aims to assess the features of IE in children with CHD compared to those without CHD.
Method: Patients aged <18 years with CHD and structurally normal hearts treated for IE at the National Heart Hospital, Sofia, Bulgaria, for 14 years (01.01.2009-31.12.2022) were included. The diagnosis of IE was based on the modified Duke criteria. We assessed the demographics, clinical characteristics, laboratory results, microbiology, localization of the vegetations, therapeutic approach, and outcome of the patients.
Results: We identified 26 children (mean age 8.1 years (12 days - 17 years, SD 5,5)), 20 of them (72%) with underlying CHD, and 6 (28%) with structurally normal hearts. The mean age of the patients with CHD (group 1, n=20) was 9.12 years, and of the non-CHD (group 2, n=6) – 3.5 years (p=0.02). The leading pathogens by group 1 were Gram-negative bacteria (30%) and by group 2 - S.aureus (33.33%) (p=0.52). The infection was community-acquired in 85% of the patients in group 1 and in 50% of those in group 2 - hospital-acquired (p=0.21). The vegetations were localized in the left heart in 50% vs. 83.33% of the patients, respectively (p=0.23). The symptoms were as follows: fatigue (95% vs.50%, p=0.11); fever (90 vs 83%, p=0.83); heart failure (40% vs 33%, p=0.83), and neurological manifestations (50% vs 20%, p=0.23). Valvular replacement was needed in 20% of the children in group 1 compared to 33.3% of those in group 2 (p=0.64). Mortality was 10 vs. 16% respectively (p=0.83).
Conclusions: Even though the only statistically significant difference between children with CHD and structurally normal hearts is age, in patients with CHD there is a tendency for subacute clinical presentation and more community-acquired infections, whereas non-CHD patients tend to have an acute presentation, more frequent neurological symptoms, hospital-acquired infections, and a greater need for valve replacement. It should be noted that the number of our cases is too small to draw firm conclusions.

Keywords: infective endocarditis, congenital heart defects, structurally normal hearts

PP-658

The role of maternal diabetes and overweight in the morbidity of children with congenital heart defects

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Background and Aim: Maternal overweight and diabetes are risk factors for congenital heart defects (CHDs) in the offspring. The aim of this study was to study if the effects of these two risk factors extend beyond pregnancy. We analyzed how maternal diabetes and overweight affect the morbidity in terms of need for hospital care children with CHDs during the first year of life.
Method: This national register-based study included all children born with isolated CHD in Finland between 2006-2016. Data on patient characteristics, CHDs, maternal diabetes diagnoses and maternal pre-pregnancy body mass index (BMI) were collected from nationwide registries: the Medical Birth Register, the Register on Congenital Malformations and the Register on Reimbursed Drug Purchases maintained by the Social Insurance Institute of Finland. Morbidity was defined as the number of hospitalizations, outpatient visits, emergency clinic visits and number of inpatient hospitalization days collected from Care Register for Health Care. Poisson regression analyses were used to analyze the association between maternal overweight and diabetes and the morbidity of offspring.
Results: A total of 10 254 children (52% male) were included. Of their mothers, 15% had gestational diabetes (GDM), 2.2% had type I diabetes (T1DM), 0.32% had type II diabetes (T2DM), 21% were overweight and 13% were obese. GDM, T1DM and T2DM increased risk for hospitalizations, and outpatient visits when compared to mothers without diabetes. Only GDM (RR 1.20 (95% CI 1.11-1.29)) was associated with significant risk for emergency visits. Maternal overweight and obesity increased risk for emergency visits, hospitalizations, and outpatient visits. Maternal overweight (RR 1.14 (95% CI 1.13-1.16)), obesity (RR 1.50 (95% CI 1.48-1.52)) and all types of DM (GDM (RR 1.28 (95% CI 1.26-1.29)), T1DM (RR 1.68 (95% CI 1.63-1.72), T2DM (RR 2.49 (95%CI 2.36-2.63)) were significantly associated with more days spent in the hospital during the first year of life when compared to mothers with normal weight and no DM.
Conclusions: In this nationwide Finnish register-based study, we demonstrate that maternal diabetes and overweight/obesity were associated with increased inpatient and outpatient hospital care need in infants with CHD during the first year of life. This is important when determining the follow-up of these children.

Keywords: congenital, heart, defect, diabetes, obesity, morbidity

PP-659

The assessment of depression in children with ventricular extrasystoles and structurally normal hearts

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Background and Aim: Ventricular extrasystoles (VE) in children with structurally normal hearts are quite common finding and considered to be a benign condition. However, the problem of

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arrhythmia existence can be distressing for patients and their representatives. The AIM: to assess the signs of depression in children with structurally normal hearts and VE and to compare the results with the number of VE in 24-hour electrocardiogram (ECG) and left ventricular function.
Method: The inclusion criteria of prospective study in Vilnius University Hospital Santaros Klinikos were ≥5% VE in 24-hour ECG. The exclusion criteria - diagnoses of hemodynamically significant congenital heart diseases, confirmed diagnoses of cardiomyopathies and accessory pathways. The symptoms and depression assessed with modified pediatric PHQ-9 questionnaire. The interpretation: ≤4 - no signs of depression; 5-14 - mild-moderate depression; ≥15 - severe depression. The questionnaires were completed by the patients' representatives to assess their children's complaints. Children ≥12 years old completed the questionnaire themselves. All children underwent 24-hour electrocardiogram (ECG) and echocardiography.
Statistical analysis of the data was performed with R software. Nominal variables tested for normal distribution with Shapiro-Francia test. Nominal values were presented with average and standart deviation. Student's t-test used to compare means between normally distributed nominal variables. Chi-square test used to compare categorical variables. The p value <0,05 was considered statistically significant.
Results: 60 representatives and 31 children ≥12 years old completed the questionnaire. The average age was 12.6±3.5 years; 33 (55%) were girls. Palpitations experienced 26 (43,3%), chest pain 13 (21,7%), exercise intolerance 15 (25%) children. The average number of VE in 24-hour ECG was 12±5,7%. Left ventricular function, assessed by Simpson biplane method, average was 64,8 ±6.4%. The average score of the PHQ-9 test completed by the representatives was 3,6±3,9; by children ≥12 years - 5,5±4.9, with a maximum score of 18 points. There were no significant differences between the numbers of VE, left ventricular function and the questionnaire results.
Conclusions: According to the PHQ-9 questionnaire, 3 patients showed signs of severe depression. There were no significant differences between the numbers of extrasystoles, left ventricular function and questionnaire results.

Keywords: ventricular extrasystoles, children, depression.

PP-660
Left ventricular function in acute lymphoblastic leukaemia survivors

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Background and Aim: The majority of children with acute lymphoblastic leukaemia (ALL) survive and the received chemotherapy may be associated with later organ dysfunction. Our country has joined the ongoing ALL-STAR trial which analyses longterm organ-specific health among ALL survivors who received treatment according to the NOPHO ALL 2008 protocols. The aim of this study is to assess the cardiac function of our national cohort and to corelate the results with received doses of antracyclines.
Method: A prospective study performed between 20th April 2023 and 20th November 2023 in Vilnius University Hospital Santaros Klinikos. The study was Confirmed by Regional Ethics Committee.We included children ALL survivors who received treatment according to NOPHO ALL 2008 protocols. We grouped the patients according to received anthracyclines doses: I group - <100 mg/m^2; II group - 100-250mg/m^2; III group >250 mg/m^2. All patients underwent echocardiography. Left ventricular function assessed with Simpson biplane method; global longitudinal (GLS) strain - by speckle- tracking echocardiography. Statistical analysis performed with R software. Nominal variables tested for normal distribution with Shapiro-Francia test. Nominal values were presented as mean and standard deviation or minimum, maximum, median respectively. Welch test used to compare means between normally distributed nominal variables. Fisher exact test used to compare categorical variables. The p value <0,05 was considered statistically significant.
Results: Twenty-three children (60,9% boys) were included in the study. The median age was 13 years (7-17 years). The median age of ALL diagnosis was 4 (1-9) years, median survival time of 9 (4-13) years. We included eleven patients to I group, 8 - to II group; 4 to III group. All children had normal left ventricular function with median LV EF 62.7 (56-68.5)% and median GLS -22.9 (-30.7 - -16.9)%. We did not find a significant difference between LV EF, GLS and groups of anthracyclines doses.
Conclusions: The evaluation of left ventricular function remains the actual theme for clinicians and needs further research. In our study, we did not find significant differences in left ventricular function and the groups of anthracyclines doses.

Keywords: left ventricular function, acute lymphoblastic leukaemia, children, anthracyclines

PP-661
The impact of trisomy 21 on late surgical management results in patients with common atrioventricular septal defect

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Background and Aim: Atrioventricular septal defect (AVSD) has a strong conidiation with Down syndrome (DS) and occurs in 30-40% of those children. Different early and late outcome of surgery in children with or without 21 trisomy are suggested. Patients after correction suffer from mitral regurgitation (MI) in 21% of them, what is the reason for reoperation, years after. Less often patient require pacemaker or left ventricular outflow tract obstruction (LVOTO) relieve. The aim of the study was to evaluate late outcomes after surgical correction in patients with and without DS.
Method: We analyzed 86 patients with AVSD undergoing surgery in 1996-2022. Patients with unbalanced AVSD were excluded. In analyzed group was 48 males age 1.9 to 27.5 years (median 10.3). Median age at the surgery was 5 months (0,1 – 8 years). In group 68,6% [n=59] of them were diagnosed with DS (33M/26F), others 31,4% [n=27] had normal karyotype (14M/13F). The median follow up time was 8 years (1 to 27 years); median 9.5 in DS group and 8 years in patients without.
Results: All 87 underwent correction. Pulmonary banding was in 9 patients (10,3%) similarly in both groups [10% vs 11.1%] (RR=0.9; p=0.87288). In patients with trisomy - 42% MI was mild, 44% moderate and 14 % severe after surgery. In patients with normal karyotype 37 % mild, 42% moderate and 21% severe – with no significant differences. Eleven patients had insignificant residual ventricular septal defect, 6 among DS and 5 in normal karyotype, five patients had LVOTO. Surgical relief had 2 patients - 1 with DS. Reoperations were required in 11 patients - mitral plasty or replacement and LVOTO relieve. More often in normal

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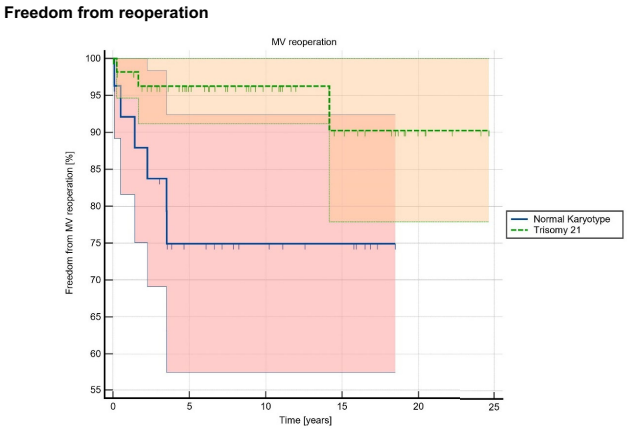
karyotype [6.78% vs 25.93%] (RR=3.82; p=0142). Nine patients underwent mitral valve repair after complete correction, more often without DS [22,22% vs 5.08%] (RR=4.37; p=0.0166). Pacemakers were implanted in 11.63% [n=10] of patients. In 9 cases DS with only 1 with normal karyotype [15.25% vs 3.7%] (RR=4.12; p=0.1231).
Conclusions: Surgical AVSD correction has good long term result. Patients with Down syndrome require more often pacemaker but patients with normal karyotype mitral valve reoperation.

Keywords: atrioventricular septal defect, trisomy 21



Freedom from reoperation in time - Down Syndrome and Non Down Syndrome Patients

PP-662
Anakinra as treatment in an infant with refractory kawasaki disease complicated by multiple giant coronary aneurysms

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Background and Aim: Kawasaki disease (KD) is a rare, acute, inflammatory, multisystemic vasculitis, of unknown etiology that mainly affects children under 5 years of age and is the leading cause of acquired heart disease in developed countries. KD mostly damages medium-sized vessels and have a selective tropism for coronary arteries. The prognosis of KD depends mainly on coronary damage, which with early high-dose immunoglobulins (standard treatment) has been reduced to <5%. But a subset of patients who are resistant to IVIG treatment are at increased risk for coronary artery aneurysms (CAAs), and its optimal treatment remains controversial. KD recognizes IL-1 as the therapeutic target to block systemic symptoms and coronary artery lesions. Therefore, Anakinra, a recombinant IL-1 receptor antagonist, which negatively regulates IL-1-mediated inflammation may be an effective treatment.
Method: We present a previously healthy 12-month-old male, who despite presenting good clinical-analytical progress with timely and aggressive therapy with immunoglobulins, steroids, and aspirin, was complicated by multiple giant CAAs in the outpatient control visit 15 days after the diagnosis. Multiple giant CAAs were located in both proximal tracts of the coronary arteries and were confirmed by CT angiography, as well as the presence of thrombi was excluded. Given the severity of the size of CAAs, with the aim of preventing further cardiac damage, and possibly reversing those already established, 2 weeks after diagnosis we decided to start the subcutaneous administration of anakinra at 2 mg/kg/day together with double antiplatelet therapy (clopidogrel and aspirin), anticoagulation with heparin, without stopping the gradual tapering of oral prednisone. Treatment with anakinra was carried out for 1 month.
Results: The patient was closely monitored by biweekly echocardiography over the following weeks to control the size and possible complications of thrombosis of the aneurysms. In these controls, size stability was observed without associated complications. Three months after treatment, a clear decrease in the size of the aneurysms was observed without observing side effects of anakinra.
Conclusions: In our experience, anakinra was effective in decreasing the size of multiple giant- to medium-sized aneurysms, even when initiated outside of clinically active phase of KD and at the lowest dose described in the literature.

Keywords: kawasaki disease, anakinra, IL-1 receptor antagonist, Coronary artery aneurysms.

PP-663
Effect of home-based exercise training on functional capacity of patients with fontan circulation

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Background and Aim: Patients with Fontan circulation despite systemic and pulmonary circulation continue to have limited functional capacity. A variety of institutional exercise training programs have been shown to improve functional capacity and peak oxygen consumption (VO2 max). A similar non-institutional home-based exercise training (HBET) is more practical if proven useful. Hence, in this prospective observational study, we sought to study the effect of HBET on functional capacity and VO2 max in patients with Fontan circulation.
Method: Patients with Fontan circulation aged 10-18 years with total cavopulmonary connection (Fontan circulation) performed within the preceding 5 years were included. Those with cardiac surgery within the prior 6 months, high-risk or fenestrated Fontan and neuromuscular disability were excluded. A total of 14 patients underwent 6-minute walk distance (6MWD), cardiopulmonary exercise testing (CPET) using modified Bruce protocol and brain natriuretic peptide (BNP) measurement at the baseline and 12 weeks after a pre-defined HBET protocol which included self-paced daily walks, step-up calf exercises and incentive spirometry. The adherence to HBET protocol was ensured remotely with daily updates of physical activity on smartphones, online video sharing and weekly video calls.
Results: Compared to the baseline, following 12 weeks of HBET, the subjects could exercise longer with the mean duration of CPET increased from 12.9 ± 2.2 to 15 ± 1.7 (p=0.0002) achieving higher METS (8.3 ± 0.9 vs 9.42 ± 1.16; p=0.0004). Mean VO2 max increased from 28.4 ± 3.8 ml/kg/min to 32.0 ± 5.2 ml/kg/

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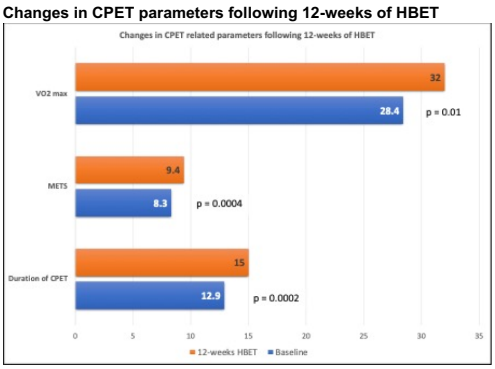
min (p=0.01). The 6-minute walk distance (6MWD) also increased from 380.9 ± 68.5 m to 440.9 ± 44.3 m (p=0.001). Interestingly, BNP levels showed a significant increase from 31.9 ± 47 pg/ml to 45.8 ± 66.3 pg/ml (p=0.014) both the values, however, were within normal range. No patient reported adverse events.
Conclusions: HBET is feasible, safe and effective in improving the functional capacity and VO2 max of patients with Fontan circulation. Larger studies with longer follow-ups are needed to evaluate the long-term benefits of HBET.

Keywords: Fontan circulation, Home-based exercise training, Functional capacity



Following 12 weeks of home-based exercise training (HBET) there was a significant improvement in the duration of exercise, METS achieved and maximal oxygen consumption

PP-664
Genetic profile of children with congenital long qtc syndrome born in northern ireland between 2005-2023

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Background and Aim: Congenital Long QT syndrome (LQTS) is an inherited channelopathy which if not treated can lead to sudden cardiac death. Major advances have been made in the understanding of the genetic basis of LQTS with 17 putative disease-causing genes having now been identifiedi. 75% of LQTS worldwide is caused by three major genes (KCNQ1, KCNH2 and SCN5A). Loss of function genetic mutations cause LQTS in KCNQ1 and KCNH2, gain of function mutations cause SCN5A. KCNE1 has limited evidence of causing LQTS but is strongly associated with acquired Long QT Syndrome (aLQTS).
Method: We conducted a retrospective review analysing the genetic mutations of all children diagnosed with LQTS attending the Inherited Cardiac Conditions Clinic born between 2005 and 2023. Testing for the main genes causing LQTS (KCNQ1, KCNH2, SCN5A, KCNE1 & KCNE2) was undertaken using a commercial gene panel until 2019, thereafter a more extended panel (10 genes) was used.
Results: 192 children with genes causing LQTS & aLQTS were identified between 2005 and 2023. Loci affected included 112 KCNQ1, 36 KCNH2, 13 SCN5A, 29 KCNE1. Two patients had multiple genes affected, which included a case of Jervell Lange Neilson Syndrome (JLS). Two patients had a prolonged Qtc but were negative following a genetic panel. Twenty-one mutations were identified in KCNQ1 patients, fourteen mutations in KCNH2, two in SCN5A and seven in KCNE1. The children with mutations of (KCNQ1, KCNH2 & SCN5A) had evidence of LQTS. Those with KCNE1 had mutations associated with aLQTS, some who had initially been treated with beta blockers were able to have their treatment transitioned to advice of avoiding QT Prolonging medicines.
Conclusions: The genetic profile of children with LQTS in Northern Ireland is broadly similar to that seen worldwide. There are numerous mutations of the three main genes that are associated with the LQTS phenotype. In addition, there are a number of patients at risk of aLQTS due to mutations in KCNE1, who can likely be treated with advice only.

Keywords: Long QTc, Arrythmia, Channelopathy, Genetics

PP-665
Association of heart rate variability and anxiety in children with mitral valve prolapse

Selen Karagözlü[1], Mehmet Gökhan Ramoğlu[2], Safiye Zeynep Tatlı[3], Özlem Bayram[2], Mustafa Yılmaz[2], Batuhan Bakırarar[4], Tayfun Uçar[2] and Hasan Ercan Tutar[2]
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Background and Aim: Mitral valve prolapse (MVP) is usually asymptomatic, but it may be rarely symptomatic. Increased sympathetic activity and decreased vagal tone in MVP patients may lead to impaired heart rate variability and impaired cardiac automaticity. In this study, the relationship between echocardiographic parameters, anxiety and depression status and heart rate variability in children with MVP was prospectively evaluated.
Method: The study group consisted of 64 children (aged 8-18 years) with isolated MVP and the control group consisted of 64 healthy children. Demographic and clinical information of the patients were evaluated. The study group was further classified according to the presence of MVP syndrome. All the cases in this study were evaluated with echocardiography, electrocardiography and revised Children's Anxiety and Depression scale (RCADS- long version), Quality of Life scale (QOL). The study group were also evaluated with 24-hour rhythm Holter for heart rate variability.
Results: The mean age of the study group (54.1% females) was 14.78±2.65 years and the control group (45.9% females) was 13.94±2.09 years. Demographic and clinical information of the patients are shown in image 1. 56.3% of the study group had MVP syndrome.There was no significant difference between the patients with MVP and the control group in QOL, and RCADS. In the MVP group, borderline RCADS T-scores were detected in 4 patients (6.2%) and clinically significant RCADS T-scores which required treatment were detected in 12 patients (18.7%). There was a statistically significant difference between SDNN, rMSSD, pNN50, LF and HF values in children with and without MVP syndrome (respectively, p=0.004; p=0.011; p=0.004; p=0.042; p=0.025). There was a slight positive correlation between the children's QOLs and the LF/HF ratio (p=0.039, r=0.259).
Conclusions: Decrease in heart rate variability is thought to be one of the mechanisms leading to anxiety disorders in patients with heart diseases. As a result, MVP may also be accompanied by significant arrhythmia. Disruption of autonomic balance in patients

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with MVP syndrome may lead to psychiatric disorders such as anxiety and depression but further studies with larger sample are necessary to outline the mechanisms linking heart disease and anxiety disorders.

Keywords: Heart rate variability, Mitral Valve Prolapse, Anxiety

Image 1

Number:64		Frequency (n)	Percentage (%)
Gender	Female	46	71,9
	Male	18	28,1
Height Percentile	< 3	5	7.8
	3-10	1	1.5
	10-25	3	4.6
	25-50	12	18.7
	50-75	15	23.4
	75-90	12	18.7
	90-97	10	15.6
	>97	6	9.3
Weight Percentile	< 3	9	14,1
	3-10	11	17,2
	10-25	9	14,1
	25-50	16	25
	50-75	8	12,5
	75-90	5	7,8
	90-97	2	3,1
	>97	4	6,3
Clinics			
MVP syndrome	yes	36	56,3
Skeletal abnormalities	yes	12	18,8
Palpitation	yes	21	27,6
Atypical or non-anginal chest pain	yes	20	26,3
Dizziness or syncope	yes	1	1,3
Numbness or tingling	yes	0	0
Dyspnea	yes	7	9,5
Exercise intolerance	yes	3	4,1
Panic and anxiety disorders	yes	4	6,2
Abnormal resting and exercise electrocardiograms	yes	2	3,1
Findings			
Mitral Valve thickness at Echocardiography	<5 mm	45	70,3
	≥5 mm	19	29,7
Mitral insufficiency at Echocardiography	non	10	15,6
	mild	36	56,3
	moderate	16	25
	Moderate-severe	2	3,1
Significant arrythmia	Yes	8	12.5

Demographic and clinical information of the patients

PP-666
29-Years natural history of perimembranous vsds and restrictive vsds: a single center study

Selen Karagözlü[1], Mehmet Gökhan Ramoğlu[2], Tayfun Uçar[2], Semra Atalay[2] and Hasan Ercan Tutar[2]
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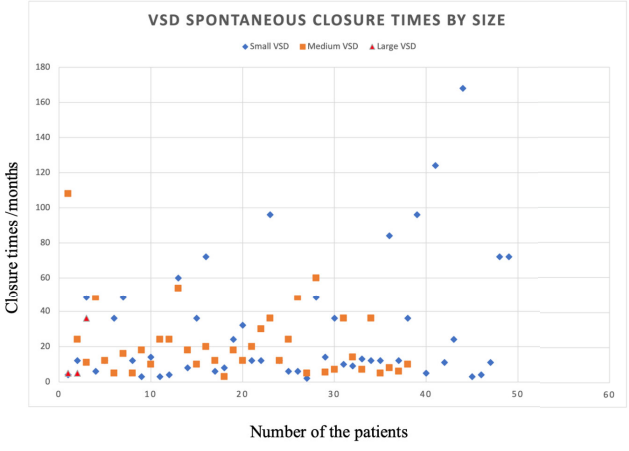
Background and Aim: Congenital heart disease is the most common birth defect and ventricular septal defect (VSD) is the most common congenital heart defect that accounts for approximately 32% of all heart defects diagnosed in the first year of postnatal life. Perimembranous ventricular septal defects (VSDs) are the most common VSD subtype in the United States, occurring in 75-80% of cases.The aim of this study is to shed light on perimembranous VSD and restrictive VSDs whose management still remains controversial.
Method: Children diagnosed with isolated perimembranous ventricular septal defect (pm-VSD) from 1991 to 2020 were enrolled in this retrospective study. The follow-up period was from one year to 29 years from the first diagnosis of the patients.The patients were classified according to the size and location of VSD.Echocardiographic findings, clinical conditions, follow-up period, treatment, data of the intervention and the surgery was evaluated.
Results: Four hundred and forty-eight patients (46.2% female) were evaluated. The mean age at first diagnosis a was 26.5±47.6 months.168(37.5%) of the patients had small VSD, 216(48.2%) had medium, and 64(14.3%) had large VSD. 81.3% of the large VSD had surgical closure.41.2% of the medium VSD was surgically closed and 17.6% is spontaneously closed. 29.2% of small VSD's closed spontaneously. There were 368(82.2%) patients with restrictive PM-VSD. 28.2% of those patients were treated (ACEI or/and diuretic or/and digoxin). 86(23.3%) cases of the restrictive PM-VSD closed spontaneously. 81(22%) patients had surgical and 2 had transcatheter VSD closure. LVEDD z-scores of all patients were evaluated during the follow-up period.The LVEDD z-score of 130 patients were above +2 at the initial, but 96(94.1%) of them were observed that LVEDD z-score decreased in the follow-up. Spontaneous closure times of VSD according to size are shown in Image 1.
Conclusions: Ventricular septal defects are among the most common congenital heart lesions. Although we have definitive knowledge about PM-VSD and restrictive VSDs for several years, we are still lacking accurate information on PM-VSD and restrictive VSDs. There is no debate that large VSDs require surgery. Spontaneous closure rate of the PM-VSD and restrictive VSD are lower than other conditions and they are still mainly open.

Keywords: Perimembranous, VSD, Restrictive

Image 1



Spontaneous closure times of PM-VSD according to size

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PP-667

Establishing a coronary artery anomalies program: Lessons learned from a standardized approach

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Background and Aim: Anomalous aortic origin of a coronary artery (AAOCA) is a rare congenital heart anomaly affecting up to 1% of the general population. Though this condition is rare, it is the second leading cause of sudden cardiac death in the young athlete, with left AAOCA carrying the highest risk. Optimal risk stratification in AAOCA remains a challenge. We aimed at establishing a dedicated program with a standardized approach to evaluation and management of patients with AAOCA.

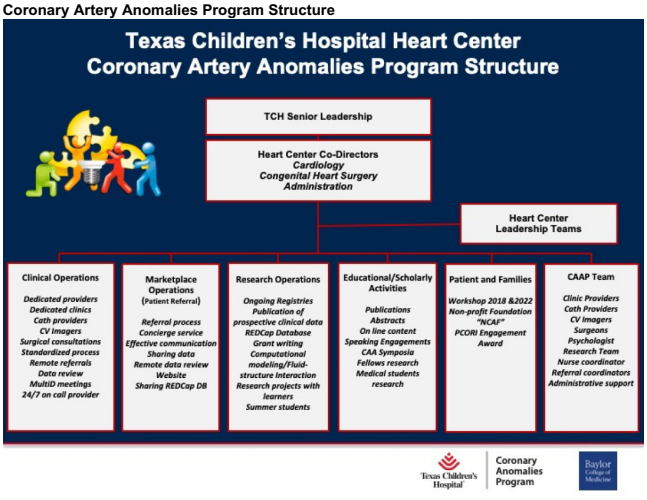
Method: A multidisciplinary team comprised of cardiology providers, cardiovascular surgeons, cardiovascular radiologists, nursing, outcomes and research staff was establishes in December 2012. Initial meetings were held and a standardized approach for evaluation and management decision-making was defined based on available knowledge of AAOCA. Quality Assurance and Program Report meetings were established every 1.5-2 years intervals. Assessment of data gathered, evolution of decision-making and lessons learned were compiled over a 10-year period.

Results: Following the development of this multidisciplinary team, several approaches culminated in a robust program with international reach, including: 1. Development of dedicated clinics and referral system network for gathering and collection of data for patients referred to the program; 2. Uniform imaging protocol with CT angiography for determination of key morphologic features thought to confer high-risk; 3. Establishment of reliable advanced perfusion imaging to determine inducible perfusion abnormalities upon provocative stress; 4. Evolvement of invasive coronary artery testing techniques and their implications; 5. Collection of data on quality of life following diagnosis of AAOCA and determination of psychological support for patients and families; 6. Evolution of surgical techniques according to morphologic features of AAOCA; 7. Evaluation of clinical and surgical outcomes; 8. Analyses and publication of robust data in AAOCA; 9. Institution of national scientific meeting; and 10. Engagement of patients and families with establishment of national (non-profit) foundation.

Conclusions: Establishment of a dedicated program with multidisciplinary team engagement and a standardized approach to evaluation and management, with data gathering and periodic reassessment of lessons learned, provided evidence-based clinical decision-making. This approach has empowered patients and families affected by AAOCA for shared decision-making on management decisions that affect their quality of life.

Keywords: anomalous coronary artery, AAOCA, standardized approach, program development

Coronary Artery Anomalies Program Structure



Comprehensive structure of the Coronary Artery Anomalies Program after 10 years aligns with the Heart Center Leadership structure at Texas Children's Hospital.

PP-668

A rare pediatric case of cardiac hydatid cyst

Simona Antoniu[1], Evelyn Maria Kecskes[1], Ana Maria David[1], Veronica Marcu[4], Mihaela Axente[5], Eliza Cinteza[6], Irina Margarint[2], Youssef Tammam[2], Monica Popescu[3] and Cristina Filip[6]

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Background and Aim: We present the case of a 15 years old female patient, transferred to our clinic with a suspicion of myocarditis after presenting to a secondary center for a syncopal episode followed by psychomotor agitation. Echinococcosis is a larval infection that affects over 1 million people worldwide, particularly in rural areas, and is characterized by the growth of hydatid cysts in internal organs, typically the liver and lungs. While hypersensitivity reactions are the main complication, severe complications can arise if the cyst grows in atypical locations such as the heart.

Method: Multiple imagistic, radiological, and biologic exams were performed (echocardiography, abdominal echography, cardiac and brain MRI, coronary angiography, serologic test (hydatid cyst antibodies) with enzyme-linked immunosorbent assay (ELISA), electrocardiography.

Results: The electrocardiography revealed ST elevation on the inferolateral leads with positive cardiac enzymatic shift. On echocardiography, a mass in the lateral wall of the left ventricle was noted alongside hypokinesia. On admission, the patient was stable with right side hemiparesis, aphasia and patent coronary arteries (coronary angiography was performed). A brain magnetic resonance imaging (MRI) revealed multiple ischemic strokes on the lateral frontal cortex and cerebellum. Following the betterment of symptoms, a cardiac MRI identified the mass as a hydatid cyst and an IgG antibody test confirmed the finding. Consequently, Albendazole was started followed by surgical resection of the cystic mass. Given the neurological involvement and the hemorrhagic risks, surgery was delayed for a month. The evolution was favorable with the motor deficit subsiding, cognitive function

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improving and an increase of contractility. Thus, the decision was made to discharge the patient under home treatment.
Conclusions: Despite the diagnosis difficulties, the outcome was favorable. The localization of the hydatid cyst in the left ventricle represents an extremely rare presentation of Echinococcosis in a pediatric patient, causing severe hydatid embolization of coronary and cerebral vessels resulting in aforementioned symptoms. Overall, this patient's case underlines the importance of early diagnosis and interdisciplinary management of rare and complex medical conditions, such as echinococcosis, in order to avoid severe and life-threatening complications.

Keywords: echinococcosis, cardiac hydatid cyst, hydatid embolization

PP-669

Non-traditional cardiometabolic risk factors in children and adolescents with metabolic syndrome may predict early onset atheromatosis

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Background and Aim: Cardiovascular disease is one of the leading causes of mortality worldwide. The aim of our study was to investigate whether "non-traditional" cardiovascular risk factors in children and adolescents with Metabolic Syndrome (MetS) may be used as biomarkers for predicting early atheromatosis, and evaluate their response to the implementation of a multidisciplinary, personalized, lifestyle intervention program for 1 year.
Method: One hundred forty-nine (n=149) children and adolescents [91males (61.07%), 58 females(38.93%), 32 prepubertal (21.48%), 117 pubertal (78.52%)] were studied prospectively. Subjects were classified as having MetS (n=89; mean age ± SE:13.19 ± 0.2 years) or not (n=60; mean age ± SE:12.05 ± 0.28 years) according to the International Diabetes Federation criteria. All participants underwent echocardiography, ultrasound of the carotid arteries to determine carotid intima-media thickness, liver ultrasound, and determination of biochemical, endocrinologic and "non-traditional" cardiometabolic risk factors (adiponectin, homocysteine, hs-CRP, leptin, IL-2, IL-4, IL-6, IL-10, IL-17A, TNF, IFN-γ) at the beginning of the study and following 1-year of intervention.
Results: MetS patients had significantly higher concentrations of pro-inflammatory cytokines IL-6, IL-17A and INF-γ compared to the control group ($P<0.05$). After intervention, patients with MetS showed a significant reduction in TG/HDL ($P<0.05$), ApoB/ApoA1ratio ($P<0.01$), leptin ($P<0.01$) and IL-6 ($P<0.01$) concentrations. Carotid triplex recorded a mean intima-media thickness (cIMT) of common carotid artery in the MetS group of 0.65 ± 0.03 mm at baseline, a value higher than normal for age (normal range: 0.49 ± 0.03 mm) and with a statistically significant difference from the control group 0.50 ± 0.02 mm). A significantly improvement in the MetS was recorded over one year of intervention in the MetS group($P<0.01$). In the echocardiogram, subjects with MetS had higher values in left ventricular septal thickness during diastole and systole, and larger end-diastolic and end-systolic diameters than the control group. Liver U/S revealed NAFLD in the MetS population at a rate of 51.5%, while a parallel improvement in steatosis, cIMT and IL-6 concentration was observed after the intervention. Multiple linear regression analysis indicated that independent variables associated with adipose inflammation, such as lL-6 and HOMA-IR index, were positive predictors of early-onset atheromatosis in children and adolescents.
Conclusions: These findings indicate that adolescents with MetS may have a greater risk for developing atheromatosis early in life, while early lifestyle intervention is crucial for preventing the arteriosclerotic process in youth.

Keywords: atheromatosis, cardiovascular risk-factors, carotid-intima-media-thickness, cytokines, metabolic-syndrome.

PP-671

Age-adjusted zlog-nt-probnp as a marker of heart failure symptoms in newborns with native hypoplastic left heart syndrome

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Background and Aim: As a marker of cardiac wall stress, NT-proBNP may be a useful indicator to quantify heart failure symptoms in newborns with native hypoplastic left heart syndrome (HLHS). However, little is known about NT-proBNP in this specific population and reference intervals are highly age-dependent. Therefore, the aim of the present study was to investigate its association with clinical findings, especially heart failure symptoms, in neonates with native HLHS.
Method: Biomarker levels were evaluated using an age-adjusted z-score (so called "zlog-NT-proBNP"), whose reference interval ranges from -1.96 to +1.96 independent of age. A total of 62 newborns with HLHS and at least one NT-proBNP measurement before Norwood surgery or hybrid approach were enrolled. Correlations between zlog-NT-proBNP and clinical variables (age, sex, height, weight, gestational age), lactate level, echocardiographic findings such as restriction of the atrial septum, grade of tricuspid regurgitation, and tricuspid annular plane systolic excursion (TAPSE), New York University Pediatric Heart Failure Index (NYU PHFI), and cardiothoracic ratio (CTR) were determined to identify key factors affecting (zlog-)NT-proBNP.
Results: NT-proBNP (median 35,000 ng/L, range 4,900–57,700 ng/L) was measured at a median age of 6 (IQR, 4–8) days. Corresponding zlog-NT-proBNP levels ranged from 1.76 to 4.50 (median 3.76) and were significantly correlated with the

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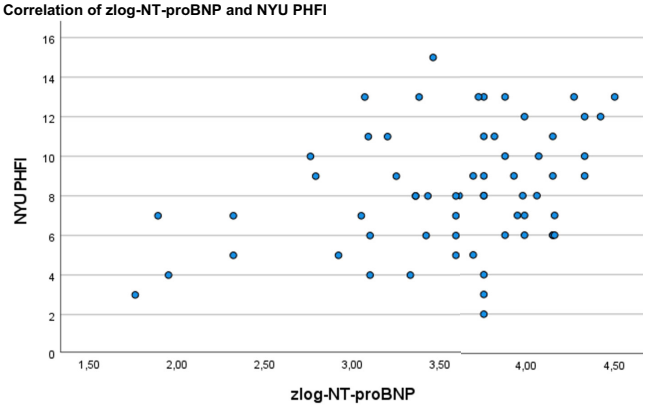
NYU PHFI (r=0.33, p=0.01). In contrast, the absolute NT-proBNP concentration did not show a significant correlation with the NYU PHFI (p=0.38) or other parameters except oxygen saturation (p=0.037). However, other variables associated with higher zlog-NT-proBNP levels were a higher CTR (r=0.26, p=0.042), age (logarithmized; r=0.30, p=0.02), oxygen saturation (r=0.24, p=0.063), respiratory rate (r=0.26, p=0.043), and dyspnea (p=0.016). No correlation was observed between zlog-NT-proBNP and echocardiographic findings, lactate level (p=0.78), gestational age (p=0.52), or weight (p=0.54). The NYU PHFI was most strongly associated with the grade of TR (r=0.38, p=0.002).

Conclusions: Age-adjusted zlog-NT-proBNP overcomes the disadvantages of absolute NT-proBNP concentrations with their strongly age-dependent reference intervals. It reliably reflects and helps to quantify heart failure symptoms in newborns with native HLHS.

Keywords: zlog, HLHS, NT-proBNP, heart failure, newborn, NYU PHFI



Age-ajusted zlog values ("zlog-NT-proBNP") correlated significantly to NYU PHFI in contrast to the absolute NT-proBNP.

PP-672

A landscape analysis of paediatric and congenital heart disease services in africa

Thomas Aldersley[1], Sulafa Ali[2], Adila Dawood[1], Frank Edwin[3], Alexia Joachim[1], John Lawrenson[1], Darshan Reddy[4], Amy Verstappen[5], Bistra Zheleva[6], Liesl Zühlke[7] and On Behalf Of All Respondents[8]
[1]University of Cape Town, Division of Paediatric Cardiology; [2]University of Khartoum, Division of Paediatric Cardiology; [3]School of Medicine, University of Health and Allied Sciences, Ho, Ghana; [4]University of Kwa-Zulu Natal, Division of Cardiothoracic Surgery; [5]Global Arch; [6]Children's Heart Link; [7]South African Medical Research Council; [8]Respondent countries: Algeria, Angola, Benin, Botswana, Burkina Faso, Burundi, Cameroon, Chad, Comoros, Democratic Republic of the Cong, Egypt, Eswatini, Ethiopia, Gabon, Gambia, Ghana, Kenya, Libya, Malawi, Mali, Mauritania, Mauritius, Morocco, Mozambique, Namibia, Niger, Nigeria, Rwanda, São Tomé and Príncipe, Senegal, Seychelles, Sierra Leone, Somalia, South Africa, South Sudan, Sudan, Tanzania, Togo, Uganda, Zambia, Zimbabwe.

Background and Aim: There is geographic disparity in the provision of Paediatric and Congenital Heart Disease (PCHD) services. Previous studies show that North America and Western Europe account for 74% of the world's cardiothoracic surgical capacity. In contrast, Africa accounts for only 1% of the total global capacity. However, PCHD training and service provision in Africa has increased. As such, we conducted a cross-sectional electronic survey to evaluate PCHD services in Africa.
Collaborators: Children's Heart Disease Research Unit, Pan-African Network for Paediatric and Congenital Hearts, and the African Society for Paediatric and Congenital Heart Surgery.
Method: Respondents were selected by purposive sampling and included paediatric and adult cardiologists and cardiothoracic surgeons, paediatricians, and non-specialists, involved in PCHD care. The survey included respondent, institution, and national-level queries related to human and infrastructure resources for paediatric cardiology, cardiac catheterisation, and cardiothoracic surgery.
Results: There were 119 respondents, from 92 institutions and 43 different countries in Africa. Aggregated country level data showed that 80% (34/43) of countries had some form of cardiac service, of these 17 (40%) provided a full PCHD service including interventional paediatric cardiology and paediatric cardiac surgery, 11 (26%) provided paediatric cardiac surgery services but no interventional paediatric cardiology service and 1 provided an interventional paediatric cardiology service but no cardiac surgery. Nine countries (20%) had no PCHD service (Figure 1a). There was median 1 (IQR:0–3.75) paediatric cardiothoracic surgeon per country or 0.04 (IQR:0.00–0.13) per million population, far below the recommended ratio of 1.25 per million population (Figure 1b). Similarly, there were median 3 (IQR:1–10) paediatric cardiologists per country or 0.18(IQR:0.03–0.35) per million population, below the recommended ratio of 2 per million population (Figure 1c).
Conclusions: Only 17(40%) countries had a full PCHD service including cardiac surgery and interventional cath. However, a further 12 countries provided some form of cardiac intervention, making a total of 29(67%) countries with interventional cardiac services. The capacity of these services, however, remains low with the ratio of paediatric cardiologists and cardiothoracic services well below international recommendations and no country, except Mauritius, with sufficient paediatric cardiologists and cardiothoracic surgeons to meet the recommended ratio per million population.

Keywords: Global Health, Africa, Congenital Heart Disease, Cardiothoracic Surgery, Survey, Landscape Analysis

Figure 1

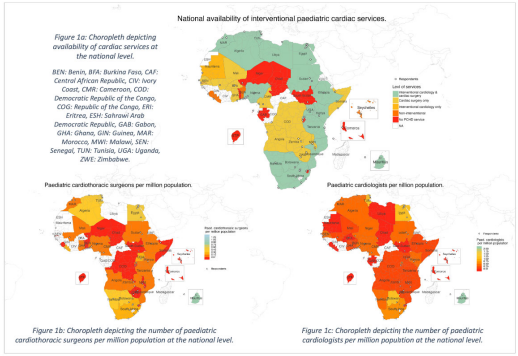


Figure 1: Choropleths depicting (A) availability of cardiac services at the national level, (B) the number of paediatric cardiothoracic surgeons per million population at the national level, and (C) the number of paediatric cardiologists per million population at the national level.

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PP-673

Should somatostatin be used as first-line agent in management of congenital chylothorax?

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Background and Aim: Congenital chylothorax is defined as abnormal accumulation of lymphatic fluid in the pleural space and may be either congenital or an acquired condition. Although congenital chylous effusions are relatively rare in infancy, they have serious clinical consequences and can be potentially life-threatening disorder. In our case the accumulation of chylothorax has been treated successfully after the administration of octreotide. We aim to provide guidance for the optimal management of Congenital Chylothorax in Infancy.
Method: A premature baby born at 30 weeks gestation, diagnosed antenatally with trisomy 21 syndrome and severe bilateral congenital pleural effusions which subsequently confirmed after birth as chylothorax. Bilateral thoracentesis were performed and bilateral chest tubes were inserted soon after birth due to the size of the effusion which compromised the respiratory system. Quantification of drainage used to determine clinical improvement and also used as a guide to fluid imbalance and replacement of daily losses. Expressed Brest milk and Medium-chain triglyceride(MCT)" formula was introduced in the first week of life. However Chylothoraces re-accumulated which required another bilateral thoracen-tesis and bilateral chest tubes. Congenital chylothorax was treated successfully after administration of Octreotide infusion along with intercostal decompression of the pleural effusion and total parental nu-trition as adjunctive therapy. In our case there was no any complications of with the use of octreotide.
Results: The MCT diets have met with variable success in the treatment of chylothorax. This is because any oral enteral feeding increases lymph flow. Octreotide is a synthetic, long-acting somatostatin analogue, It has been successful in treating chylothorax in conjunction with other modalities "TPN, effusion drainage" in conservative management.
Conclusions: This case is of particular interest because it provides an evidence for the efficacy of octreotide in the management of chylothorax. Therefore, octreotide may be used as first-line agent along with adjunc-tive therapy of parental nutrition and intercostal decompression of the pleural effusion. The early ad-ministration of Octrotide may allow the patient to avoid invasive procedures

Keywords: Congenital chylothorax, chylomicrons, Octreotide

PP-674

Rheumatic valve disease in two hospitals in central africa: Diagnostic, therapeutic and prognostic aspects

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Background and Aim: Rheumatic valve disease is a serious public health problem in low- and middle-income countries. They are secondary to rheumatic fever and can lead to serious complications if not diagnosed and treated early enough. The objective of our study was to study the diagnostic, therapeutic and prognostic aspects of rheumatic valve disease in two hospitals in the city of Yaounde.
Method: This was an analytical cross-sectional study conducted over a period of 8 months, from November 2022 to June 2023. Were included in our study all patients with a diagnosis confirmed by heart ultrasound of rheumatic valve disease in the cardiology departments of the central hospital and the Chantal Biya Foundation in Yaoundé. We collected socio-demographic, clinical, paraclinical, therapeutic and patient outcome data, data entry and analysis were done using SPSS software and the data were considered statistically significant for a P value < 0.05 with a 95% confidence interval.
Results: We included 41 children and the majority of patients was aged from10 to 14 years old. There was a notion of tonsillitis in 74.5% of patients. Complications were the most found circumstance of discovery. Patients had dyspnea and a heart murmur in 85.8% of cases. Monovalvular involvement predominated in 61.3% of cases, and the mitral valve was the most affected with 82.1% of cases of mitral regurgitation. A surgical indication was recommended in 69.3% of patients and 29.2% were operated. The proportion of deaths in the population was 6.3%. Factors associated with poor prognosis were age, patients who did not receive antibiotics for their tonsillitis and signs of heart failure.
Conclusions: Notwithstanding the progress in health care and the availability of antibiotic therapy, rheumatic valve diseases remain present and pose a problem of screening and management in our context. Primary prevention should focus on the treatment of sore throat and screening for rheumatic heart disease by health personnel.

Keywords: rheumatic valve disease, children, cameroon

PP-675

Left coronary birth defects from the aorta: Diagnostic and therapeutic challenge of a sudden death risk in a limited-resource country. A case report

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Background and Aim: Coronary artery birth and pathway anomalies, particularly those where the artery originates from the contralateral coronary sinus with a pathway between the aorta and the pulmonary artery, represent a high risk of sudden death. Early diagnosis and management are essential for a good prognosis.
Method: Clinical CASE: 9 year old boy brought to a paediatric cardiology consultation for loss of consciousness during physical effort with cardiac arrest, having required in situ cardiopulmonary resuscitation two days ago. It is was the second such episode within a month. No particular personal or family history. Physical examination revealed good general condition with 100% room air saturation, stable haemodynamic parameters and normal anthropometric parameters for age. Heart sounds are regular, no murmur, symmetrical femoral pulses regular and of good volume. The rest is unremarkable. In view of this normal physical examination and the notion of malaise during the effort, the suspicion of anomaly of the coronary artery pathway is evoked and a cardiac echography is carried out: left coronary artery coming from the

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right aortic sinus with a trunk pathway between the aorta and the pulmonary artery. Normal flow in both coronary arteries. Good overall cardiac function with an ejection fraction of 64%. A cardiac CT scan confirmed the origin of the left coronary from the right coronary sinus at 2 o'clock, which travels along the anterior aspect of the aorta in the inter-aorto-pulmonary, then divides into the anterior inter-ventricular and circumflex arteries at the level of the left atrioventricular fold. As the technical platform did not allow for surgical management in Cameroon, a medical evacuation was organised for unroofing of the left coronary.
Results: We presented a case revealed by a sudden cardiac arrest, in a country where diagnosis and management remain a real challenge due to limited technical facilities.
Conclusions: We presented a case revealed by a sudden cardiac arrest, in a country where diagnosis and management remain a real challenge due to limited technical facilities.

Keywords: coronary anomaly, child, sudden death, cameroon

PP-676
Infective endocarditis - the challenge of differential diagnosis

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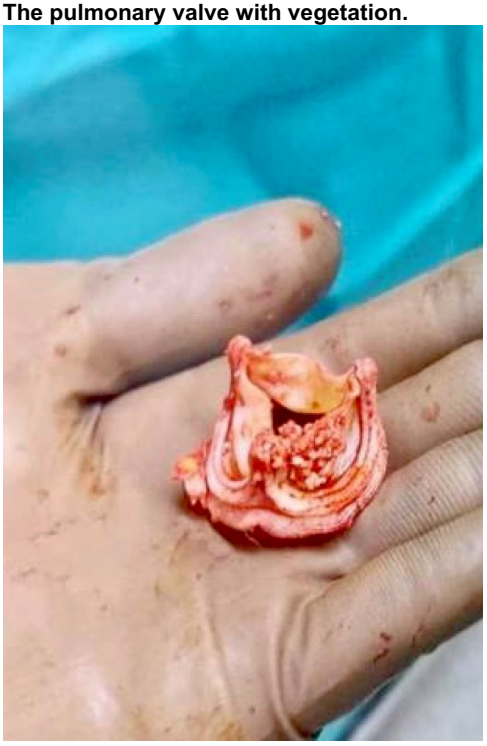
Background and Aim: Infective endocarditis (IE) is a life-threatening disease. However, early diagnosis may be challenging in some cases due to polimorphic clinical presentation. The aim of this article is to present unusual clinical manifestation of IE.
Method: 15-year-old girl who had a complete repair of Tetralogy of Fallot with implantation of Resilia Edwards Valve was admitted to Cardiology Department due to recurrent fever, anemia, microhematuria with casts erythrocytes in urine and proteinuria for the last six months. C-reactive protein and erythrocyte sedimentation rate were slightly elevated and estimated glomerular filtration rate (eGFR) was mildly low. The rheumatic factor (RF), antinuclear antibodies (ANA) and the anti-neutrophil cytoplasmic proteinase 3 antibodies (PR3-ANCA) ware positive. The serial blood cultures were all negative. Transthoracic echocardiography revealed a mass which may be compatible with vegetation on the pulmonary prosthesis and severe pulmonary stenosis and regurgitation. The positron emission tomography/computed tomography (PET/CT) demonstrated only mild perivalvular uptake.
Results: The patient was classified as "possible" IE according to the modified Duke criteria with associated glomonephritis. A biopsy was used to confirm a diagnosis of glomonephritis.
The antibiotic therapy with cloxacillin, ampicillin and gentamicin was administered and intravenous methylprednisolone pulse therapy started. A surgical intervention was planned. After the pulmonary valve replacement surgery a significant improvement in symptoms of glomerulonephritis was observed.
Conclusions: IE may present with various clinical situations. The physicians must maintain a high clinical suspicion in patients with symptoms of glomerulonephritis even if the blood cultures are negative.

Keywords: infective endocarditis, glomonephritis

The pulmonary valve with vegetation.



PP-678
Unilateral absent pulmonary artery: A rare and complex entity

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Background and Aim: Unilateral absent pulmonary artery (UAPA) is a rare congenital anomaly which may be associated with other malformations. Typically, a UAPA arises from a ductus arteriosus, which originates from the contralateral aortic arch.
Method: We describe two cases to illustrate the range of this malformation and to raise awareness for this rare diagnosis.
Results: Our first case is an asymptomatic three-month-old boy who underwent a routine cardiological evaluation because of bradycardia during inguinal herniotomy. The right pulmonary artery (RPA) was absent on echocardiography. CT demonstrated a residual peripheral RPA of narrow calibre fed by accentuated bronchial arteries. There was no continuity with the pulmonary trunk. The right lung was hypoplastic, but structurally regular. No right-sided patent ductus arteriosus was detected, but an outpouching at the base of the innominate artery was noted. As favourable outcome has been reported compared to staged repair, we opted for primary unifocalisation. The operation was performed without complications, using a pulmonary flap and a

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pericardial patch to create continuity between the pulmonary trunc and the RPA. As expected, a ligamentum arteriosum was found. The patient recovered rapidly. CT before discharge demonstrated a patent RPA with a localized narrowing. We proceeded with a ballon angioplasty 2 months later, which led to a significant improvement in RPA size and blood flow. Our second case was a preterm baby (35 weeks of gestation, birth weight 2350 g) who presented with recurrent pulmonary infections. Work up revealed congenital diaphragmatic hernia, agenesia of the RPA, hypoplasia of the right lung and consecutive dextrocardia. CT angiography showed multiple collaterals supplying the right lung but no peripheral RPA. Therefore, surgical re-anastomosis was not possible and repair of the congenital diaphragmatic hernia was performed.
Conclusions: Here, we demonstrate two cases with unilateral RPA agenesia. We aim to raise awareness for this rare congenital disorder. Patients can be asymptomatic as demonstrated by our first case description. Careful assessment and further diagnostic measures of suspected absence of the RPA is essential to avoid delaying treatment for this patient group.

Keywords: pulmonary artery, intervention, unifocalisation

PP-679

Cardiac dysfunction associated with chemotherapy in pediatric cancer patients and severity assessment based on esc guidelines

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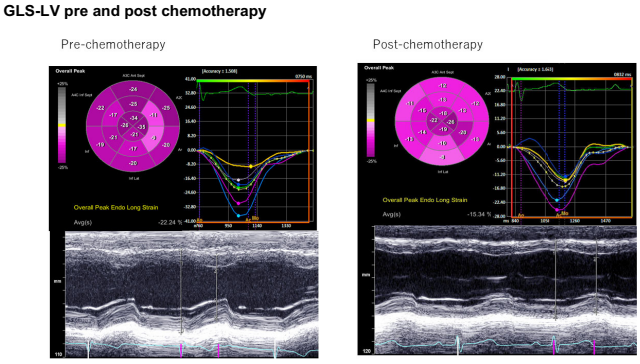
Background and Aim: Chemotherapy for pediatric cancer patients can cause cancer therapeutics-related cardiac disorder (CTRCD). In 2022, the European society of cardiology (ESC) proposed a CTRCD severity classification that includes Global Longitudinal Stain (GLS-LV) of the left ventricle based on echocardiography. The purpose is to evaluate the cardiac function of cancer treatment patients based on ESC guidelines and examine its usefulness.
Method: We analyzed Pediatric cancer patients treated at our hospital from 2020 to 2023 and underwent cardiac function evaluation from the start of treatment. In addition, 37 healthy children (age 0-17 years) served as a control group. Severity was classified based on echocardiography results and serological parameters. In statistical analysis, the t-test was used if the data variables followed normal distribution, otherwise, the Mann-Whitney U test was used, and $P < 0.05$ was considered significant.
Results: Twenty-six cases (15 males) of pediatric cancer patients were matched. They were age (median 5, IQR1-13 years), observation period (median 7.5, IQR 5-10 months), and total Adriamycin equivalent dose (ADM dose) (median 106, IQR 68-236 mg/body surface area). 15 cases were diagnosed with CTRCD (group A) and 11 cases were without CTRCD (group B). None of the target patients had congenital heart disease other than patent foramen ovale, and there was no significant difference in LVEF before the start of treatment compared to the control group. There were no significant differences in age, gender, observation period, or amount of ADM between Group A and Group B. All patients in group A were diagnosed with asymptomatic mild CTRCD during or at the end of treatment, but LVEF was >50% and there was no sustained increase in serological parameters until the end of treatment. Three patients in group A showed improvement in GLS-LV during follow-up, but one patient showed a decrease in LVEF in the distant period, leading to the diagnosis of asymptomatic moderate CTRCD.
Conclusions: Mild CTRCD was observed in 53% of the target patients, of which 1 progressed to moderate CTRCD in the distant stage. Evaluation of CTRCD including GLS-LV was useful as an indicator for starting close follow-up.

Keywords: chemotherapy-related cardiac dysfunction, Global longitudinal strain, cardiac failure, CTRCD,



Absolute values of GLS-LV reduced after chemotherapy.

PP-680

Cardiovascular complications in kawasaki disease. Ukrainian experience

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Background and Aim: Kawasaki disease (KD) is a leading cause of acquired heart disease in children in the world. The diagnosis of KD in Ukraine was often based on the presence of coronary artery lesions (CAL), but not on the clinical criteria. The aim of the study were to evaluate the level of KD patients in Ukraine, to analyze risk factor of complications in KD, to examined cardiovascular complications in KD, to evaluate the results of the KD program in Ukraine.
Method: In the Ukrainian Children`s Cardiac Center from 2013 to 2023y were examined 108 patients with KD aged from 3 mo. to 13 yr. Most patients with KD were diagnosed in Kyiv region - 78% (n=85).
Results: From 2013 to 2016 KD was diagnosed only in 3 patients and 2 of them had CAL (66%). After 2016 the diagnosis of KD improved and we had 105 patients and 22 (21%) of them had CAL. From total number of KD that we saw, 25 patients (23%) had CAL: small coronary artery aneurysm - 68% (n=17); medium-sized aneurysms - 12% (n=3); giant aneurysms - 20% (n=5). Of them, 12 (11%) had pericarditis, 5 (4.6%) – myocarditis and 2 (1,8%) - mitral insufficiency. One patient (0.9%) with giant coronary aneurism had cardiac surgery due to myocardial ischemia. Mortality was 0.9% (n=1). Fourteen patients had complete regression of coronary aneurysms during the follow-up (58%).
Conclusions: High incidence of serious CAL (medium-sized, giant aneurisms) is associated with late diagnosis and late treatment. Start of KD program in Ukraine showed good results of diagnostics but

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level of CAL is still high (23%). Age younger 5 years was significant risk factor for CAL (29%). Data from UCCC showed KD is not yet diagnosed at a high level.High incidence of serious CAL (medium-sized, giant aneurisms) is associated with late diagnosis and late treatment.

Keywords: Kawasaki disease, Coronary artery lesions, giant aneurisms.

PP-681

Infective endocarditis in children with structurally normal hearts

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Background and Aim: Although infrequent, infective endocarditis (IE) can be seen in children with structurally normal hearts. The risk factors include prematurity, prolonged intensive care unit stay, use of central venous catheters, and immunosuppression. Our study aims to assess the features of IE in children without underlying heart disease.
Method: Patients aged <18 years with structurally normal hearts who were treated for IE at the National Heart Hospital, Sofia, Bulgaria, for 14 years (01.01.2009-31.12.2022) were included in the study. The diagnosis IE was based on the modified Duke criteria. The demographics, clinical characteristics, predisposing conditions, clinical symptoms, laboratory results, microbiology, localization of the vegetations, therapeutic approach, and outcome of the patients were analyzed.
Results: 6 children (mean age 3.5 years (12 days - 9 years, SD 2.9)), 50% male without underlying heart disease were diagnosed with IE during the 14-year period. 3 of them (50%) were aged <1 year and were born prematurely. 5 out of 6 children had predisposing factors: two of them were premature, one developed IE after tonsillectomy, one had acute hepatitis A, and one patient had dental caries. The most frequent symptoms were fever and neurological manifestations. In all cases, the levels of inflammatory markers were increased. In the majority of patients, the left-sided heart valves were affected: the mitral valve (n=2), the aortic valve (n=2), both the mitral and the aortic valve (n=1), and by one premature newborn with central venous line the tricuspid valve was involved. The course of the disease was progressive, with rapid destruction of the valves and the need for valve replacement in two cases. There was one relapse of IE and one lethal outcome.
Conclusions: IE can affect children with structurally normal hearts, these cases are usually caused by aggressive pathogens, with acute course of the disease with extensive destruction of the valves, and need for valve replacement in a high percentage of cases. This is why IE should be considered in the differential diagnosis of patients with fever of unknown origin, and blood cultures should be taken before the start of antibiotic therapy.

Keywords: infective endocarditis, structurally normal hearts

PP-682

The effect of the COVID-19 pandemi on childhood myocarditis

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Background and Aim: Myocarditis may develop in children during COVID-19 infection, and MIS-C (Multisystem inflammatory syndrome), or after after COVID-19 vaccine. The aim of the study was to evaluate the effect of Covid 19 pandemic in childhood myocarditis compared to the children diagnosed as myocarditis in prepandemic period.
Method: In this study, the files of children, diagnosed as myocarditis and hospitalized between the 2017-2022(two and a half years before and after pandemi) were retrospectively analyzed. Sociodemographic characteristics of the patients, anthropometric data, vital signs at the admission, complaints at admission, laboratory and imaging results, disease severity and prognosis, need for intensive care unit, treatment methods, morbidity-mortality data, risk factors for intensive care need were evaluated.
Results: A total of 61 children, 41 boys (67%) and 20 girls (33%), were included in the study. Groups of patients diagnosed with myocarditis before and after pandemi were compared. Patient groups diagnosed with post-COVID-19 myocarditis were subgroped as COVID-19 related, MIS-C related, Covid 19 vaccine related and COVID-19 unrelated. In our study, 18% (n=11) of 61 patients were diagnosed with pre-COVID-19 and 82% (n=50) post-COVID-19 myocarditis. Post-COVID-19 patients had significantly higher heart rate, respiratory rate, body temperature, and significantly lower systolic and diastolic blood pressures at the time of admission. While disease severity was mostly mild in pre-COVID-19 patients, disease severity was moderate-severe in post-COVID-19 patients. The length of hospital stay was longer in the post-COVID-19 group. The need for ntensive care unit was found to be significantly higher after COVID-19. Septic shock, permanent cardiac systolic dysfunction and multi-organ involvement were found to be significantly higher after COVID-19. The variables that best predicted the need for intensive care unit were determined as CRP and ProBNP.
Conclusions: The COVID-19 pandemic has increased the incidence of myocarditis. The severity of the disease was more severe in patients diagnosed with myocarditis after COVID-19. With the effect of the COVID-19 pandemic, the need for intensive care units has increased. Patients diagnosed with myocarditis after COVID-19 have a worse prognosis.

Keywords: Myocarditis, COVID-19 pandemi, children

PP-683

A case of a 2-year-old boy with kawaski disease with atypical onset

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Background and Aim: Kawasaki disease is an inflammatory disease of medium caliber vessels. It is crucial to initiate appropriate treatment quickly to prevent complications such as coronary artery aneurysms.
Method: A 2-year-old boy was admitted to the Pediatric Department due to dehydration, diarrhea and fever up to 40.5 C lasting for 6 days. The child complained of mouth and abdomen pain. One day before admission, on the boy's skin appeared a small-papular rash on the abdomen, back and legs, as well as redness and bleeding of the conjunctiva of the eyes. Additionally, runny nose, red lips, dry mucous membranes, a tongue covered with white coating and a red throat were found.
Results: Laboratory tests showed high inflammation level (CRP 109 mg/l), blood count with a neutrophilic smear, elevated liver enzymes (AST 48 U/l, ALT 170 U/l, GTP 110 U/l). Rota and

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adenoviral, HIV, CMV, EBV infections and hepatitis A, B and C were excluded. Abdominal ultrasound didn't show any pathology. After obtaining blood, urine and stool cultures, treatment was started with third-generation cephalosporin and intravenous hydration. Despite the treatment, the fever persisted, and only on the second day of hospitalization did swelling of the eyelids and feet appear. Diagnostics for MIS-C and Kawasaki disease have been expanded. The tests showed an increase in D-dimer concentration and the presence of SARS-CoV-2 IgG (121 BAU/ml), no IgM. Due to suspicion of Kawasaki disease, immunoglobulin infusions were used for treatment. Soon, an improvement in the patient's clinical condition and normalization of laboratory parameters were observed. The results of the cultures taken were negative. In a cardiac ECHO examination performed during hospitalization, no changes in the coronary arteries were observed. The patient was discharged home in good general condition, with a recommendation to perform a follow-up ECHO in a month.
Conclusions: Since the beginning of the SARS-CoV-2 pandemic, the differential diagnosis of MIS-C syndrome and Kawasaki disease has been very difficult. Both conditions should be remembered in the case of prolonged fever without an identifiable cause, even if the patient does not present characteristic symptoms from the beginning, as was the case in the presented case.

Keywords: Kawasaki disease, Multisystem inflammatory syndrome in children (MIS-C), SARS-CoV-2, COVID, immunoglobulines

Genetics, Basic Science and Myocardial Disease

PP-684
Noncompaction cardiomyopathy in pediatric patients: Single centre experience

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Background and Aim: Noncompaction cardiomyopathy (NCCM) is a disease characterized by hypertrabeculation of left ventricle due to an arrest in compaction during fetal development. Although NCCM is a rare disease, it is the third most common cardiomyopathy (CMP) in the pediatrics. In 2006, NCCM was classified as a distinct form of CMP by the American Heart Association. In pediatric population NCCM can present as isolated and most frequently non-isolated phenotype (with concomitant congenital heart disease, metabolic or neuromuscular disease).
Method: Participants were children who were examined on an outpatient basis or hospitalized in period between 1st January 2020 to 1st October 2023 and fullfed Rotterdam echocardiographic criteria.
Results: During this period there were 16 patients with diagnosis of NCCM, 8 was males and 8 females. The age range was from 3 months to 18 years of age. Most participants have normal electrocardiographic (ECG) findings and two have nonspecific changes such as biventricular hypertrophy. Continuous 24 hours ECG were performed in 9 participants, 8 of them had sporadic ventricular ectopy in unsignificant count. All participants had fullfed Rotterdam echocardiographic criteria. Ten participants underwent of cardiac magnetic resonance (CMR), and Petersen diagnostic criteria was met in nine. Genetics testing was performed in six participants while five had pathological mutation. Metabolic disorder as underlined factor for development of cardiomyopathy was confirmed in 2 patients. Eight participants had non-isolated form of NCCM. Complications of NCCM such as ventricular arrhythmias, complete AV block, thromboembolic incidents in the observed group were not recorded. Fourteen patients had medicament therapy. Tree patients underwent heart transplant.
Conclusions: NCCM is novel cardiomyopathy. Diagnosing NCCM can be challenging due to non-uniform diagnostic criteria. Although NCCM was considered a rare form of cardiomyopathy in the pediatric population in our study group, it accounted for 20% of all cardiomyopathies in the observed period. The diagnosis is based on echocardiography and CMR. CMR is gold standard for diagnosis. There are not specific ECG changes for NCCM. Considering the proportion of positive findings of gene processing, we would like to emphasize the importance of genetic and metabolic testing in patients with NCCM. There is no specific medical or surgical treatment.

Keywords: cardiomyopathy, non-compaction cardiomyopathy, hypertrabeculation.

PP-685
Blood pressure and aortic dilation in girls with turner syndrome

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Background and Aim: Turner syndrome (TS) is caused by a partial or complete absence of an X-chromosome and is associated with significant cardiovascular morbidity and mortality. Aortic dissection occurs six times more frequently and at a younger age compared to the general population. Many risk factors for dissection, such as the presence of a bicuspid aortic valve (BAV), cannot be modified. However, blood pressure (BP), aortic dilation and stiffness can be monitored and hypertension can be treated. This study describes the evolution of these parameters in paediatric TS patients.
Method: retrospective longitudinal study based on data extracted from the Electronic Patient Records of all paediatric TS patients followed at the paediatric department of the Ghent University Hospital. Parameters included: biometry, age, BP, aortic diameter, aortic stiffness, aortic valve morphology, and genotype. (45,X; 45, X/46,XX;other mosaic genotypes) BP and echocardiographic measurements were compared at childhood (4-7 years), pubertal age (10-13 years) and adolescent age (15-18 years).
Results: 72 patients were included (mean age at consultation 11.8 ±4.3 year; range 29 days – 21 years; 17 with BAV; 5 with aortic coarctation). 31.7% of in office systolic blood pressure measurements (SBP) and 18.3% of diastolic (DBP) measurement were ≥p95 for age, sex and height. 12.9% of SBP and 13.7% of DBP measurements were ≥p90 and<p95 for age, sex and height. No significant difference was found in BP comparing different genotypes or aortic valve morphology. A significant higher Z-score of the ascending aorta diameter was found in patients with a BAV compared to patients with a tricuspid aortic valve ($p<0.001$). Paired tests showed a significant increase in blood pressure percentile ($p<0.001$), between pubertal and adolescent age ($p<0.001$). The aortic stiffness index significantly increased with age ($p=0.05$), whereas the Z-score of the aorta did not change significantly.

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Conclusions: : compared to the general population, young TS patients have an increased blood pressure warranting close monitoring from a young age. Although z-scores of the aorta are also higher than average, they show little progression over time during childhood. This is reassuring as aortic dilation is considered a precursor of aortic dissection.

Keywords: Turner syndrome, blood pressure, aortic dilation, aortic stiffness, longitudinal, childhood

PP-686

Cardiac complications in children with inflammatory connective tissue diseases. A single centre experience

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Background and Aim: Inflammatory connective tissue diseases include systemic lupus erythematosus (SLE), juvenile dermatomyositis (JDM), systemic sclerosis (SScl) and mixed connective tissue disease (MCTD). Each of them may be associated with cardiac complications such as pericarditis, myocarditis, valvular disease, diastolic dysfunction, pulmonary hypertension, premature atherosclerosis or conduction disorders. Their prevalence has been reported in 3 – 40% of adult patients depending on the specific disease. Our aim was to find out the burden of cardiac complications in our cohort of children with inflammatory connective tissue disease and to raise awareness of diagnostic and therapeutic challenges in these patients.
Method: All patients diagnosed with SLE, JDM, SScl and MCTD from 2012 until 2023 at our institution were included in the study. Their medical reports together with ECG and echocardiography records were retrospectively analyzed.
Results: Altogether 33 children (31 girls and 2 boys) aged median 14 years were diagnosed with SLE. Among these, 11/33 (33%) developed cardiac complications such as pericarditis (4/11), significant repolarization changes on ECG (3/11) and mitral valve insufficiency probably related to Libman-Sacks endocarditis (2/11). One of the latter patients required an urgent mitral valve replacement. One patient died from myocardial infarction and another one succumbed to basilar meningitis complicated by severe heart failure. Among children with JDM (13 girls, 8 boys, aged median 7 years), 2/21 (10%) developed a mild myocarditis with preserved ventricular function. The diagnosis of SScl and MSTD/overlap was established in 5 and 4 girls, respectively. None of them has manifested heart involvement so far.
Conclusions: The burden of cardiac complications in children with inflammatory connective tissue diaseses is relatively low. However, in exceptional cases it may lead to severe complications and even require surgical intervention. The study shows the cardiac complications in the context of the severity of the respective diseases, their extracardiac manifestations and their immunological profile.

Keywords: inflammatory connective tissue disease, pericarditis, Libman-Sacks endocarditis

PP-687

Novel nkx2-5 pathogenic variant in a family with atrial septal defect, non-compaction cardiomyopathy, atrioventricular block and sudden death

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Background and Aim: Mutations in the NKX2-5 gene have been associated with congenital heart disease, mainly atrial septal defect (ASD), atrioventricular block (AVB), noncompaction cardiomyopathy and sudden death (SD).
Method: The index case is a 5-year-old girl with bradycardia secondary to advanced 2:1 AVB, requiring pacing at age of 14. One of the proband's sister had first-degree AV block and her mother had percutaneous closure of ASD. The grandfather, who had ASD, suffered SD at 54 years old and the great-grandfather died at 55 years due to cyanotic heart disease.
A cousin of the index case, now 12 years old, required surgical closure of ASD and he had a first-degree AVB and non-compaction cardiomyopathy as well as prolonged sinus pauses. His father, maternal uncle of the index case, had undergone surgery for ASD and presented hypertrabeculation of the left ventricle on MRI without meeting diagnostic criteria for noncompaction, with preserved systolic function. On holter, atrial fibrillation and non sustained ventricular tachycardia were detected. During the process of completing the study, he presented SD at age of 35.
Results: The genetic study of the deceased showed a possibly pathogenic mutation, p.Ala103Serfs*5, in the NKX2.5 gene, not previously described. A family genetic cascade study was performed, confirming pathogenicity by genotype-phenotype segregation. During follow-up, a transvenous implantable cardioverter defibrillator (ICD) was indicated for primary prevention in the proband, her sister and mother, as well as in the first cousin.
Conclusions: In patients associating ASD and AVB of various degrees or in familial cases with these phenotypes, it is important to indicate genetic study of NKX2-5, considering the therapeutic implications of the diagnosis. It would be advisable to perform multicenter studies of this genetic disease to allow better clinical characterization, stratify the risk of sudden death, and develop indications for ICD in primary prevention, given the high risk of malignant ventricular arrhythmias.

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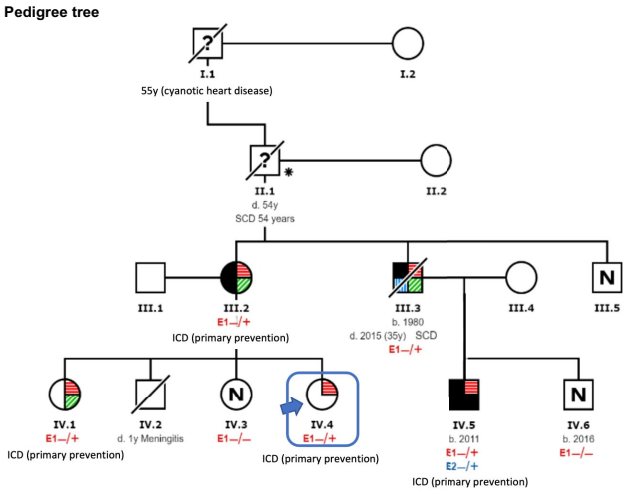
Keywords: NKX2-5, atrial septal defect, non-compaction cardiomyopathy, atrioventricular block, sudden death, cardiogenetic.



Pedigree tree. ICD: implantable cardioverter defibrillator. SCD: Sudden cardiac death.

PP-688

Cardiopulmonary exercise test as a key tool for the diagnosis of mitochondrial disease in an adolescent with exercise intolerance and lactic acidosis

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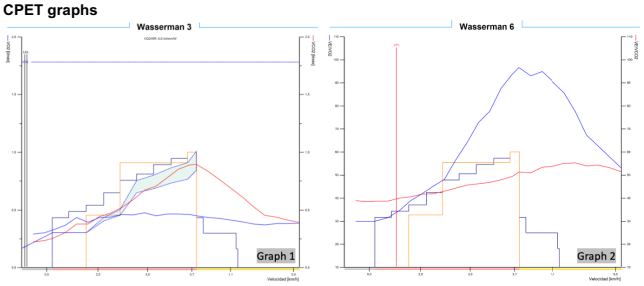
Background and Aim: Exercise intolerance and early fatigue are the most common symptoms in mitochondrial myopathies (MM) due to an increased dependence of skeletal muscle on anaerobic metabolism, generating excessive lactate.
Method: We report a 15-year-old girl with dyspnea on minimal efforts and exercise intolerance since early childhood. No other relevant antecedents. Transthoracic echocardiogram (TTE) and electrocardiogram (ECG) were normal. Cardiopulmonary exercise test (CPET) was performed on a treadmill, which stopped due to dyspnea at 5:21 min.
Results: CPET showed severely decreased aerobic capacity (VO2max 0.48L/min; 10.2ml/kg/min, 27% of predicted VO2). Important ventilatory inefficiency stands out (VE/VCO2 47), reaching the anaerobic threshold in very early stages, which reflects early metabolic acidosis, confirmed by post-exercise blood gas analysis (pH 7.17, lactate 8 mmol/l). These findings were suggestive of aerobic metabolism impairment and mitochondrial disease. Metabolic study was requested, highlighting elevated acidosis metabolites and coenzyme Q10 (CoQ10) depletion. Empirical treatment with cofactors (carnitine, thiamine, riboflavin, CoQ10, biotin) was undertaken with clinical improvement in daily activities. The genetic study revealed two heterozygous variants in SLC25A26 gene, causing mitochondrial disease due to oxidative phosphorylation defect, compatible with clinical phenotype.
Conclusions: Combined oxidative phosphorylation deficiency is caused by a mutation in the SLC25A26 gene, which encodes a mitochondrial transporter. It is an autosomal recessive multisystem disorder associated with mitochondrial dysfunction, affecting RNA stability, protein modification, mitochondrial translation, and biosynthesis of CoQ10 and lipoic acid. The phenotype is variable, including episodic metabolic decompensations with early lactic acidosis, mild muscle weakness, cardiorespiratory failure, developmental delay, or even death. CPET has been used to assess musculoskeletal oxidative capacity in patients with MM. Differences between healthy subjects and patients with mitochondrial dysfunction are found in numerous steps of oxygen transport and consumption: abnormal ventilation rates, reduced maximal oxygen consumption, altered oxygen delivery. It is a useful non-invasive tool in the diagnosis of mitochondrial disorders, especially in patients with mild symptoms. In our case, it was the key that led to the final diagnosis. Early suspicion and diagnosis of these diseases is important because, although there is no curative treatment, improvement of symptoms has been described with treatment with enzyme cofactors in some subjects.

Keywords: lactic acidosis, mitochondrial disease, exercise intolerance, Cardiopulmonary exercise test



Graph 1 shows a very flattened O2 consumption curve and a severely decreased maximal consumption. It also shows a CO2 production disproportionate to O2 consumption from very early stages of the effort, translating an important participation of anaerobic/lactic metabolism with minimal efforts. The data agree with what is observed in the second graph (O2 and CO2 equivalents), the first ventilatory threshold appears as soon as the effort begins. These graphs are characteristic and should raise the suspicion of mitochondrial disease in patients without known cardiovascular disease.

PP-689

Exon 3 deletion of ryanodine presenting with phenotype from fetal life: Expanding the clinical spectrum of ryanodinopathies

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Background and Aim: Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a cardiac channelopathy characterized by ventricular arrhythmias triggered by exercise or psychological stress with risk of sudden death. The most common genetic cause is gain-of-function variants in the ryanodine gene (RYR2), but the disease may also be due to variants in the cardiac calsequestrin gene, tight junction triadin, calmodulin and KCNJ2. In classic CPVT, there is no structural heart disease and the baseline ECG does not usually show significant alterations.

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Method: The proband was studied during fetal life because of marked sinus bradycardia (80 beats per minute). Profound sinus bradycardia was confirmed at birth and congenital long QT syndrome (corrected Qt 500 msec) was suspected, motivating beta-blockers treatment. Echocardiography at birth showed non-compaction cardiomyopathy and dilatation of the ascending aorta. In the following months, normalization of the QT interval was observed but the ECG still showed pathological repolarization with negative T waves in left precordial leads, so treatment with nadolol was continued. The initial genetic study for long QT syndrome and cardiomyopathy was negative. During follow-up, pathological sinus bradycardia persisted with a mean heart rate of 45 bpm. At age 12, exercise testing showed bidirectional extrasystoles triggered by physical exertion. Flecainide was added to nadolol. The patient remained asymptomatic.
Results: A new genetic study was requested, including analysis of copy-number variation (CNV), and p.Asn57_Gly91del, exon 3 deletion of RYR2 gene, was detected. This variant was not present in the asymptomatic parents, both with normal cardiological study, and was therefore de novo.
Conclusions: Exon 3 deletion of ryanodine is a rare cause of CPVT, with less than 50 cases reported in the literature. It has been described to be associated with sinus node dysfunction (58%), atrioventricular conduction disturbance (22%), non-compaction cardiomyopathy (31%) and sudden death (11%). The case presented demonstrates how sinus bradycardia can be detected even before birth and noncompaction cardiomyopathy can be present from birth. The association with dilated ascending aorta has not been previously described. This rare genetic entity should be suspected when such phenotypes' association is found. Further studies are needed to know the optimal management of this disease.

Keywords: ryanodine, noncompaction cardiomyopathy, RYR2, Catecholaminergic polymorphic ventricular tachycardia, genetic cardiology

PP-690

Loeys-dietz syndrome- experience of one medical center

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Background and Aim: Loeys-Dietz syndrome (LDS) is a rare, genetically conditioned connective tissue disorder with characteristic triad of symptoms including bifid uvula, hypertelorism and vascular disorders. The vessels abnormalities including aneurysms and tortuosity can be found in every artery. This can impact patients survival rate, by leading to the arterial dissection and premature death. The aim of this study is to present the data on LDS syndrome specific for the Polish population based on one center study.
Method: The retrospective data on patients with recognized LDS were collected from the visits in the Pediatric Cardiology and Congenital Heart Defects Department since 2012 till 2023. The patients had undergone detailed medical family history interview, physical examination with focus on characteristics typical to connective tissue disorders, echocardiography, radiological examinations e.g. whole body vessels CT/MRI, genetic testing and ophthalmology consultations when necessary.
Results: Data on 26 patients with genetically confirmed LDS or clinically confirmed LDS with presence of the positive family mutation history were collected. The age range of patients varied between 11 months till 51 years. The mutations present in the polish society included TGFBR1 (19%), TGFBR2 (19%), SMAD 3 (15%), TGFB2 (15%), TGFB3 (11%), SMAD2 (4%). Four families were recognized, the rest of patients presented somatic mutations. In echocardiography, aortic root dilation happened the most frequently (61%), distal part of the aorta dilation less frequently (19%), aortic annulus and STJ dilatation both in 8% of patients. Other cardiological defects included: biscupid aortic valve (15%), patent ductus arteriosus (15%) and atrial septal defect (19%). 23% of patients showed vessels tortuosity. 4 of patients (15%) underwent together 6 cardiosurgeries. 3 (11%) patients underwent 1 cardiosurgery, 1 patient (4%) underwent 3 cardiosurgeries. One patient (4%) went through aortic dissection type A.
Conclusions: LDS is a very severe connective tissue disorder that can lead to life- threatening consequences e.g. aneurysm dissection. From our experience many of the patients were observed for Marfan syndrome, however due to the new genetic possibilities LDS was confirmed. As a result of long-term observation the patients are given frequent, multidisciplinary care which prevents them from preterm fatal consequences.

Keywords: LDS, Loeys-Dietz, HCTD, connective tissue

PP-691

Hypertrophic cardiomyopathy in raf1-related noonan syndrome treated with mek inhibitors: Two levels of success depending on the mutation type

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Background and Aim: Hypertrophic cardiomyopathy (HCM) is a frequent and life-threatening complication in patients with RAF1-related Noonan syndrome. The RAF1 gene regulates many crucial cell processes including proliferation, differentiation, and apoptosis of cells. It consists of three main conserved regions (CR), with different functions. Missense mutations in RAF1 gene affect functioning of the extracellular-signal-regulated kinase and mitogen-activated protein kinase (ERK/MAPK) pathway. However, mutations in different regions have different phenotypes and symptoms.
Method: Two female patients (5 and 16 years old) with Noonan syndrome and HCM were enrolled for experimental treatment with MEK inhibitor (Trametynib) due to the absence of alternatives. The patients had different mutations in RAF1 gene, 5-year-old one in CR2 and 16-year-old in CR3 region. After receiving approval of clinical ethics board and informed parental consent, Trametinib treatment with 0.025 mg/kg/day dose was introduced.
Results: In both patients very prompt clinical and cardiac improvement was observed after 4 months of treatment. In a 5-year-old patient left ventricular outflow tract (LVOT) maximal systolic pressure gradient dropped from 140 to 78mmHg, and NT-proBNP from 30,000 to 3384 pg/ml. The left ventricle global longitudinal strain (LV GLS) increased (before vs after treatment): A4CH: 15,7% vs 19,4%; 3CH: 10,2%, vs 12,8%, 2CH: 16,1% vs 21,9%, Avg - 14% vs 18%. In 16-year-old-patient, LVOT maximal systolic pressure gradient dropped from 59 to 26 mmHg, NT-proBNP from 1397 to 62 pg/ml, and LV GLS increased respectively: A4CH: 17,8% vs 20%; 3CH: 16,5% vs 15,3%; 2CH: 11,2% vs 18,2%; Avg: 15,2% vs 17,8% In follow-up CMR of both patients no further progression of hypertrophy was observed

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(1 mm reduction of hypertrophy was considered within margin of error). Improvement of physical performance was reported by parents of both patients.
Conclusions: Treatment with trametinib in two patients with Noonan syndrome and HCM with different mutations in RAF1 gene was associated with cardiac and clinical status improvement in a short treatment period. MEK- inhibition might represent a successful alternative treatment in RASopathies. The response to therapy depends on the mutation type.

Keywords: Noonan syndrome, Hypertrophic cardiomyopathy, MEK-inhibitors

PP-692
Present ck levels do not correlate with cardiac function in dmd

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Background and Aim: The development of dilated cardiomyopathy is a slow, yet an inevitable process in Duchenne muscular dystrophy (DMD). With heart failure recognized as a leading cause of death in this group of patients, identification of risk factors and optimalization of prophylactic treatment are subjects of ongoing research. The aim of this study was to determine the utility of biomarkers for risk stratification in DMD.
Method: Left ventricle function based on ejection fraction (LVEF) in echocardiography (Teichholz method) and biomarkers serum levels in male individuals with genetically confirmed DMD were analyzed in a single-centre observational study. The echocardiography was performed within 2 days after CK, CK-MB and NTproBNP analysis. Statistical analysis was performed using Wizard 2.0 (Evan Miller, Chicago, IL). Data are presented as median (range).
Results: Forty records acquired from patients aged 10.75 (4.25-17.6) years, one record each, were analyzed. The LVEF was 64 (48-75)%, decreased (<55%) in 2 patients. In all patients CK and CK-MB were exceeding reference values: 6472 (672-32172) U/l and 65,25 (11.9-722.6) U/l respectively. NTproBNP was 53 (7-374) pg/ml, increased (>125 pg/ml) in 6 cases. LVEF, CK, CKMB were negatively correlated with age, P values <0.001 each. For NTproBNP no similar correlation was found (P=0.310). Furthermore, negative correlation was found between CK levels and LVEF (P<0.001) and NTproBNP levels and LVEF (P=0.036). Yet, after correction for age no correlation of biomarkers and LVEF was found, P values of 0.426 for CK, 0.217 for CKMB and 0.055 for NTproBNP. In case of NTproBNP there was a clear negative trend and lack of statistical significance may be due to small sample size (underpowered).
Conclusions: Present biomarker levels do not correlate with cardiac function in DMD. Further studies including prolonged observation and biomarkers trends are warranted to identify individuals at higher risk of unfavorable course of the disease and, eventually, application of intensified prophylactic management.

Keywords: Duchenne Muscular Dystrophy, Heart Failure, Biomarkers

PP-693
Genotype and cardiac outcome for patients with cardio-cutaneous syn-drome (NAXOS and NAXOS-like disease) in Saudi Arabia

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Background and Aim: Cardiocutaneous syndrome (Naxos disease and its variant) is genetic diseases caused by desmoplakin and plakoglobin genes mutation, and usually manifest with palmoplantar keratoderma, woolly hair and cardiomyopathy, and are found to have a high risk for uncontrolled arrhythmia.
Method: a cross-sectional retrospective cohort study included 10 Saudi children with clinical manifestation of Naxos disease variant. The medical records of the patients were analyzed. Echocardiography parameters (for ventricular function assessment); electrocardiography (ECG) and 24 hours Holter (for arrhythmias) and genetic analysis results were collected.
Results: We report 10 Saudi children with cardiocutaneous syndrome who presented with manifestation of severe dilated cardiomyopathy. All the patients had woolly hair, and half of them had also palmoplantar keratoderma. They all had severely dilated and de-pressed left ventricular systolic function, and nine of them had also depressed right ventricular systolic function. Frequent premature ventricular tachycardias (PVCs) were reported in nine cases, and an implantable cardioverter defibrillator (ICD) was im-planted for 3 patients for uncontrolled ventricular tachycardias. Moreover, 4 patients underwent heart transplantation, and 3 died suddenly while waiting for heart donation. Finally, in 8 patients, genetic studies were homozygous for the desmoplakin gene (DSP), confirming the diagnosis.
Conclusions: DSP gene mutation found to be the main genotype for Saudi Arabi patients with cardi-ocutaneous syndrome (mainly Naxos disease variant) and it is associated with high risk of arrhythmia and sudden cardiac deaths, so family members of proband need

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exten-sive genetic work up for identification of gene carriers for counselling, especially in our Arab countries where consanguineous marriage is common. Moreover, hair and skin phenotype in a child should alert for signs of cardiomyopathy manifestation.

Keywords: Naxos disease, Woolly hair, palmoplantar keratosis

PP-694

Persistent pulmonary hypertension in an infant. A congenital parvovirus B19 infection or related to a novel vars2 variant?

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Background and Aim: Congenital Parvovirus-B19 infection could potentially be a life-threatening condition in neonates. We describe a case, combined with persistent pulmonary hypertension and possible VARS2-related mitochondrial disorder.
Method: A full-term neonate, with birth weight (2.6 kg-3rd centile) and Apgar score (1':8, 5':9) developed respiratory distress a few hours after birth, requiring intubation and mechanical ventilation.
Results: The first echocardiogram ruled out major congenital heart defects, but revealed high suprasystemic pulmonary pressures, D-shaped left ventricle and right to left shunt through a patent foramen ovale. Treatment for pulmonary hypertension was initiated. The neonate continued to suffer from pulmonary hypertensive crises and desaturation episodes, that were difficult to control with pharmacotherapy. Thereafter, progressive right ventricular remodeling was observed: right ventricular hypertrophy (RVH) and heart displacement in the middle. Control of the pulmonary hypertension was achieved in the first month of life and sildenafil was stopped. Due to concomitant anemia (requiring multiple transfusions), failure to thrive and unexplained episodes of fever with negative cultures, the patient underwent screening test for infections. The PCR for Parvovirus-B19 was positive in both the mother and the neonate. However, serology testing revealed negative Parvovirus-IgM and positive IgG antibodies, indicating a recent infection (possible in third trimester of pregnancy). At the age of 2 months, the echocardiogram revealed recurrence of high right ventricular pressures, so sildenafil was restarted. Then, a gradual decrease in pulmonary pressures was observed and complete resolution was achieved, after the age of six months. However, RVH remained similar with no progress. Further investigation with whole exome sequencing revealed compound heterozygosity for two novel VARS2 variants: c.1587G>C (p.Gln682*) and c.2044C>T (p.Trp529Cys). The first variant is classified as likely pathogenic and the second one as a variant of unknown significance. Functional studies in muscle biopsy are currently underway to confirm the diagnosis.
Conclusions: Pathogenic and likely pathogenic variants in the VARS2 gene are associated with a mitochondrial disorder that could potentially cause hypertrophic cardiomyopathy and pulmonary hypertension in early infancy. Nevertheless, the genotype/phenotype correlation still remains unclear. Our case reflects the potential role of a crucial viral infection during late pregnancy/neonatal period as a triggering factor.

Keywords: persistent pulmonary hypertension, parvovirus infection, VARS2 gene

PP-695

Surviving child with malignant myosin heavy chain mutation of restricted cardiomyopathy three years after cardiac transplant

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Background and Aim: Restrictive malignant cardiomyopathy in infants is characterized by diastolic dysfunction caused by an impaired ventricular filling with increase of ventricular pressure and atrial dilatation. It is a very rare form of cardiomyopathy in infants with a very early onset and very short evolution to death. We report 3 years surviving after transplant child, with a rare and malignant restrictive cardiomyopathy secondary to a de novo mutation of the cardiac myosin heavy chain gene MYH7.
Method: The patient had clinical examination, electrocardiogram, Doppler echocardiography, blood sample, catheterize. Informed consent was signed.
Results: Family history of both paternal side were unremarkable. Healthy 8 years old daughter. Pregnancy and deliver without problems. Full term male new-born.
Clinical aspects: At 2 months of age was examined for systolic murmur. Biventricular not obstructive hypertrophic cardiomyopathy was detected in Doppler echocardiography. Differential diagnosis was made, all metabolic diseases discarded. At 6 month of age hypertrophy cardiomyopathy evolved to restrictive type and genetic study was done.
Genetics: A de novo mutation in heterocigosis of the beta Cardiac myosin heavy chain gene MYH7 was detected. (NP_000248.2p. Pro838Leu NM_000257.3: c.2513C>T NC_000014.8: g23894144G>A. This mutation change the protein structure and also its function. Clinical deteriorating was observed in the following months. Transthoracic Doppler Echocardiogram showing hypertrophic dysfunctional systolic and diastolic ventricles and enlarged atriums (fig 1) ProBNP elevating. ECG enlarging P waves and repolarization abnormalities with inverted T waves in V5 and V6. Treatment with carvedilol and acetyl salicylic acid began at 7 months old and captopril, sildenafil, at 12 months. At 13 months old after confirming catheterize he was included in transplantation list. At two years old, cardiac orthotropic transplant with bicave technique was performed. After 3 years of follow-up he is in functional class I, with tacrolimus and aziatropina treatment. No rejection was detected.
Conclusions: This MYH7 de novo mutation has only been reported in three families with severe prognosis. This child survives after 3

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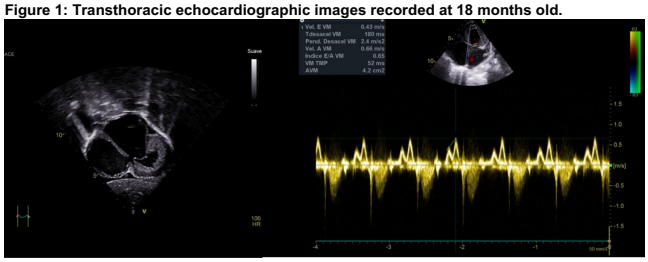
years of cardiac transplantation. Genetic mutation must be early suspected in infants with deteriorating restrictive cardiomyopathy. Nowadays transplantation is the only option for this patients.

Keywords: Restrictive cardiomyopathy, myosin heavy chain, MYH7, mutation.

Figure 1: Transthoracic echocardiographic images recorded at 18 months old.



(A) Subcostal view showing enlarged atriums and restricted hypertrophic ventricles. (B) Mitral Doppler showing Inverted E/A.

PP-697
Hereditary connective tissue disorders and qtc interval prolongation: Report of 2 families

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Background and Aim: INTRODUCTION
QTc interval prolongation increases the risk for life-threatening arrhythmias. QTc prolongation has been described in patients with connective tissue disorders (CTD), such as Marfan (MFS) and Loeys-Dietz (LDS) syndromes. However, this relationship is still not well established. We describe two families with concomitant CTD and prolonged QTc interval.
Method: CASE REPORT FAMILY 1. A 14-year-old girl, the first child of non-consanguineous parents. The mother has a mitral valve prolapse (MVP) and myopia. At 11 years, she was admitted to the Paediatric Cardiology Clinic due to the diagnosis of LDS type 4 (maternally inherited 4Mb deletion in 1q41 encompassing the "*"TGFB2"*" and "*"KCTD3"*" genes). Genetic testing had been performed in the context of intellectual disability, joint laxity and facial dysmorphisms. She has MVP with moderate regurgitation without aortic root dilation. A 24-hour Holter revealed Long QT Syndrome (LQTS). Magnetic resonance angiography revealed no cerebral, thoracic or abdominal arterial aneurysms to date. A Next Generation Sequencing (NGS)-based panel for LQTS is underway.
Results: FAMILY 2 Two sisters born to non-consanguineous healthy parents were observed in the Paediatric Cardiology Clinic. Prolonged QTc was detected in the younger (13 years old) in routine sports medicine examination, posteriorly confirmed in 24-hour Holter. The NGS-based panel for LQTS was negative. Her phenotype (pectus excavatum, scoliosis, joint laxity and flat feet) is suggestive of MFS, which was molecularly confirmed (heterozygous likely pathogenic variant c.7754T>C in the "*"FBN1"*" gene, maternally inherited). The older sister (21 years old) had the same familial variant, confirming the MFS diagnosis. 24-hour Holter revealed QTc prolongation as well; therefore, a beta-blocker was prescribed. Both sisters are under clinical surveillance.
Conclusions: In Family 1, the 1q41 deletion encompasses a gene encoding a protein that regulates potassium channels ("*"KCTD3"*") yet unrelated to LQTS, presumably sensitive to loss-of-function. In Family 2, an alternative diagnosis was not established. This work enlightens the association between CTD and QTc prolongation, a known risk factor for sudden death. Therefore, recognizing this potential association may have implications for the management and follow-up of these patients and families. Further studies are needed to substantiate this theory.

Keywords: Connective tissue disorders, Marfan syndrome, Loeys-Dietz syndrome, QTc interval prolongation.

PP-698
Williams-beuren syndrome – a case report with a fatal outcome

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Background and Aim: Introduction. Williams-Beuren Syndrome (WS) is caused by a recurrent 7q11.23 contiguous gene deletion involving the elastin gene, with an autosomal dominant inheritance. In 80% of cases, WS is characterized by cardiovascular disease, commonly presenting as supravalvar aortic or pulmonary stenosis. We describe a case of a 4-month-old infant with WS experiencing sudden cardiac death.
Method: CASE REPORT. This case involves the second child of non-consanguineous healthy parents (GII PII), born at 37 weeks of gestation. The infant was evaluated by paediatric cardiology at 8 days old because of a murmur, being diagnosed with mild supravalvar pulmonary and aortic stenosis. Some WS facial features were recognized. During follow-up, supravalvar stenoses worsened, leading to severe pulmonary and aortic stenosis with left ventricular hypertrophy at 3 months old. Genetic evaluation confirmed the diagnosis of WS (del 7q11.23).
Results: CASE REPORT. At 4 months, the infant experienced an episode of cyanosis, pallor, and hyporeactivity, which demanded an observation at the Paediatric Emergency Department. The electrocardiogram showed sinus tachycardia associated with diffuse repolarization abnormalities in the left and inferior member leads, with a supravalvar aortic echocardiographic gradient of 109 mmHg, with preserved ventricular function. A cardiac computed tomography angiography scan was requested, and a cardiac catheterization was scheduled for the next day. Unexpectedly, he presented a cardiac arrest. Despite initial revival, subsequent arrests in the Paediatric Intensive Care Unit were irrecoverable, resulting in the infant's demise. Autopsy findings confirmed severe aortic supravalvar and pulmonary artery stenosis. Additionally, diffuse coronary artery stenosis and biventricular concentric hypertrophy, associated with myocardial ischemic lesions, were observed. Histologically, the arterial intima and media of the aorta and coronary arteries were thick and associated with disorganized and fragmented elastic fibres (figure 1).
Conclusions: WS's elastin anomaly, causing arterial narrowing, is widespread. The literature describes WS-related coronary ostial

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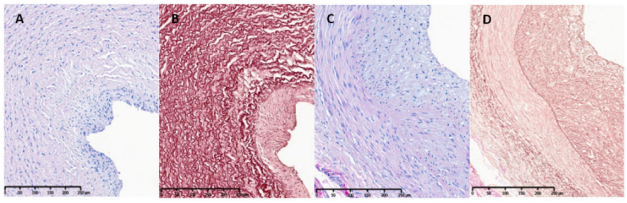
stenosis leading to life-threatening myocardial ischemia in infancy and childhood. Unexpectedly, this case emphasizes the need for heightened suspicion and prompt treatment. Tragic events, like cardiac arrests, may be triggered by diagnostic or therapeutic procedures requiring anaesthesia, highlighting WS as a risk factor for sudden cardiac death.

Keywords: Williams-Beuren Syndrome, elastin anomaly, coronary arteries, cardiac arrest.

Figure 1



Histological findings in the aorta (A, B) and coronary arteries (C, D).

PP-699

Childhood hypertrophic cardiomyopathy caused by thin filament sarcomeric variants

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Background and Aim: Hypertrophic cardiomyopathy (HCM) caused by variants in thin filament proteins represents up to 20% of childhood sarcomeric HCM, but data on genotype-phenotype correlations are limited. This study describes the natural history and outcomes of children with thin filament associated HCM and compares it to thick filament associated disease.

Method: A retrospective, longitudinal cohort of children ≤ 18 years evaluated at Great Ormond Street Hospital Centre for Inherited Cardiovascular Disease 1991- 2019 with a disease-causing variant in a thin-filament gene was established. Clinical data on presentation, symptom status, medications, echocardiography, electrocardiographic and cardiac magnetic resonance imaging data were collected. Primary outcome was a Major Adverse Cardiac Event (MACE).

Results: Of 40 children with a disease-causing thin-filament variant, 21 (female n=6, 35.5%) were diagnosed with HCM at a median age 13.0 years (IQR 8.3-14.0). Seven patients (33.3%) had an additional sarcomeric gene variant and were more likely to be diagnosed in infancy (n=2 vs n=0, p=0.042). Nine patients (42.9%) were symptomatic at baseline. One patient, with an additional variant of unknown significance (VUS) variant in ACTC1, had left ventricular outflow tract obstruction (LVOTO). Over a median follow up 5.0 years (IQR: 4.0 – 8.5), 3 (14.3%) experienced one or more Major Adverse Cardiac Event (MACE) (out of hospital arrest n=2, appropriate Implantable Cardiac Defibrillator (ICD) therapy n=8). One gene carrier died suddenly aged 9 years. Compared to a cohort of thick-filament disease (MYH7 n=68, MYBPC3 n=62), children with thin filament variants more commonly experienced NSVT (n=6 (28.6%) vs n=14 (10.8%), p=0.024) or underwent ICD insertion (thin, n=13 (61.9%) vs thick, n=50 (38.5%), p=0.040). However, there was no difference in the incidence of MACE [thin 2.47/100 pt years (95% CI 0.80 – 7.66) vs thick 3.63/100 pt years (95% CI 2.25 – 5.84)] or an arrhythmic event [thin 1.65/100 pt years (95% CI 0.41-6.58) vs thick 2.55/100 pt years (95% CI 1.45 – 4.48), p value 0.43)].

Conclusions: This study suggests adverse events in thin-filament disease are predominantly arrhythmic, and can occur in the absence of hypertrophy, but overall short term outcomes do not differ significantly from thick-filament disease.

Keywords: childhood hypertrophic cardiomyopathy, sarcomeric cardiomyopathy, thin filament variants

Comparison of characteristics at baseline of patients with thin- and thick-filament disease.

	Thin filament (n=21)	Thick filament (n=130)	P-value
Age at diagnosis (median)	13.0 (IQR: 8.3 – 14.0)	10.6 (IQR: 5.0 – 14.1)	0.361
Symptoms at baseline	9 (43.9%)	51 (39.2%)	0.753
Medication at baseline	10 (47.6%)	48 (36.9%)	0.350
Baseline echocardiogram			
LVOT gradient ≥ 30 mmHg	1 (4.8%)	18 (13.8%)	0.244
Median LVOT gradient (mmHg)	6.0 (IQR: 5.0 – 10.0)	8.0 (IQR: 5.0 – 14.5)	0.257
MLVWT (median, IQR) at baseline	15.0 (IQR: 9.5 – 25.5)	15.5 (IQR: 11.0 – 23.)	0.864
MLVWT Z-score (median, IQR) at baseline	7.6 (IQR: 1.5 – 16.7)	9.6 (IQR: 5.3 – 15.9)	0.227
LA diameter, mm (mean +/- SD) at baseline	28.7 ± 9.3	32.1 ± 8.0	0.390
LA Z-score (mean +/- SD) at baseline	-0.3 ± 3.0	1.6 ± 2.1	0.035
Outcomes			
Median age at last follow up	18.0 (IQR: 15.0 – 20.0)	15.8 (IQR: 11.4 – 17.4)	0.009
ICD insertion	13 (61.9%)	50 (38.5%)	0.043
Myectomy	1 (4.8%)	9 (6.9%)	0.712
OOHCA	2 (9.5%)	8 (6.2%)	0.564
NSVT on ambulatory ECG	6 (28.6%)	14 (10.8%)	0.026
MACE	3 (14.3%)	18 (13.8%)	0.957
Death	0	8 (6.2%)	0.243
Cardiac transplantation	0	5 (3.8%)	0.361

LVOT – Left Ventricular Outflow Tract, MLVWT – Maximal Left Ventricular Wall Thickness, LA – Left Atrium, ICD - Implantable Cardiac Defibrillator, OOHCA - Out Of Hospital Cardiac Arrest, NSVT - Non-sustained Ventricular Tachycardia, MACE - Major Adverse Cardiac Event. Values expressed as median with interquartile range (IQR) or mean with standard deviation (SD).

PP-700

Notch1 pathogenic variants in tetralogy of fallot

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Background and Aim: Tetralogy of Fallot (TOF) is the most common form of cyanotic congenital heart disease, characterized by a ventricular septal defect, over-riding of the aorta, right ventricular outflow obstruction, and right ventricular hypertrophy. The pathophysiology of TOF is complex with both genetic and environmental contributions, and the etiology remains unknown in the majority of the patients. Notably, pathogenic variants (PVs) in *NOTCH1* have been identified as a molecular cause of bicuspid

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aortic valve (BAV), and more recently with TOF. Here we focus on *NOTCH1* variation to further unravel the molecular etiology of TOF.
Method: In 32 probands with isolated TOF and normal copy number variant analysis, we performed trio exome sequencing (ES) on genomic DNA followed by analysis of a virtual congenital heart disease gene panel including 471 genes. *NOTCH1* (NM_017617.5) variants were classified according to the American College of Genetic and Genomic Medicine (ACMG) guidelines.
Results: We identified three heterozygous (likely) PVs and one suggestive variant of unknown significance in *NOTCH1* (12.5% of total TOF cohort). Index 1 presented with TOF with pulmonary valve atresia (PA) and harbored a splice-site PV in *NOTCH1*, c.5693-1G>A. Notably, the variant was inherited from the seemingly asymptomatic father. Index 2, also presenting with TOF with PA harbored a de-novo p.Cys74Tyr PV in *NOTCH1*. Index 3 showed TOF with a pulmonary valve stenosis (PS) and a right-sided aortic arch and showed a p.Asn802ThrfsTer68 PV in *NOTCH1*, with unknown inheritance (parental DNA unavailable). Finally, index 4 showed a TOF with PS and persistent left superior vena cava. He harbored a p.Gly1476Asp *NOTCH1* missense variant (GnomAD v4: 3/10^6, in-silico predicted pathogenic, ACMG criteria: PM2_PP, BP1, PP3, PP1), inherited from his mother and maternal grandmother, both diagnosed with BAV.
Conclusions: CONCLUSION: Our data indicate that PVs in *NOTCH1* are an important contributor to the etiology of TOF. The observation of both TOF and BAV in one family and the occurrence of non-penetrance argues for additional modifiers of phenotypic variability.

Keywords: Tetralogy of Fallot, NOTCH1, Genetics

PP-701
Danon disease: a case report emphasizing the importance of genetic analysis in diagnosis and interprofessional management

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Background and Aim: Danon disease (DD) is a rare x-linked dominant multisystemic disorder characterized by a lysosomal-associated membrane protein-2 (LAMP2) gene defect. The clinical phenotype includes severe cardiomyopathy, skeletal myopathy, and mental retardation, with additional systemic involvement reported in males. This case report presents an 8-year-old male diagnosed with DD, emphasizing the significance of genetic analysis in confirming the diagnosis and guiding appropriate therapeutic interventions.
Method: At 8 months of age, the patient was diagnosed with hypertrophic cardiomyopathy (HCM), muscle lysis, and hepatocytolysis. Extensive investigations targeting storage diseases, neuromuscular disorders, and metabolic abnormalities yielded negative results. The patient, currently asymptomatic, has been maintained on Propranolol since the HCM diagnosis. Fluctuations in hepatocytolysis and muscle enzyme levels have been observed, necessitating liver protectors. Cardiac evaluations consistently reveal left ventricular hypertrophy, medioventricular and left ventricular outflow tract gradients, and mitral valve systolic anterior motion. Genetic testing at the age of 8 identified a hemizygous nonsense variant LAMP2 c. 877C>T p.(Arg293*), confirming the diagnosis of Danon disease.
Results: The clinical presentation of this case underscores the pivotal role of genetic analysis in diagnosing Danon disease accurately. Despite the absence of syncope and a low 0.1% risk of HCM-related complications at 5 years, the patient's genetic profile necessitates ongoing monitoring and appropriate management. The interprofessional team's collaboration, comprising cardiologists, geneticists, and other specialists, is imperative for comprehensive evaluation and treatment.
Conclusions: In conclusion, Danon disease presents a complex clinical scenario, necessitating a multidisciplinary approach for accurate diagnosis and optimal management. Genetic analysis stands as the gold standard, guiding treatment strategies and assessing the genetic risk for family members. This case report emphasizes the critical role of the interprofessional team in evaluating and treating this rare multisystemic disorder.

Keywords: Danon disease, lysosomal-associated membrane protein-2, hypertrophic cardiomyopathy, genetic analysis, interprofessional management.

PP-702
Rare case of simultaneous aortic and chest surgery in patient with loeys-dietz syndrome

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Background and Aim: The Loeys-Dietz syndrome is a rare connective tissue disorder that affects the cardiovascular system resulting in the occurrence of arterial aneurysms. The condition is genetically determined, caused by the mutation in the TGF-β signaling pathway. The syndrome is characterized by clinical findings eg. craniofacial abnormalities including bifid uvula, cleft palate and hypertelorism. The skeletomuscular deformities such as pectus excavatum/carinatum, pes planus/clubfeet are also characteristic. The presence of both aortic aneurysm and chest deformation can escalate the risk of premature mortality. The aim of this study is to show the case report on the rarity and remarkableness of simultaneous surgical procedures including David surgery regarding aortic aneurysm and Nuss method chest surgery.
Method: Retrospective data collection on the case of 9-year-old boy with genetically confirmed Loeys-Dietz syndrome was performed. The patient was under the care of the Pediatric Cardiology and Congenital Heart Defect Department since the

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age of 4. In the echocardiography the aortic dilation was diagnosed, with the aortic root size of 26 mm (Z-score +3,36), atrial septal defect II and biscupid aortic valve (BAV). At the age of 8, significant dilatation of the distal part of the ascending aorta was shown- 45 mm (Z-score +7,76), BAV with moderate valve insufficiency and hemodynamically non relevant PDA. Additionally, patient had pectus excavatum, in CT scan the aortic aneurysm was adjacent to the sternum.
Results: Due to the significant aortic dilatation and fast progression comparing to previous examination, the patient was qualified for the cardiosurgery. David procedure was performed. As a result of pectus excavatum being close to heart and main vessels, the surgery on correcting the sternum alignment with metal tile (Nuss procedure) was performed immediately after aneurysm surgery
Conclusions: The shut of the sternum after cardiological surgery without Nuss procedure could be impossible or could lead to accrete of both bone and heart structures, which would forbid opening the chest in the future. Performing chest surgery before cardiological surgery could have led to vessels damage. Due to the reasons stated above simultaneous David and Nuss procedures were performed.

Keywords: Loeys-Dietz, connective tissue, David procedure, Nuss procedure, pectus excavatum

PP-703
Efficacy and safety of ace-inhibitors in pediatric hypertrophic cardiomyopathy
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Background and Aim: The course paediatric hypertrophic cardiomyopathy (HCM) is highly variable. Symptoms of heart failure are predominantly due to left ventricular diastolic dysfunction. Some studies suggest that ACE-inhibitors (ACEi) could improve hypertrophy, diastolic function, and fibrosis in HCM. The aim of this study is to demonstrate the efficacy and safety of the use of ACEi in pediatric HCM.
Method: This is a single centre retrospective study. A total of 241 pediatric patients were evaluated. Inclusion criteria were age < 18 years; diagnosis of hypertrophic cardiomyopathy; increased diastolic pressure at echocardiogram or NTproBNP>600pg/ml and treatment with ACEi. Data were collected at baseline, after 3 months, 6 months and 1 year of treatment with ACEi.
Results: A total of 45 patients meet inclusion criteria and were enrolled in the study (42% male, mean age 13.9+/-5.8 years, mean FU 2.4+/-1.6 years). 40% were primitive, 13% mytochondrial, 36% syndromic, 11% idiopathic, 29% showed obstruction, 46% of patients were in NYHA I, 47% in NYHA II and 7% in NYHA III. Main wall thickness (MWT) was 9.4 +/- 7 Boston z-score (range 2-34); main ventricular mass was 85 +/- 48 (n.v.< 45); main diastolic filling pressure E/E' was 10.3 +/- 4 (range 2-21); main NTproBNP level was 3338 +/- 4935 pg/ml. After starting treatment, NTproBNP showed a trend towards reduction (main value was 3187pg/ml, 2977pg/ml, 2558pg/ml and 1845pg/ml at 3, 6 months, 1 and 3 years). MVT slightly decrease to 7.4 +/- 5 (-2 points of z- score), ventricular mass decreased to 69 +/- 21. E/E' values remain stable (main value of 9.9 +/- 3), good responders were patients with diastolic pressure >15. NYHA class improves (69% Class I, 24% class II, 7% class III). No adverse events in term of hypotension or renal dysfunction were detected during FU and main LV outflow gradient remain stable (39.5 +/- 21.9 mmHg and 38.8 +/- 23.5 before and after treatment).
Conclusions: • The use of ACEi in pediatric patients with HCM, seem to be effective in reducing NTproBNP levels, NYHA class and ventricular mass. • Treatment with ACEi was safe and no adverse events or increasing in LV outflow gradient were detected during FU.

Keywords: Hypertrophic Cardiomyopathy, Ace-inhibitors, Treatment, Pediatric

PP-704
RISk factors for adverse outcome in pediatric dilated cardiomyopathy
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Background and Aim: Paediatric dilated cardiomyopathies (DCM) are a heterogeneous group of disorders, including inherited pathogenic variants, inborn errors of metabolism, neuromuscular disorder, inflammation and toxic cause. The outcome of DCM in children is highly variable, ranging from complete recovery to heart transplantation (HT) or death. In this study we aim to identify risk factors for adverse outcomes in a large cohort of pediatric patients affected by DCM.
Method: This is an observational retrospective multicentre study. We enrolled all children with DCM diagnosis referred to Bambino Gesù Children's Hospital (Rome) and Monaldi Hospital (Naples). We analysed data regarding survival (defined as freedom from cardiac death or heart transplantation) through Kaplan Meier (KM) curves and Cox Regression analysis.
Results: We collected 141 pediatric DCM patients, M 59%, mean age at diagnosis was 6.59 +/- 6.57 years (30% diagnosed in the first year of life). 68% were primitive form, 33% idiopathic and 12.9% secondary. At presentation, 21% of patients were equally in NYHA class I and II, 25% in class III and 20% in class IV; LVEF was < 40% in 62% of patients; moderate and severe mitral valve regurgitation (MVR) were detected in 25% and 15% respectively; inferior cava vein (ICV) was dilated in 15%; mean NTproBNP value was 4780 +/- 8570 pg/ml; 44% patients were treated with inotropes, whilst 52% received oral chronic therapy. The KM analysis showed a survival of 86% at 1 year of age; of 60% at 14 years and of 30% at 20 years of age. Univariable analysis identify as risk factors for adverse outcome: female sex, younger age at diagnosis, primitive or idiopathic form, advanced NYHA functional class, lower EF, moderate-severe MVR. At the multivariable analysis the only independent factors for death or HT were lower age at diagnosis, severe LV dysfunction and NYHA class IV at presentation.

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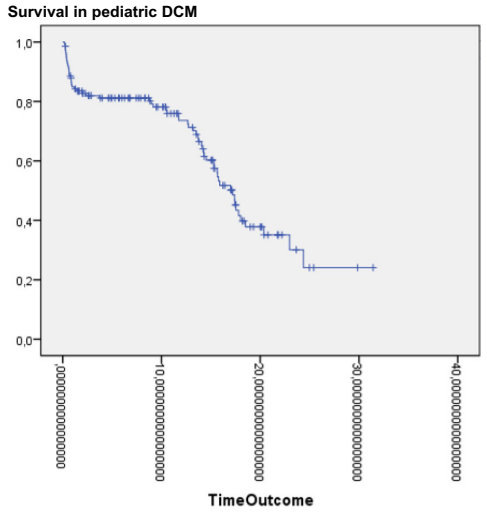
Conclusions: Data from this study showed as survival decrease significantly in the first year of age and after adolescence. Lower age at diagnosis, severe LV dysfunction and NYHA class IV at presentation may represent important risk factors for adverse outcome in pediatric DCM.

Keywords: Dilated Cardiomyopathy, Risk Factors, Adverse Outcome, Pediatric



Overall survival and freedom from adverse events in pediatric DCM, represented by Kaplan Meier curve.

PP-705
Exercise testing in children with rasopathy-associated hypertrophic cardiomyopathy

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Background and Aim: Exercise testing (ETT) either in the form of traditional blood pressure, ECG, and symptom assessment, or cardiopulmonary exercise testing (CPET) which combines this with ventilatory expired gas analysis, is known to provide enhanced information on the severity of hypertrophic cardiomyopathy (HCM) and the mechanisms of the patients' symptomatology. The data on children is limited, with no data on syndromic aetiologies of HCM. To describe the baseline ETT in children with RASopathy-associated HCM
Method: Retrospective multi-centre (UK, Munich, Naples) cohort of children with a diagnosis of HCM secondary to an overarching diagnosis of a RASopathy syndrome presenting in a paediatric cardiology centre and undergoing ETT/CPET.
Results: Of the 49 patients identified, 15 (30.6%) underwent simple ETT and the remaining 34 (69.4%) a CPET, at a median age of 11.4 years (IQR 8.88-13.68). Thirty-five patients (71.4%) had a diagnosis of Noonan syndrome (NS), the remaining Noonan syndrome with multiple lentigines (NSML). Seventeen patients (34.7%) had a variant in PTPN11, 9 (18.4%) RAF1, 3 (6.1%) RIT1, 2 (4%) LZTR1 and one (2%) MEK 2. The majority were asymptomatic (N=35, 71.4%), while on medication (N=26, 53.1%), primarily b-blockers (N=23, 46.9%). Nine patients (18.4%) displayed submaximal effort [RER 1.04 (IQR 0.89-1.15)] with a median peak HR of 100.1% (IQR 85.3-100.4) of predicted. Most patients had an abnormal BP response to exercise [flat N=22 (44.9%), hypotensive N=5 (10.2%)]. The median VO2 consumption was 32mL/(min*kg) (IQR 25.8-39) and VE/VO2 slope was 33 (IQR 29.3-35.1). There was no statistically significant difference in ETT/CPET performance by syndrome. Over a median follow up of 195.7 months (IQR 116.6-216), 2 patients (4.2%) died, 15 patients (31.3%) underwent myectomy, 3 patients (6.3%) were admitted to hospital due to congestive heart failure symptoms, 5 patients (10.4%) had an implantable defibrillator/cardioverter inserted and 3 (6.3%) suffered a major arrhythmic cardiac event. No predictors of outcome were identified from the ETT/CPET parameters.
Conclusions: We describe the baseline ETT/CPET in paediatric RASopathy-associated HCM, demonstrating overall low normal cardiovascular fitness with mildly impaired ventilation response, which are likely multifactorial. Further and larger studies are needed to best understand the correlation with phenotype and clinical implications.

Keywords: exercise test, Noonan syndrome, hypertrophic cardiomyopathy, paediatric, genetics

PP-706
Cardiac magnetic resonance imaging in children with noonan syndrome and associated hypertrophic cardiomyopathy

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Background and Aim: Cardiovascular magnetic resonance imaging (CMRI) provides important information on the diagnosis and risk stratification of hypertrophic cardiomyopathy (HCM) in adults with limited data in children and absent data in syndromic aetiologies. To describe the baseline CMRI in children with Noonan syndrome (NS).
Method: Retrospective multi-centre (UK, Naples) cohort of children with a diagnosis of HCM secondary to an overarching diagnosis of a NS presenting in a paediatric cardiology centre and undergoing CMRI.
Results: Fourty patients with NS and HCM who underwent a CMRI at a median age of 13.4 years (IQR 9.3-16.4) were identified, with 28 (70%) being males. Eleven (27.5%) patients had a variant in PTPN11, 10 (25%) in RAF1, 4 (10%) in RIT1 and one (2.5%) in LZTR1, MEK2, and HRAS respectively. The

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majority of patients (N=21, 52.5%) displayed an asymmetric pattern of left ventricular (LV) hypertrophy with 10 (25%) having concomitant right ventricular (RV) hypertrophy. The mean maximal LV wall thickness was 18mm (SD 8.7mm), most commonly at the basal septum (N=17, 42.5%) with an overall normal sized LV (N=11, 73.3%) with hyperdynamic systolic function (N=25, 62.5%). The mean LV mass was 116.4g (SD 82.1g). Seven (17.5%) patients had late gadolinium enhancement (LGE) present, all patchy and in the hypertrophied segments. No statistically significant differences were found by gene variant identified.
Conclusions: We present the baseline characteristics of CMRI in patients with NS-associated HCM. Further and larger studies are needed to identify its correlation with the phenotype and clinical outcomes.

Keywords: cardiac magnetic resonance imaging, Noonan syndrome, hypertrophic cardiomyopathy, paediatrics, genetics

PP-709

Demographics of samples in a north indian congenital heart disease biorepository

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Background and Aim: Congenital Heart Defects (CHDs) are most common birth defects with prevalence in Asia of about 9.3 per 1000 live births(Saxena A. et al., 2018). Every year around 2 lakh children are born with this disease. Critical CHD cases with a birth prevalence of 1.5-1.7/1000 live births do not survive till first year of life and hence are not accounted for in prevalence studies(Saxena A 2018). Children born with CHD face considerable challenges including lifelong disabilities like retarded growth, nutrition, delayed developmental milestones, inability to perform normal activity, heart rhythm problems, sudden cardiac arrest, stroke, breathing problem and many more leading to poor quality of life. Several factors accounted by lack of awareness, delayed diagnosis, improper distribution of resources, financial constraints hinder paediatric cardiac care to reach the masses (Saxena A. et al., 2018).
Method: All the patients undergoing surgery in the dedicated tertiary pediatric cardiac hospital were considered for recruitment into the study. Blood and tissue samples from patient and parents were collected with informed consent for genetic studies. Demographic Data was gathered in Redcap software with self declared retrospective history recall for the disease. Blood samples for more than 4000 CHD cases and 600 heart tissue samples were collected in a span of 4 years.
Results: 50% of the patients were from the state of Uttar Pradesh. 90% of the samples were nonsyndromic with 75 familial cases and 20 one affected twin pairs. 50% of the CHD cases were first borns with 56% being males. 30% of the mothers did not take any iron folic acid supplementations during pregnancy. The most common CHD types were Ventricular Septal Defects, Atrial Septal Defect and Tetrology of Fallot. The most commonly observed symptoms included frequent coughing and breathing difficulty.
Conclusions: This is the only dedicated congenital heart disease biorepository in north India with a ever growing patient load. In such scenario it is a repository of immense value to do research on CHDs in the growing Indian papulation.

Keywords: Congenital Heart Disease, North India, Demographics, Disease Burden

PP-710

Role of MIR-122 in pathogenesis of congenital heart defects during pregestational diabetes

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Background and Aim: Pregestational maternal diabetes increases the risk for congenital heart defects (CHDs) in infants by over four times. Diabetes may alter maternal/fetal microRNA (miRNA) profiles leading to the pathogenesis of CHDs. An elevation of miR-122 has recently been reported in patients with impaired glucose tolerance, insulin resistance and obesity. miR-122 is highly abundant in the liver and acts as tumor suppressor. The aim of the present study was to determine if miR-122 is elevated in embryonic hearts from diabetic female mice, and to examine its role in pathogenesis of CHDs during pregestational diabetes.
Method: Diabetes was induced by streptozotocin to adult female C57BL/6 mice. Diabetic females were treated with a locked nucleic acid (LNA) antimiR-122 or scramble LNA control (10 mg/kg, SC x2), and their offspring's hearts were examined at E18.5 for morphology and function. Gene expression in the fetal heart was assessed by RT-qPCR analysis.
Results: miR-122 expression was upregulated in E12.5 hearts of offspring from diabetic dams. In cultured E12.5 hearts, treatment with miR-122 or high glucose inhibited cell proliferation and epicardial EMT, and increased apoptosis. These effects of miR-122 and high glucose were abrogated by antimiR-122 transfection. Downregulation of genes critical to cell cycle progression, angiogenesis, and heart development, such as Cyclin D1, Snail1, Gata4 and Hand2 under high glucose conditions, was also prevented by anitmiR-122 transfection. Furthermore, in vivo antimiR-122 treatment to diabetic dams decreased the incidence of CHDs and improved cardiac function of fetuses compared to scramble LNA controls.
Conclusions: Upregulation of miR-122 results in CHDs in mice. Our study reveals for the first time a critical role of miR-122 in CHD pathogenesis in mice, and may have therapeutic implications in preventing CHDs during pregestational diabetes.

Keywords: Congenital heart defects, miR-122, pregestational diabetes

PP-711

Double stress of pressure and hypoxia attenuated mitochondrial damage in cardiomyocytes compared to single stress of pressure or hypoxia stress in RAT

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Background and Aim: Recent advances in the management of congenital heart disease have dramatically improved survival rates, particularly for cyanotic congenital heart disease (CCHD). On the

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other hand, residual stenotic lesions and cyanosis can lead to intractable heart failure requiring close management, mostly in the right ventricle (RV). Despite the accumulation of knowledge in RV failure, RV remodeling in CCHD remains unclear. Our aim is to assess histological and physiological changes in RV with hypoxia and pressure stressed rats.
Method: Three-week-old male Sprague-Dawley rats were kept in a hypoxic environment (13%), underwent pulmonary artery banding (PAB) at 4-week-old, and continued until 7-week-old. After exposure to hypoxia, echocardiography and catheterization were performed, and ventricles were harvested for histological analysis. To compare them, non-operated or normoxia rats, respectively. 2-way ANOVA was used for statistical analysis.
Results: Subjects were hypoxia with PAB (PBH) n=15, normoxia (PBR) n=17, hypoxia control without PAB (CH) n=11, normoxia control (CR) n=12. In PAB model, RV systolic pressures were similar (%RV/LV pressure; PBH76.9±4.8, PBR91.9±8.9, p=0.24), and the fibrosis rates (%) were similarly increased in single stress groups, but, in PBH group no further fibrosis was observed (PBR3.8±0.4, PBH4.4±0.3, CR2.9±0.3, CH4.4±0.7, p=0.02). In electron microscopy, mitochondria morphology showed the swelling and disruption due to pressure or hypoxia stress, however maintained the size in PBH (area (μm2) PBR0.85±0.16, PBH0.75 ±0.07, CH1.09±0.04, CR0.79±0.07, p=0.07).
Conclusions: Mitochondria are an organelle responsible for ATP production through oxidative phosphorylation and glycolysis in cells, and are particularly abundant in cardiomyocytes and essential for the maintaining of cardiac function. Excessive pressure or ischemic stress can cause injury, leading to fibrosis via mitophagy and heart failure. The present results showed that pressure or hypoxia stress caused mitochondrial injury, whereas double stress maintained mitochondrial morphology and no further fibrosis. These findings are possible to maintain and improve cardiac function in CCHD, and should be understood the mechanisms to potential therapeutic interventions.

Keywords: Right ventricle failure, Hypoxia, Mitochondria

PP-712
Exploring the 12-lead ecg characteristics of pediatric and young adult carriers of desmoplakin gene variants

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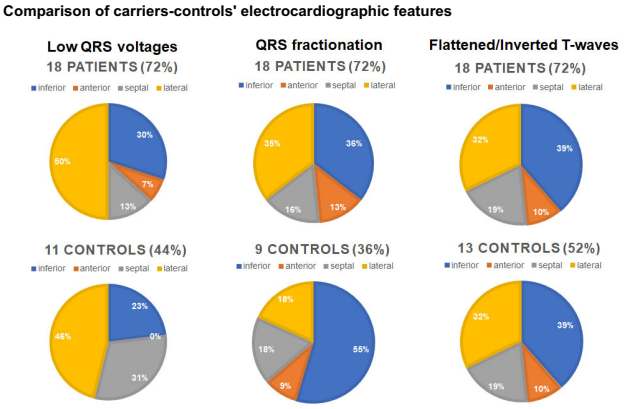
Background and Aim: The DSP gene (locus. 6p24.3) encodes desmoplakin, a quintessential desmosomal protein for cellular adhesion, particularly within cardiac tissues. DSP gene variants can cause dilated cardiomyopathy, non-dilated left ventricular cardiomyopathy (NDLVC) and arrhythmogenic righ ventricular cardiomyopathy (ARVC), cardiac conditions known to trigger malignant ventricular arrhythmias and cause sudden cardiac death in young individuals. Nevertheless, the electrocardiographic (ECG) patterns in children carrying DSP variants are underexplored.
Method: A retrospective analysis was conducted involving paediatric and young adults DSP gene variant carriers (</=20 years), followed at Great Ormond Street Hospital from 2006 to 2023. Their 12-lead ECGs were characterised and compared with those of an age and gender-matched healthy control group (χ^2, p-value<0.05).
Results: 25 DSP carriers (64% females; 12.6±4 years) were identified [including 4 (16%) with an overt cardiomyopathy phenotype] and compared to 25 healthy gender and age-matched controls (12.7±4.3years). Compared to the control group, DSP carriers exhibited a higher prevalence of low QRS voltages (72% vs 44%, p= 0.041), QRS notching (72% vs 26%, p= 0,014) and flattened/inverted T waves (72% vs 52%, p= 0,026). All 4 patients with an overt cardiomyopathy phenotype had widespread low QRS voltages and QRS fractionation, as well as flattened/inverted T waves, particularly in the inferior and lateral leads.
Conclusions: Predominant ECG features observed in paediatric patients with DSP gene variants included low QRS voltages, QRS notching, and flat or inverted T waves. Our findings suggest that ECG features may precede the development of an overt cardiomyopathy phenotype in children with DSP variants, but further studies with larger cohorts are needed to confirm this and to determine their prognostic significance.

Keywords: DSP mutation, arrhythmogenic cardiomyopathy, electrocardiogram, paediatric



QRS low-voltages, QRS fractionation and flattened or inverted T-waves distributions among the electrocardiographic leads in patients carrying DSP variants and gender and age-matched controls.

PP-713
Myocarditis or arrhythmogenic cardiomyopathy? which comes first? a difficult case

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Background and Aim: A characteristic feature of arryhthmogenic cardiomiomopathy (ACM) in paediatric patients is the presence of recurrent episodes of myocarditis like, the so-called "hot phase". Cases of ACM misdiagnosed as myocarditis, or conversely, have

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also been reported. The genetic defects in structural proteins of the cardiomyocyte could be responsible for an increased vulnerability to pathogen-induced myocardial inflammation, or conversely, the infectious-inflammatory trigger of the myocarditis could slatentize ACM in genetically predisposed individuals.
Method: We report the case of a 14-year-old patient with occasional findings of supraventricular and ventricular extrasystoles (PSVC, PVC). In family history, mother with resuscitated cardiac arrest undergoing implantable cardioverter defibrillator (ICD). The cardiac magnetic resonance (CMR) documented sub-epicardial late gadolinium enhancement (LGE) in the posterior septum: previous myocarditis was diagnosed.
Results: The girl underwent sports medicine examinations: at ECG, rare isolated monomorphic PVCs. In stress test appeared PSVC and isolated PVCs at the start of the test and in the recovery. Echocardiogram and cardiac enzymes were normal. CMR showed extensive subepicardial/intramyocardial LGE at the interventricular septum and the inferior mid-apical wall of the left ventricle, with a pattern suggestive of previous infectious-inflammatory phenomena. In consideration of the family history, a cardiogenetic evaluation was requested and genetic test resulted positive for desmoplakin mutation (DSP): c.5851C>T (p.Arg1951Ter), class 5, pathogenic, maternal segregation. Diagnosis of left-dominant ACM was made due to the presence of 3 major criteria: LGE on cardiac MRI, pathogenic mutation, non-sustained ventricular tachycardia (NSVT). At subsequent controls PVC and NSVT were detected and a S-ICD was implanted (class 2A).
Conclusions: -ACM is a rare condition in paediatric age.
-Paediatric onset may be most frequently associated with 'hot phase', mimicking myocarditis in terms of symptoms and clinical presentation.
-A scrupulous personal and family history and a critical review of instrumental data are necessary in order to identify the 'red flags' that direct the diagnostic suspicion.
- In patients with suspected diagnostic cardiomyopathy, it is necessary to consider genetic investigations, which are generally not routinely performed in myocarditis.

Keywords: myocarditis, arrhythmogenic cardiomyopathy, genetics, desmoplakin, PVC

PP-714
The role of cardiac magnetic resonance in paediatric patients with friedreich's ataxia

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Background and Aim: Friedreich's ataxia (FA) is a rare, autosomal recessive disease that is often diagnosed in childhood or adolescence. Progressive hypertrophic cardiomyopathy (HCM) and myocardial fibrosis are responsible of 60% of FA-related deaths. Currently, staging of HCM is based on ECG, high-sensitivity troponin T (hsTNT) and echocardiographic data. However, additional cardiac magnetic resonance (CMR) parameters could play a role in cardiac disease staging. The aim of this study is to describe the CMR characteristics of patients with FA.
Method: This is a single centre retrospective study including all consecutive patients with FA diagnosis admitted to Bambino Gesù Children's Hospital. Cardiac data were collected and analysed, including ECG parameters, maximum wall thickness (MWT) at echocardiogram, hsTNT values and CMR parameters.
Results: A total of 40 patients were evaluated, of whom 19 performed CMR (58% male, mean age 17.8 +/- 8.6 years). All patients showed mild to moderate concentric left ventricle hypertrophy. At the ECG mean heart rate was 80 bpm, 89% of patients presented abnormalities of ventricular repolarisation (usually infero-lateral negative T waves). At the echocardiogram, MWT was 10.8 +/- 3.2 mm; main left ventricular mass index (LVH) was 55,6 +/- 13 gr/m2; main ejection fraction (EF) was 61.4 +/- 8,9 %. At CMR, MWT calculated was 11.1 +/- 2.2 mm; LVH was 71,7 +/- 16 gr/m2 and main EF was 59.9 +/- 9.5 %. 8 out of 19 patients presented late gadolinium enhancement (LGE), 50% on the interventricular septum, and the other 50 % on the left ventricular lateral wall. Main hsTNT level was 46,7 +/- 53 pg/ml at CMR time.
Conclusions: Our study shows that the main MWT and LVH values are higher when detected at CMR compared to echocardiogram. Among the 8 patients presented with LGE at CMR, 2 showed mild reduction of EF, and one progressed to end-stage dilated cardiomyopathy. Thus, CMR in these patients seems able to detect the cardiac mass index more accurately and earlier than echocardiogram. The presence of LGE may be an indicator of cardiac disease progression and a risk factor for the develop of end-stage cardiomyopathy.

Keywords: CMR, Friedreich's ataxia, hypertrophic cardiomyopathy, LGE

PP-715
Impact of the 2023 european society of cardiology (ESC) guidelines for cardiomyopathies on the diagnosis of paediatric arrhythmogenic cardiomyopathies

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Background and Aim: The 2023 European Society of Cardiology (ESC) guidelines for the management of cardiomyopathies propose a diagnostic approach that emphasises the importance of systematically evaluating functional and morphological traits to accurately describe the myocardial phenotype at presentation. Distinct cardiomyopathy phenotypes, including dilated cardiomyopathy (DCM), arrhythmogenic right ventricular cardiomyopathy (ARVC) and non-dilated left ventricular cardiomyopathy (NDLVC), are described for patients who previously may have been classified under the umbrella term 'arrhythmogenic cardiomyopathy (ACM)'. It is currently unknown if these distinct phenotypes exist in paediatric populations. This study aimed to assess this new classification and explore the phenotype in affected paediatric patients.
Method: We describe a cohort of patients with a clinical diagnosis of dilated ACM or ARVC, attending a dedicated ACM clinic between 2015 and 2023. Data were retrospectively collected on

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baseline clinical characteristics and outcomes, including arrhythmic events [aborted cardiac arrest, sustained ventricular tachycardia (sVT) and nonsustained VT (NSVT)], implantable cardioverter defibrillator (ICD) implantation, cardiac death or heart transplantation.
Results: Of 661 patients seen in the ACM clinic, 23 (female n=14.6%) were diagnosed at a mean age of 14±3 years. Following phenotypic reclassification according to the new guidelines, 11 were reclassified as DCM, 6 possible ARVC, and 6 NDLVC. A pathogenic/likely-pathogenic genetic variant was identified in 6 (30%) patients: 3 in DSP, 1 in DSG2, 1 in PKP2, and 1 in SCN5A. Sixteen (69%) were taking at least one medication, and only 1 was not taking beta blockers. Sixteen (69%) had at least one episode of arrhythmia (5 atrial tachycardia, 8 NSVT, 1 sVT, 2 VF). 4 underwent successful ablation for atrial tachycardia. 7 patients underwent ICD implantation (primary prevention n=5, secondary prevention n=2). One patient had received a cardiac transplantation, and 2 further patients remain on the cardiac transplant waiting list.
Conclusions: Our findings are consistent with the recent ESC guidelines, demonstrating that cardiomyopathies are a continuum from childhood to adulthood. These data show that disease presenting in childhood can be severe and associated with life-threatening arrhythmias or end-stage heart failure and supports the need for early screening of paediatric first-degree relatives.

Keywords: cardiomyopathies, arrhythmogenic ventricle cardiomyopathy, dilated cardiomyopathy, non dilated left ventricular cardiomyopathy

PP-716

Genetic variant screening and association study of *nkx2-5* in congenital heart disease patients from north india

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Background and Aim: Globally 1% of the live births are affected by some form of congenital heart anomaly. Genetics and environment both play a role in its causation but very little of these aspects are explored from the Indian subcontinent. One of the first and key transcription factors required for the formation of the heart during development is *NKX2-5*. Several mutations in this gene have been identified for Congenital Heart Diseases (CHDs). In this study, we screened for known and novel variants to understand their role in CHDs.
Method: Two exons and flanking 3' and 5' UTR regions of *NKX2-5* were Sanger sequenced using 4 primer pairs in n= 71 CHD cases from north India after Institution Ethics Committee (IEC) approval. Case-control test of association and haplotype study was performed with the publicly available South Asian (SAS) population database.
Results: Only 3 known variants namely rs2277923 (c.63A> G) in exon 1, rs3729753 (c.606G>C) in exon 2, and rs703752 (c.61G>T) in 3'UTR were identified in a total of n= 69 cases. Case-control test of association for the 3 variants revealed no significant allelic or haplotypic association. A genotypic association was observed for rs703752 in a recessive model (p=0.03), along with a trend of association for rs3729753 (p=0.064) and rs703752 (p=0.082).
Conclusions: Although we did not identify any new mutations in the coding regions of NKX2-5 gene, our findings are important observations and incite for establishing the association between *NKX2-5* variants and cardiac defects in the context of the north Indian population. There is a need to explore the role of other transcription factors and cardiac developmental pathways to establish their interaction and role in disease biology in the Indian Subcontinent.

Keywords: Congenital Heart Disease, NKX2-5, Sanger sequencing, north India, Genetic variant, Case-control association

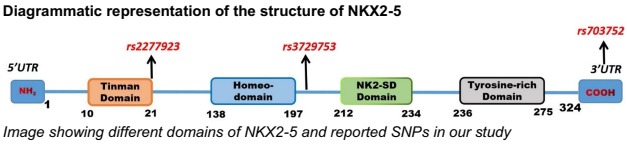

Image showing different domains of NKX2-5 and reported SNPs in our study

PP-717

The c.1617del variant of tmem260 is identified as the most frequent single gene determinant for persistent truncus arteriosus in japanese population

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Background and Aim: Recent advances in genetic analysis have facilitated the exploration of causative genes for congenital heart disease. We aim to explore the clinical impact of TMEM260 variants, which have been reported to be associated with a cardiac outflow tract (OFT) defect, persistent truncus arteriosus (PTA).
Method: We performed whole-exome sequencing on two Japanese patients with PTA in a family to detect a possible pathogenic variant of TMEM260, and searched for this variant in 26 individuals from the genome bank of Japanese patients with PTA. We also characterized the detected variant by western blotting and immunocytochemistry. The developmental expression pattern and transcriptional regulation of Tmem260 were analyzed by in situ hybridization with embryos and reporter assays using a putative enhancer region of the Tmem260 genome, respectively.
Results: We identified the c.1617del variant of TMEM260 in both patients of our familial case and in five (19%) of 26 individuals from the genome bank. Intriguingly, the public database indicated that the c.1617del of TMEM260 is a rare variant found only in Japanese and Korean populations. The phenotype of patients with c.1617del appeared predominantly in the heart although TMEM260 was reported as the responsible gene for the SHDRA syndrome (OMIM# 617478) associated with structural heart defects, renal anomalies, and nerve symptoms. Further, our functional assays using proteins translated from the mouse Tmem260 variant synonymous with the human c.1617del variant exhibited truncation, downregulation, and abnormal aggregation. The expression of TMEM260 was documented in the developing OFT, and it was shown to be regulated by an approximately 0.8 kb

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genomic region in intron 3 that contains highly conserved binding sites for essential cardiac transcription factors.
Conclusions: The c.1617del variant of TMEM260 is a major cause of PTA in the Japanese population with its frequency next to that of the 22q11.2 deletion, the most well-known genetic cause of PTA. The pattern and regulation for expression of TMEM260 are compatible with PTA. Genetic testing for TMEM260 should be considered in patients with PTA lacking the 22q11.2 deletion not only in Japan but also in Europe to elucidate its impact for the European population with PTA.

Keywords: Cardiac outflow tract defect, heart development, exome sequencing, truncation, enhancer

PP-718

Vascular structure and stiffness in pediatric mulibrey nanism using ultra-high frequency ultrasound

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Background and Aim: Mulibrey nanism (MUL) is a recessive disorder particularly prevalent in the Finnish population presenting with severe growth delay and pericarditis-related congestive heart failure determining prognosis. We aimed to delineate arterial and venous morphology, and arterial stiffness in a representative pediatric MUL cohort.
Method: 23 MUL and 23 individually sex and age matched healthy controls were prospectively assessed in a cross-sectional study with ultra-high frequency ultrasound (48-70 MHz).
Results: Heart failure was present in 7 MUL patients, with severe congestive heart failure in 2. Pericardiectomy had been performed due to non-inflammatory pericarditis in 6 MUL. Arterial lumen diameters and arterial wall layer thickness (intima-media thickness and adventitia thickness) were smaller in MUL patients, but appropriate for body size when compared with controls. Systolic and diastolic blood pressure, aortic and carotid compliance, stiffness as well as central aortic pulsed wave velocity were all similar in MUL compared with controls. Plasma pro-BNP levels were variably elevated (>300 ng/l) in 9/23 MUL patients and in 4/18 MUL patients older than 5 years of age. Internal jugular vein (mean difference 0.054 mm, CI95% 0.024-0.084) and cubital vein (0.046 mm, CI95% 0.013 - 0.078) total wall thickness was elevated in MUL compared with controls. Myocardial motion abnormalities and persistent congestive heart failure were common (3 out of 6) in MUL with history of pericardiectomy consistent with myocardial restriction. There were no statistically significant relations between vascular parameters and clinical or laboratory signs of heart failure or pericardiectomy.
Conclusions: MUL disease presents with significant constriction related diastolic dysfunction. Arterial lumen, wall layer thickness and stiffness are appropriate for body size in MUL, and like healthy controls. Mild venous wall thickening in the upper body region may be due to increased venous pressures related remodelling caused by diastolic heart failure. Myocardial abnormalities were prevalent among MUL with pericardiectomy performed and consistent with progression of myocardial disease in a significant proportion of patients.

Keywords: Mulibrey nanism, Trim 37, pericardial disease, arteries, stiffness, veins

PP-719

Identification and validation of gene module along with immune cells infiltration patterns in calcified aortic stenosis via transcriptome analysis

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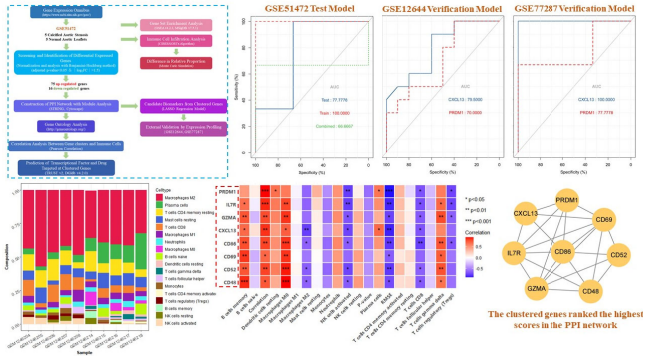
Background and Aim: Calcific aortic valve disease (CAVD) is currently the most prevalent valvular disease. However, the pathological mechanism of CAVD has not yet been fully elucidated. This study aimed to explore the differentially expressed genes (DEGs) along with infiltrating immune cells landscape and their potential mechanisms in the progression of calcific aortic valve disease.
Method: In this study, three CAVD-related microarray datasets were downloaded from the NCBI-GEO database. The gene set enrichment analysis was performed for interpreting the biological insights of gene expression data. The CIBERSORTx algorithm was implied to infer the relative proportions of infiltrating immune cells of the CAVD samples via R software. The protein interaction network was constructed via STRING and visualized by the Cytoscape. Gene ontology (GO) enrichment was performed via geneontology.org. The least absolute shrinkage and selection operator (LASSO) logistic regression and receiver operating characteristics (ROC) analyses were performed to build machine learning models for evaluating the capability of clustered genes in predicting CAVD. The expression levels of the cluster genes were validated in two external cohorts.
Results: Pathways related to immunoinflammatory responses were identified according to GSEA analysis, and the detailed fractions of infiltrating immune cells were compared between the normal and calcified aortic valves. Additionally, we identified 91 DEGs in CAVD progression ($|\log2FC| \geq 1.5$ and adjusted $p < 0.05$). They were mainly enriched in GO terms relating to inflammatory response and immune response. A cluster of eight genes, namely IL7R, GZMA, CD48, CD86, CXCL13, PRDM1, CD69 and CD52, were found to be significant, and their correlations with infiltrating immune cells were calculated. The cluster genes were also validated to be differentiated expressed in two external cohorts. Moreover, PRDM1 and CXCL13 may exert predictive pathogenic functions in the process of CAVD, as the machine learning prediction model showed a preferable result.
Conclusions: We reanalyzed the transcriptomic signature of CAVD development with the landscape of the immune cell, as well as revealed new insights and specific prospective DEGs for the investigation of disease-associated dynamic molecular processes and their regulations with immune cells, holding potential insights for the investigation of curative therapies.

Keywords: Calcified aortic stenosis, Immune cells infiltration, Machine learning, Microarray, CXCL13, PRDM1

Figure 1 Central illustration



The schematic information and the pathophysiological relations in training datasets and validation datasets.

Interventional Cardiology

PP-720

Atrial septal defect (ASD). How large is too large for thranscatheter closure? Our experience

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Background and Aim: Transcatheter device-based closure is considered the first line therapy for secundum atrial septal defect (ASD). There are still uncertainties regarding the feasibility of performing percutaneous closure in small children with large defects. Some experts consider a device size/patient weight ratio greater than 1.5 as the cut-off value to recommend surgical repair. This study evaluate the safety, efficacy, and short term follow-up results of patients who underwent trans-catheter closure of secundum ASD in small children.

Method: Single-center, retrospective cohort study of children weighing less than 15 Kg undergoing percutaneous closure of ASD. Patients' demographic and clinical data were collected. Indications for closure were patients with right ventricular enlargement and symptoms. Associated co-morbidities were prematurity in 2 patients (8%), genetic syndromes in 3 patients (12.5%) and 3 patients (16%) had other associated cardiac defects. The procedure was carried out under general anesthesia with fluoroscopy and transesophageal control (TEE). A device 0mm to 2mm larger than the ASD measurement assessed by TEE was chosen. The Amplatzer® Atrial Septal Occluder (St. Jude Medical Inc.–St. Paul, United States) was the device used.

Results: Twenty-four patients were treated between January 2022 and November 2023. Nine patients were male. The average age and weight were 27 months (3-50 months) and 10 kg (3.8-15 kg), respectively. The mean defect size was 11mm (8mm-15mm), the mean device size was 12 mm (9-16 mm), and the device-to-weight ratio was 1.2 (0.8 - 2.4). 33% of patients (n=8) had a device-to-weight ratio >1.5. One patient had transient first-degree atrioventricular block, and another had mild mitral regurgitation. In both patients the device/weight ratio was greater than 1.5. We obtained a median follow-up of 2 months (1-10months). Technical success was achieved in all 24 patients (100%). There was no mortality.

Conclusions: Percutaneous ASD closure in selected symptomatic infants weighing <15kg with hemodynamic consequences and significant clinical impact is a feasible, safe and effective alternative and should be the first option therapy. No major complications were observed during the procedure. Very early closure of ASD should be reserved for those associated with symptoms, prematurity with chronic lung disease, PAH and syndromes. Long-term follow is needed.

Keywords: Atrial septal defect, thranscatheter closure.

PP-722

Patent foramen ovale closure with noblestich system after a failed device closure attempt in a patient with ebstein anomaly and arterial desaturation

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Background and Aim: Arterial desaturation due to a patent foramen ovale (PFO) with right-to-left shunt is common in patients with Ebstein anomaly. However, the complex anatomy of PFO may sometimes to compromise the procedural success. We describe a complex case of a patient with unknown Ebstein anomaly and arterial desaturation due to PFO right-to-left shunt in which a severe hypertrophy of the septum secundum didn't allow an effective device closure.

Method: A 78 years-old female patient came to the attention of our Adult Congenital Heart Disease Department because of significant arterial desaturation at rest (SaO2 75%). The patient required an immediate ventilatory support with high-flow nasal cannula (FiO2 100%). Chest X-ray and HR-CT scan excluded lung disease. However, the echocardiography showed an Ebstein anomaly (tricuspid septal leaflet apical dislocation of 1.2 cm/m2) with moderate tricuspid regurgitation, a huge right atrium and an aneurismatic interatrial septum with a right-to-left PFO shunt.

Results: Cardiac catheterization was done. A balloon occlusion test of PFO was performed. After 10-15 minutes of balloon PFO occlusion, there was a significant improvement of arterial oxygen saturation (from 82-83% to 95%) without both a significant right atrial pressure rise (from 10 to 12 mmHg) and systemic cardiac output fall. An occluder device was fenestrated to allow a mild right atrium "pOP-off"; however, the hypertrophic septum secundum (15.5 mm) prevented a good device deployment. In this scenario, we decided to try the closure with the NobleStich EL system. After S- and P- suture deployment, the KwiKnot was released. The trans-esophageal echocardiography showed an optimal result with just a trivial PFO right-to-left shunt. Already in the catheterization laboratory, the arterial oxygen saturation raised up to 93-94%. Tw day after the procedure, the patient was discharged with good arterial saturation (92-93%) with neither ventilatory nor oxygen support.

Conclusions: Ebstein anomaly may be often the cause of a significant desaturation due to a PFO right-to-left shunt. In some cases, PFO device closure may fail because of the presence of a complex anatomy. In these cases, NobleStich EL system is an effective approach to allow PFO closure with complete shunt elimination (or at least shunt reduction).

Keywords: Ebstein anomaly, Patent Foramen Ovale, Right-to-Left Shunt, Arterial Desaturation, Noblestich EL

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PP-723

Neonatal predictors of long-term follow-up outcomes in pulmonary atresia with intact ventricular septum and critical pulmonary stenosis

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Background and Aim: Pulmonary Atresia with Intact Ventricular Septum (PA-IVS) and Critical Pulmonary Stenosis (CPS) are cyanotic duct-dependent congenital heart diseases.The aim of our study is to evaluate the impact of neonatal predictive factors after right ventricle decompressionon the prognosis of these patients.
Method: From 2011 to 2022, 55 newbors with PA-IVS (30 pts) and CPS (25 pts) were admitted to our Pediatric Cardiology Department. Three patients didn't undergo pulmonary valve perforation due to either a monopartite right ventricle or a significant right ventricle – coronary connection. The others fifty-two patients (94.5% of all) underwent right ventricle decompression and they were our cohort.
Results: Twenty-seven pts (51.9%) (group I) required an arterial duct stenting in neonatal age, whereas the other 25 pts (48.1%) (group II) were discharged after right ventricle decompression. Group I showed smaller tricuspid valve diameter (9.1±1.8 vs 10.8±2 mm, p<0.01; z-score -1.95±1.07 vs -0.9±0.98, p<0.01), tricuspid/mitral valve annular ratio (0.75±0.13 vs 0.96±0.15, p<0.01) and pulmonary valvular annulus (diameter 6.1±0.7 vs 7.1±1 mm, p<0.01; and z-score -1.62±0.81 vs -0.51±0.87, p<0.01) right ventricle end-diastolic area (1.2±0.6 vs 2±0.7 cm2, p<0.01), larger right atrium end-systolic area (4±1.44 vs 2.5±1.05 cm2, p<0.01), and a worse right ventricle diastolic function, as inferred by higher E/E' ratio (9.7±4.2 vs 6.9±3.2, p<0.01) and percentage amount of atrial right-to-left shunt (75±15.7 vs 56.4±27.8%, p<0.01). At a mean follow-up of 6.3±3.1, group I had higher risk of both surgical RVOT reconstruction (6 pts, 22.2% vs 0) and percutaneous re-balloon valvuloplasty (9 pts, 33.3% vs 2 pts, 8% p- 0.03) and being therapy-dependent (furosemide and/or propranololo) (7 pts 25.9% vs 1 pt, 4%; p- 0.03). In group I, no neonatal echocardiographic parameter was predictive of surgery during follow-up.
Conclusions: Trans-catheter approach is effective to relief PA-IVS and CPS. Multiple echocardiographic parameters allow to predict the need of an arterial duct stenting during follow-up. PA-IVS and CPS patients undergone arterial duct stenting have a higher risk of percutaneous or surgical procedures and/or to be therapy-dependent during follow-up. No patient discharged without arterial duct stenting required a surgical procedure during follow-up. Among patients undergone arterial duct stenting, no neonatal echocardiographic parameter was predictive of surgery.

Keywords: pulmonary atresia, pulmonary stenosis, trans-catheter, arterial duct stenting

PP-724

The impact of percutaneous stenting on the arterial blood pressure control in native aortic coarctation and recoarctation

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Background and Aim: Native aortic coarctation and aortic recoarctation are usually associated to a significant arterial blood hypertension. Nowadays, percutaneous stenting is the first-line treatment. However, the patients show different improvements to trans-catheter dilatation. The aim of our study is to assess the impact of percutaneous stenting on the arterial blood pressure (ABP) control and to evaluate the predictive factors of "good response".
Method: This is an observational retrospective single-center study. We evaluate all patients with native aortic coarctation or aortic recoartation undergone percutaneous stenting in our Department (Pediatric Cardiology Department and Adult Congenital Heart Disease Department, "AORN dei Colli", Monaldi Hospital) from January 2006 to November 2021. Primary outcomes were: lower ABP values (at least 20 mmHg of systolic arterial pressure reduction), reduced dosage of the anti-hypertensive (AH) therapy, reduced number of AH drugs used. They were assessed both singularly as well as like composite outcome. They were evaluated one year after percutaneous stenting.
Results: Eighty-two patients underwent percutaneous stenting due to aortic coarctation and recoarctation. At one year follow-up, a significant improvement of systolic arterial blood pressure was detected (141,6±22,8 vs 122,4 mmHg, p- 0,05). Fifty-five patients (67%) showed a significant (>20 mmHg) reduction of right arm ABP (45 pts, 54,8%) and/or reduced dosage of the AH therapy (5 pts, 6,1%) and/or reduced number of AH drugs used (26 pts, 31,7%). Native aortic coarctation group showed a more significant ABP reduction (25,5±10,6 mmHg vs 13,8±4,9 mmHg, p- 0,02) and composite outcome (80% vs 54,7%, p- 0,02) than aortic recoarctation group. Multivariate analyses demonstrated a significant impact of a native aortic coarctation (p- 0.02, 95%CI 1,2-8,8) and no significant statistically impact of the age (p- 0,4) and the bicuspid aortic valve (p- 0,57) on the composite outcome.
Conclusions: Percutaneous stenting is effective to deal both native aortic coarctation and aortic recoarctation, and it is associated with a significant improvement of ABP control, above all in patients with native aortic coarctation.

Keywords: Aortic Coarctation, Percutaneous Stenting, Arterial Blood Pressure, Trans-catheter Angioplasty

PP-725

Different treatment methods for utilizing pericardium as a collagen-based biomaterial for clinical applications

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Background and Aim: Bioprostheses undergo a pre-treatment process involving cross-linking. The most widely used cross-linking method is chemical treatment via glutaraldehyde. Glutaraldehyde treatment serves several essential purposes. However, this treatment comes with some disadvantages, leading to the implantation of a non-viable prosthesis that is prone to calcification. The current study aims to investigate alternative treatment methods to glutaraldehyde for the production of heart valve prostheses using autologous pericardial tissue, to address these limitations and improve the long-term performance and durability of these prostheses.
Method: We assessed a range of cross-linking methods, encompassing chemical, physical, and biological approaches. We evaluated their efficacy in terms of cross-linking capability through techniques such as collagenase assays. Additionally, we explored their biocompatibility using various assays. Furthermore, we analyzed the impact of these cross-linking methods on tissue characteristics, including mechanical properties and fiber alignment. In addition, we investigated the remodeling behavior of the treated tissue after conditioning. Therefore, we developed a custom-designed pericardial stretching device tailored to our specific needs.
Results: The various treatment methods applied to the tissue impart distinct properties. For example, chemical fixatives yielded the highest degree of cross-linking. However, it is noteworthy that prostheses subjected to alternative cross-linking methods also exhibited mechanical properties that fell within the normative range.
Conclusions: Currently, chemical fixatives remain the preferred choice for cross-linking biomaterials due to their efficacy, as alternative methods frequently result in a diminished degree of cross-linking. Nevertheless, this reliance on chemical fixatives comes at a cost, notably in terms of the long-term durability of treated prostheses. In contrast, autologous tissue necessitates cross-linking primarily to increase its mechanical strength and mitigate enzymatic degradation, demanding a lesser degree of cross-linking in comparison to xenogeneic- or allogeneic- tissue. By the combination of a non-chemical cross-linking approach in conjunction with autologous tissue, a durable prosthetic implant could emerge. This paper reviews various options for tissue treatment and preservation and highlights the advantages and disadvantages in the context of somatic growth in our patients with congenital heart valve defects.

Keywords: Cross-linking, Implantation, Viable Prosthesis, Remodeling

PP-726

Lymphatic interventions – a case series of 9 patients

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Background and Aim: Complications related to the lymphatic system such as protein losing enteropathy (PLE), plastic bronchitis, (PB), ascites, pleural effusions, and edema formation can now be diagnosed and new interventional treatment modalities are available. We started lymphatic interventions for lymphatic complications in Copenhagen, Denmark in 2021 and here we report up to 2 years outcome for the first 9 patients.
Method: Nine patients (age range: 10-30 years, 6M, 3F)(7 Fontan, 2 without CHD) with intractable lymphatic complications (PLE, n=6; PB, n=1, pleural effusion, n=1, abdominal pain thought to be related to lymphatic compartment syndrome, n=1) were referred. Diagnostic algorithm included non-contrast magnetic resonance lymphangiography followed by dynamic contrast magnetic resonance lymphangiography (DCMRL) with contrast injections intranodally in the groins, in the liver and in the mesentery. Patency of the thoracic duct inflow into the innominate vein was confirmed with ultrasound. Anomalous lymphatic vessels connecting to the duodenum (N=5), bronchial system (N=1), pleural space (N=1) and in the hepatoduodenal ligament (N=2) were identified. Anomalous lymphatic vessels were occluded using selective intralymphatic (n=7) or direct (N=1) gluing. One patient had no treatable anomalous lymphatic vessels. Five of the Fontan patients had an additional procedure within the following week to decompress the central venous pressure. The first patient had a surgical innominate vein turn down. The following 4 patients had a catheter-based decompression by implanting a covered stent connecting the innominate vein (where the thoracic duct enters) with the atrium.
Results: All treated patients had resolution or improvement of symptoms including diarrhea, effusions, fatique, and abdominal pain. The patients report to be able to resume normal daily activities such as playing with peers, engage in mild to moderate exercise, attend school/education, and participate more hours at work. Albumin, immunoglobulins, and electrolyte levels improved or normalized. Medication was reduced.
Conclusions: With a short (up to 2 years) follow up time, our experience with interventions for lymphatic complications is promising. This group of very sick patients have gained a more normal life and the lymphatic interventions including embolisation and decompression seem to have pushed the boundaries for their activity level and well being positively.

Keywords: Lymphatic interventions, Fontan circulation, Dynamic Contrast Magnetic Resonance Lymphangiography

PP-727

Clinical usefulness of three-dimensional guidance for cardiac catheterization in congenital heart diseases

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Background and Aim: The integration of imaging overlay with 3D datasets during fluoroscopy-guided catheterizations was introduced a considerable time ago. However, there is still limited evidence regarding its quality and clinical utility. This study aims to evaluate the accuracy and clinical advantages of 3D imaging overlay during diagnostic and interventional catheterizations in congenital cardiology.
Method: A retrospective analysis was conducted on all adequately documented catheterizations in patients with congenital heart diseases at our Center, during which 3D imaging overlay was

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employed (since 2020). Procedures with 3D rotational angiography overlay were excluded. All procedures utilized the Vessel Navigator software (Philips). They were retrospectively assessed by all operators (n=4) in terms of overlay accuracy and clinical benefit, categorized into four classes: 0 – inaccurate/of no benefit, 1 – minor, 2 – moderate, 3 – accurate/of significant importance.
Results: Among over 100 procedures using 3D dataset overlay, 56 procedures in 51 patients were included. All 3D data were derived from angio-CT. The median patient age was 9 years (range: 0.4-44, 11 adults). There were 16 diagnostic (34.8%) and 40 interventional (65.2%) procedures, with pulmonary artery angioplasty and/or stenting being the majority in the latter group (n=19). All interventional procedures were successful, with one major and four minor complications unrelated to the roadmap use. The median fluoroscopy time was 16 minutes (2.8-120). Accuracy was mostly graded as 3 (53.6%), followed by 2 (16.1%), 1 (14.3%), and 0 (1.8%). Clinical advantage was graded as 3 (46.4%), followed by 1 (32.1%), 2 (19.6%), and 0 (1.8%). The benefit of 3D mapping during diagnostic catheterizations was assessed as insignificantly higher than in interventional procedures. Roadmaps to evaluate MAPCAs, shunts, and translocated coronary arteries were considered the most useful in the diagnostic group. Among the interventional group, overlay in hypoplastic pulmonary arteries angioplasty/stenting, RVOT stenting, TCPC tunnel puncture, and MAPCA closure (including one lifesaving procedure without contrast use) were assessed as the most helpful.
Conclusions: The accuracy of 3D overlay is satisfying in the majority of patients. This tool is safe and provides noticeable benefits in both diagnostic and interventional procedures, with some cases where it has become crucial for their success.

Keywords: cardiac catheterization, congenital heart disease, three-dimensional guidance

PP-728
Early experience with cardiac catheterization planning using virtual reality – case series
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Background and Aim: Virtual reality (VR) has recently emerged as an additional modality for analyzing three-dimensional (3D) datasets originating from CT and MRI studies. We aim to present our early experience with cardiac catheterization planning using VR in patients with congenital heart defects, focusing on the most interesting cases.
Method: VR was employed to analyze 101 CT-derived studies from 98 patients (20 adults) since June 2022 in a single tertiary center. Forty-four studies were created for planning subsequent catheterizations, including diagnostic in 10 and interventional in 34 patients. In the interventional group, procedures mainly involved coarctation of the aorta (CoA) stenting (n=8), pulmonary artery (PA) stenting (n=5), and closure of collateral vessels (n=4). All procedures were successful, with two minor complications unrelated to VR. We subjectively highlight four major cases where we believe VR planning was crucial for procedure success.
Results: A. A 7-year-old patient with a connective tissue defect underwent life-saving hybrid Valeo stent implantation during neonatal CoA with left subclavian artery transection. VR planning was crucial for selecting and positioning a covered 12 mm Bentley stent due to carotid artery proximity. The outcome was optimal. B. A 15-year-old patient, after PDA closure with a muscular VSD occluder due to high pulmonary pressure at the age of two, benefited from VR understanding of a unique device-related left PA stenosis. VR facilitated the selection of the stent size, and the artery was successfully dilated. C. A 19-year-old Fontan patient with chronic drug-resistant atrial tachycardia underwent VR reconstruction, revealing a small remaining fenestration for ablation and atrial anatomy recognition. Urgent temporary and later permanent electrode implantation through the fenestration into the single ventricle was required. D. A 34-year-old patient with congenital aortic valve stenosis, after Ross surgery at 14, Bentall surgery with conduit implantation in the pulmonary position at 33, and eventually severe symptomatic supravalvular stenosis, underwent successful VR reconstruction for anatomy understanding and precise stent position planning, with preservation of the pulmonary valve.
Conclusions: VR is an additional tool that enhances understanding of anatomy, aids in procedure planning, and contributes to its success. VR application is particularly useful in patients with complex morphology.

Keywords: cardiac catheterization, congenital heart disease, virtual reality, medical simulation

PP-729
Retrospective series on neonatal aortic thrombosis: Clinical persentation, therapeutic strategies and outcomes
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Background and Aim: Neonatal arterial thrombosis is a rare but extremely severe condition with high mortality rate. Clinical presentation varies according to the location of the thrombus and is usually dramatic. There are no treatment guidelines. We report a series on hyper acute presentation of aortic or arterial thrombosis in neonates with focus on treatment and outcome.
Method: All neonates with history of proven aortic or arterial thrombus without iatrogenic etiology were retrospectively reviewed in a single center series from January 2012 until september 2023. Clinical presentation was reviewed as well as diagnostic tools used. Therapeutic treatment and outcomes as well as potential etiologies were analyzed.
Results: 12 neonates were identified (4/12 female). Mean weight was 3150 gr. 8 were diagnosed between day 0 and day 1 and 3 were diagnosed between day 6 and 8. Mode of presentation were 3 antenatal ventricular assymetry, 4 cardiogenic shock, 3 limb ischemia, 1 seizure and 1 systemic hypertension. Thrombus location was thoracic aorta in 2, abdominal aorta in 4, coronaries in 2, left atrial appendage in 1 and limb arteries in 3 (sub-clavian, axillary and femoral). 2 patients had surgical removal of the thrombus in thoracic aorta, 4 had percutaneous treatment including thromboaspiration, thrombolysis and stent implantation and 6 had heparin infusion. There were no general thrombolysis. 2 patients with

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coronary thrombosis ultimately died, 1 patient had limb amputation and the other patients had complete recovery. One patient had Factor V Leiden Mutation.
Conclusions: Aortic thrombosis in neonates has various clinical presentation and should be ruled out in unusual clinical pictures. All therapeutic strategies should be discussed in multidisciplinary team ranging from heparin infusion to surgical resection Endovascular therapy is a valid option in a substantial number of patients.

Keywords: neonatal - arterial thrombosis - percutaneous treatement

PP-733
Evaluation of the effect of transcatheter asd closure on flow parameters

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Background and Aim: Pre- and post-procedural flow parameters of patients who underwent transcatheter closure of atrial septal defect (ASD) in our clinic in 2023 were analyzed and the reflection of these values on remodeling was investigated.
Method: Amplatzer Septal Occluder, Lifetech CeraFlex Septal Occluder, and Occlutech Figulla Flex II Atrial Septal Occluder devices were used for closure. Inferior vena cava (IVC) maximal and minimal diameter, IVC index, main pulmonary artery (PA) pulse wave (PW) Doppler gradient/ velocity/VTI(velocity time integral) in the parasternal short axis, and PW Doppler gradient/ velocity/ VTI values in the right upper pulmonary vein (PV) in the apical four-space window were determined as variables.
Results: In our study, female: male ratio was 1:1.45, mean age was 81.4 months(11-186), mean procedure weight was 23.89 kg(4.1-58), defect size by TTE was 11.7 mm(7-19), defect size by balloon sizing was 14.1 mm(10-25), ASD device size was 11.7 mm(12-19). When the data obtained were evaluated, it was observed that PA and PV gradient, velocity, VTI, and IVC maximum, and minimum parameters decreased statistically significantly after transcatheter closure compared to before ($p<0.01$). Although a decrease was observed in PV mean gradient and IVC index, it was not found to be statistically significant ($p>0.05$).
Conclusions: Previous studies have shown that ASD closure by transcatheter method significantly decreases flow parameters in the early period and contributes to the development of remodeling. When our study is evaluated in the light of the literature, PA and PV,VTI values stand out as a good markers reflecting the change in flow parameters after transcatheter ASD closure. The evaluation of IVC parameters in transcatheter ASD closure has not been previously reported in the literature. The significant change in the IVC parameters suggests that it may be considered an indirect indicator of the development of remodeling in the right structures. All flows in the PA,PV,and IVC returned to normal in the early period after percutaneous closure. This contributes to the decrease in both atrial volumes and the development of remodeling. This is the first study in the literature in which flow parameters of the PA, PV, and IVC were evaluated in combination in transcatheter ASD closure.

Table 1

Variables	Before (Mean +SD)	Day 1 (Mean +SD)	P Value
Pulmonary artery max gradient	9,4 ± 5,3	6,9 ± 4,6	< 0,001
Pulmonary artery mean gradient	5,1 ± 3,1	3,6 ± 2,1	<0,001
Pulmonary artery max velocity	152,7 ± 35,0	126,0 ± 35,1	<0,001
Pulmonary artery mean velocity	98,3 ± 27,5	78,5 ± 22,7	<0,001
Pulmonary artery VTI	28,4 ± 6,9	23,8 ± 6,4	<0,001
Pulmonary vein max gradient	2,5 ± 0,84	1,9 ± 1,1	0,007
Pulmonary vein mean gradient	1,3 ± 0,68	1,1 ± 0,45	0,11
Pulmonary vein max velocity	75,2 ± 14,0	61,8 ± 18,3	0,001
Pulmonary vein mean velocity	51,3 ± 10,1	41,3 ± 9,6	<0,001
Pulmonary vein VTI	25,0 ± 8,7	19,9 ± 7,1	<0,001
IVC min	7,1 ± 1,8	6,1 ± 2,2	0,004
IVC max	9,6 ± 2,7	8,2 ± 3,0	<0,001
IVC Index	0,25 ± 0,069	0,24 ± 0,063	0,3

Comparison of Pulmonary Artery, Pulmonary Vein, and IVC Flows Before and After the Procedure

Keywords: atrial septal defect, flow parameter, pediatric, transcatheter closure

PP-734
Virtual reality simulations in planning ductus arteriosus stenting in patients with duct-dependent pulmonary circulation

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Background and Aim: Ductus arteriosus (DA) stenting has become a standard for early palliative treatment of neonates with duct-dependent pulmonary circulation. Nevertheless, the procedure is complicated especially in long and tortuous ducts, which often require additional imaging and thorough planning for optimal stent length selection. Our aim was to simulate the implantation of a stent using virtual reality direct volume rendering software and to retrospectively compare the results with intraprocedural angiography.
Method: 17 patients with duct-dependent pulmonary circulation who had undergone DA stenting with CT scan performed prior to the procedure were chosen. Tomographic data was uploaded directly to the software. We assessed DA morphology and the optimal vascular access site for the procedure. Using tools provided by the software a stent covering the DA was simulated. Afterwards, the simulation was compared with descriptive data and angiography from the procedure, including stent length, position and access site.
Results: Out of 17 simulations, in 9 cases implanted stent length was identical with simulation. The simulated stent was shorter than the implanted one in 6 cases and longer in 2 cases. The access site was successfully predicted in 16 cases, albeit in one of them the simulated access was a second choice and in 2 the simulated access was aborted during the procedure. In 1 case another arterial vessel was chosen. The position of the stent was identical in 11 cases. Overall, 5 simulations presented a complete concordance of all three factors, another 4 differed in the positioning of the stent, but not its length or access site.
Conclusions: DA stenting is a procedure burdened with many variables, thus making it relatively hard to simulate. Virtual reality volume rendering allowed for acceptable prediction of stent length without intraprocedural measurements, yet an important variable is operator's selection of oversized stents for safety reasons. The simulations allowed for accurate prediction of the vascular access site based on the morphology of the aortic ostium of DA. The predictions of stent position were fairly accurate with differences depending on intraprocedural events. Overall, virtual simulations

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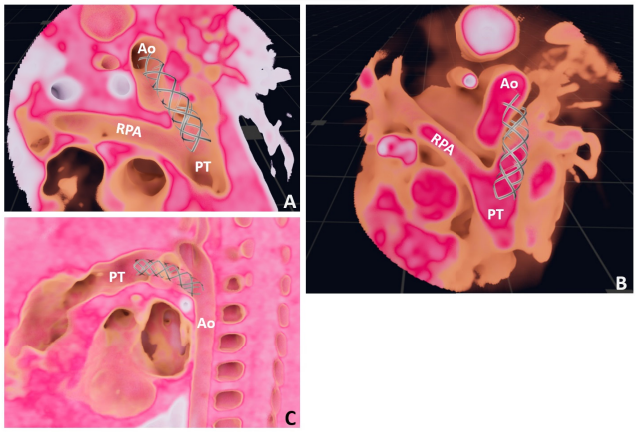
of stent implantation might prove valuable in planning real procedures, especially in more complex and controversial cases.

Keywords: virtual reality, congenital heart disease, virtual simulations, interventional cardiology

Figure 1. An exemplary successful simulation of a stent using different transfer functions



A and B present a cranial projection, C presents right lateral projection. Ao – aorta, PT – pulmonary trunk, RPA – right pulmonary artery

PP-735

Safety and efficacy of the osypka vacs®ii and vacs®iii balloon catheters in patients with congenital heart defects – a retrospective monocentric analy

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Background and Aim: Balloon dilatation (i.e. valvuloplasty and/or angioplasty) is one of the key measures to manage various congenital or residual defects in the field of congenital cardiology. Despite the long term use over decades, the results of specific types of balloons is reported only insufficiently. The choice of the balloons used hereby depends on various aspects and may include the size of the target vessel (balloon diameter size), structure or valve (balloon size and length), the potential size of the vascular access deemed favorable (requested introducer size), the estimated rigidity of the target lesion (balloon inflation pressure), the anatomy and pathology of the target lesion (i.e. calcification) or the management process (i.e. balloon sizing) during interventions)
Method: we retrospectively analyzed our database for procedures where VACS®II and VACS®III balloons were used; we choose 25 subsequent patients in each diagnostic group and analyzed balloon deployment and removal, inflation characteristics and hemodynamic results.
Results: Successful balloon inflation and removal was possible in 200 patients from newborn age to adulthood; in valvular and aortic stenosis, significant gradient reduction was documented. In peripheral pulmonic stenosis (PPS), right ventricular outflow tract (RVOT) obstruction and coarctation of the aorta (CoA), technical balloon dilatation was excellent, hemodynamic gradient reduction however varied according to the underlying nature of the stenosis (i.e. native, recurrent, after stent implantation) or intended or the intended balloon use (i.e. balloon interrogation). Stent placement was feasible using VACS®III balloons for various conditions (i.e. PPS, CoA, RVOT). 25 Additional miscellaneous procedures were documented including atrial septal dilatation, venous balloon interrogation and dilatation of venous structures as well as biological bioprostheses.
Conclusions: In the field of congenital cardiology, the VACS®II and VACS®III balloons were used successfully and without complications for various indications; successful balloon performance must be defined by the intended use such as balloon interrogation of given anatomical structures or the nature of the stenoses (i.e. tissue recoil) and not only by the amount of gradient reduction in stenotic structures. These different indications must be taken into account when assessment by regulatory bodies especially in the context of the new European medical device regulation.

Keywords: balloon valvuloplasty, pulmonic and aortic stenosis, coarctation of the aorta, stent implantation, interventional cardiology, VACSII and VACSIII balloon

PP-736

Clinical case of retrograde perforation of the pulmonary valve and rvot stenting in newborn with tetralogy of fallot, pulmonary atresia and mapcas

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Background and Aim: The management of newborns with tetralogy of Fallot with pulmonary atresia, major aortopulmonary collateral arteries (ToF/PA/MAPCAs) and especially with hypoplastic native pulmonary artery branches can be so challenging. In these patients, pulmonary valve (PV) perforation and right ventricular outflow tract (RVOT) stenting are usually performe with an antegrade approach. However, there are limited publications of PV perforation through retrograde approach from the MAPCAs.
Method: The child was admitted to our center at the age of 15 days, by weight 2.68 kg. Patient had clinical signs of heart failure, required an inotropic support and the mechanical lung ventilation. By heart catheterization ToF/PA/MAPCAs with hypoplastic native pulmonary artery branches (the Nakata index – 51.1 mm2/m2 and the McGoon index – 0.83) were diagnosed. We decided to provide recruitment of hypoplastic native pulmonary artery tree.
Results: The procedure was performed through the right femoral vein and artery. The selective angiography of the RVOT showed that it was underdeveloped, anatomically located perpendicular to the PV. MAPCA's angiography was performed and the communication (5 mm) between it and the left pulmonary artery (LPA) was visualized. An angiographic catheter MP 4Fr was inserted into the PV through the communicating MAPCA. The PV perforation (Runthrough intermediate 0.014) and the balloon angioplasty of the PV (coronary balloons from 1,25 to 2.75 mm in diameter) were performed. After it the transcardial loop was formed. The Architec 4.5x16 mm stent was implanted antegradely in the position of the RVOT, crossing the pulmonary valve. Simultaneously, embolization of the two communicating MAPCAs to eliminate competitive flow between the native pulmonary artery and the MAPCAs was underwent. Between first procedure and total repair, patient needed two balloon stentoplasty. Total repair was performed at the age of 12 months (the Nakata index increased to 169,7 mm2/m2, the McGoon index – to 1,8).

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Conclusions: If an antegrade approach is impossible in newborns with tetralogy of Fallot, pulmonary atresia and major aortopulmonary collateral arteries, retrograde perforation of the PV followed by RVOT stenting may be safe and effective. This method allows to promote the growth of native pulmonary arteries and creates conditions for total surgical repair in the future.

Keywords: Retrograde pulmonary valve perforation, right ventricular outflow tract stenting, newborn,Tetralogy of Fallot with pulmonary atresia, major aortopulmonary collateral arteries

PP-737

One center experience of first stage palliative procedure in newborn with duct-depended pulmonaty blood flow. what is better?

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Background and Aim: The duct-dependent cyanotic newborns require a secure source of pulmonary blood flow. The treatment options are the creation of systemic-pulmonary anastomosis (BTSh) or the patent ductus arteriosus stenting (PDA stent). The aim of our single-center retrospective study was to compare the advantages and disadvantages of each method.
Method: Over the last 8 years (from 2015 to 2023), 82 BTSh and 47 PDA stent were performed in cyanotic newborns with critical duct-dependent circulation. In the BTSh group 78% (n=64) and in the PDA stent group 81% (n=38) had biventricular circulation. Prostaglandin E1 infusion was used in all patients. The average age were 6.4±3.9 days (BTSh) and 7.2±5.8 days (PDA stent) and the mean weight were 3.1±0.46 kg and 3±0.54 kg, respectively. In both groups the percentage of prematured babies was similar (26.8% (22) in the BTSh group, 25.5% (12) in the PDA stent group), while the genetic/concomitant pathology was higher in the PDA stent group (27.6% (13) vs 12.2% (10)). The mean SpO2 before procedure were 79.2±10.6% (BTSh) and 65.7 ±11.1% (PDA stent).
Results: After procedure, the average SpO2 were 81%±5.5% (BTSh) and 85%±4.6% (PDA stent) and the Nakata index increased from 147.4 ± 42.8 m2/m2 to 168 ± 35.9 mm2/m2 in BTSh group and from 147.2 ± 53.7 m2/m2 to 225 ± 87.4 mm2/m2 in PDA stent group. The length of stay in ICU were 14±7 days (BTSh) and 13.6±9.4 days (PDA stent) and the time of mechanical ventilation were 165.5±94 hours and 150±113 hours, respectively. The percentage of early complications was similar (19.5% (16/82) and 21.2% (10/47) respectively). The early 30-day mortality was 7.3% (6/82) in the BTSh group and 6.4% (3/47) in the PDA stent group. The late mortality (more than 30 days) was 19.5% (16/82) and 8.5% (4/47), respectively. The total mortality in the BTSh group were 26.8% (22 patients out of 82), in the PDA stent group - 14.9% (7 patients out of 47).
Conclusions: The both therapeutic approaches are equally effective palliative procedures for cyanotic newborns with ductal-dependent circulation. However, the early and late mortality were significantly higher in patients from BTSh group.

Keywords: newborn, congenital heart disease, pulmonary blood flow

PP-738

Transcatheter closure of perimembranous ventricular septal defect using the lifetech multi functional occluder – 154 patients

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Background and Aim: We aimed to evaluate the effectiveness and outcome of transcatheter pmVSD closure using the Lifetech-multi functional occluder (MFO).
Method: Clinical features and demographic characteristics and follow-up findings of the perimembranous VSD closure with MFO were evaluated retrospectively from our center
Results: Lifetech MFO was used in 154 patients, 88% female. The mean of age was 5.3±4.2 (0.4-25) years; weight 19±13.6 (5.8-77), defect size at the LV side 9±3.5 (3-20) mm, shunt ratio 1.7±0.4 (1.1-4), mean PAP 24.8±8.7 (9-51) mmHg, fluoroscopy time 13.7±8 (3.3-60) and aortic rim 2±2.1 (0-12) mm. In 58% of them, closure was performed via the retrograde way. Major complications occurred in 13 patients (8.4%) (embolization in nine, LBBB in two, moderate aortic insufficiency in two). There was no mortality. Residual shunt flow was found 12.3%, all of them minimal. Neither complete atrioventricular block, nor other complications occurred.
Conclusions: Transcatheter closure of perimembranous VSDs in selected patients using the LifeTech MFO device seems effective. Its advantages are softer design, use of both an antegrade and retrograde approach, and an advanced smaller delivery system. Increasing the number of usage and the experience will provide more accurate data and low complication rates.

Keywords: MFO, perimembranouz VSD, childhood

PP-739

Evaluation of patients with subclavian steal syndrome and pulmonary steal syndrome: A single-center experience

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Background and Aim: To describe clinical and imaging characteristics of an isolated subclavian artery in pediatric patients and to emphasize that atypical PDA of patients with subclavian and pulmonary steal syndrome can be closed and treated transcatheterly.
Method: We retrospectively evaluated the patients diagnosed with subclavian steal syndrome (SSS) and associated pulmonary steal syndrome (PSS) in our clinic between January 2011 and October 2023.

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Results: We had 6 patients. The mean age of patients at diagnosis was 28 months (1-103 months). Two of our patients had Tetralogy of Fallot, right aortic arch, SSS and PSS and those patients had surgery. Our third patient had Pierre Robin Syndrome, supracardiac total anomalous pulmonary venous drainage (TAPVD), small VSD, large ASD, SSS and PSS. He was also operated. In our fourth patient, it was observed that there was coarctation of the isthmic aorta, only the carotid artery emerged from the arch on the left, and the left subclavian artery was filled with collaterals from the vertebral arteries in the late retrograde phase and was connected just proximal to the coarctation site. Additionally, he underwent surgery. A right aortic arch was seen in two of our patients. The descending aorta's collaterals were directed toward the left shoulder, where the internal carotid artery and left subclavian artery emerged. From there, they were connected to the main pulmonary artery via an atypical duct, which is the SSS and PSS. The pulmonary artery was simultaneously taking blood from the isolated left subclavian artery through the ductus, while the subclavian artery was taking blood from the left vertebral artery. The duct was entered antegradely through the pulmonary artery. The first patient's PDA was closed with a vascular plug, and the second patient's PDA was closed with ADO II. Both patients had no complaints after the procedure.
Conclusions: SSS is asymptomatic in most patients and does not require invasive evaluation and treatment. Two patients in whom we performed transcatheter intervention had both SSS and PSS. Since SSS and PSS may cause neurological sequelae and congestive heart failure, we recommend closure of the PDA via transcatheter in these cases.

Keywords: children, pulmonary steal syndrome, subclavian steal syndrome, transcatheter closure

Images of two patients in whom we performed transcatheter intervention had both SSS and PSS.

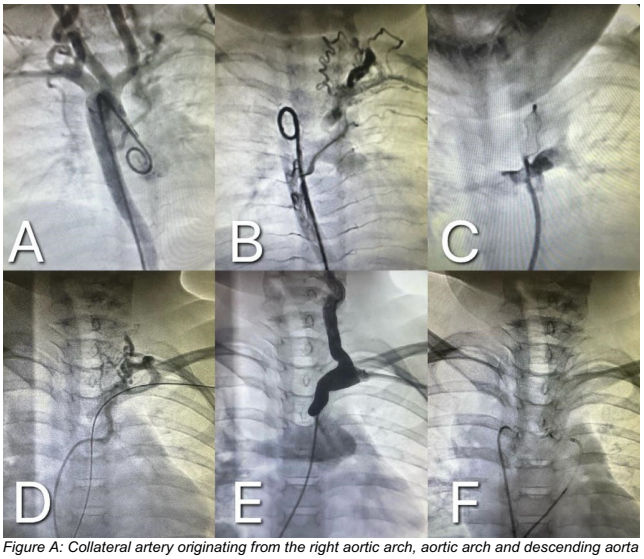

Figure A: Collateral artery originating from the right aortic arch, aortic arch and descending aorta Figure B: Stealing of blood from the vertebral artery and subclavian artery into the pulmonary artery through the vertical PDA Figure C: Vertical PDA closed by entering via antegrade route Figure D: Collateral arteries originating from the descending aorta Figure E: Stealing of left carotid artery and subclavian artery blood to the pulmonary artery with vertical PDA Figure F: Vertical PDA closed by entering via antegrade route

PP-740

Stenting of critical aortic coarctation in very low birth weight preterm neonate: A bridge to surgical correction

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Background and Aim: Surgical correction of the coarctation of the aorta (CoA) in preterm newborns with low body weight is challenging due to their frailty and high risk of complications. Interventional stenting of the aorta offers a viable therapeutic option and can serve as a bridge to surgery in this patient population.
Method: The interventional coronary stent implantation served as an effective bridge-to-surgery strategy for the very low birth weight neonate with critical CoA.
Results: We described the case of a preterm newborn from twin pregnancy with a birth weight of 1400g, born in the 32nd week with intrauterine growth restriction. The diagnosis of critical CoA was suspected on the third day of life due to absent lower limb pulses and oliguria. Prostacyclin therapy was initiated, and the patient was transferred to a cardiac center. Echocardiography confirmed the critical CoA with closed ductus arteriosus. Following escalation of prostacyclin therapy, the ductus arteriosus opened, resulting in symptoms of overcirculation and an echocardiographic gradient of 38mmHg in the isthmic region of the aorta. CT angiography confirmed tubular hypoplasia of the aortic isthmus with a length of 6mm and a diameter of 1.5mm at the narrowest point. The patient was scheduled for interventional stent implantation. Arterial access was obtained through surgical cutdown cannulation of the common carotid artery and two coronary stents were implanted (3.5mm x 8mm and 3.5mm x 13mm) to the aortic isthmus. Following the procedure, the gradient in the stented region on echocardiography measured 25mmHg. The patient remained stable during a follow-up period without clinical symptoms of recoarctation or elevation of NTproBNP. After a 15 month interval, both echocardiographic and non-invasive gradients within the isthmic region notably rose, and CT angiography revealed that the stented segment measured approximately 50% of diameter of the descending aorta. The patient underwent stent explantation surgery at the age of 17 months.
Conclusions: Aortic stenting emerges as a viable approach as a bridge to a complete surgical correction for very low birth weight neonates diagnosed with critical CoA. Surgical cutdown cannulation of the common carotid artery might protect the artery from stenosis and occlusion.

Keywords: Coarctation of the Aorta, Aortic Stenting, Congenital Heart Defect, Interventional Cardiology, Preterm Neonate

PP-741

Outcomes of transcatheter closure of congenital left circumflex coronary artery fistula

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Background and Aim: Congenital left circumflex coronary artery fistula (LCX-CAF) is a relatively rare type of coronary artery fistula,

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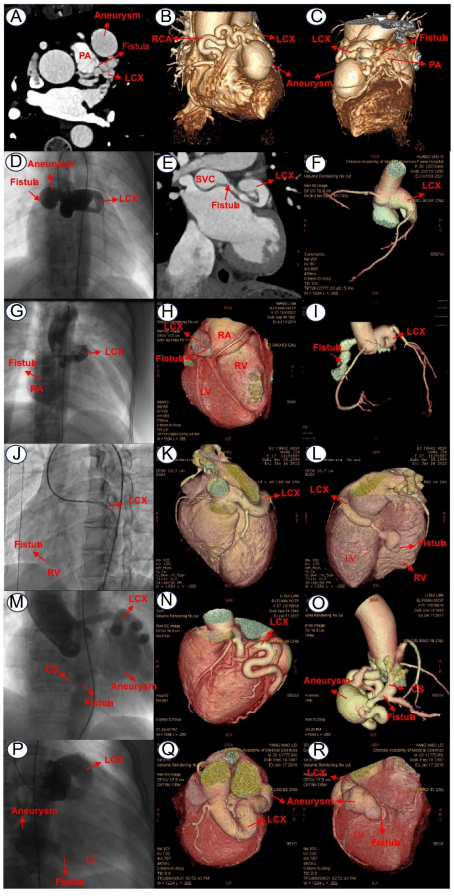
little is known about the outcomes of transcatheter closure of LCX-CAF.
Method: All consecutive patients admitted to Fuwai Hospital and scheduled for transcatheter closure of LCX-CAF from January 2012 to December 2022 were retrospectively reviewed. Anatomic characteristics of LCX-CAF, procedural techniques, and outcomes were summarized and further assessed.
Results: Twenty-five consecutive patients (age:34±20 years, 48 % males) were admitted to our hospital and scheduled for transcatheter closure of congenital LCX-CAF. The mean diameter of the fistulas was 6.99±2.04mm with 84% (N=21) having large fistulas. Transcatheter closure was infeasible for 12% (3/25) in patients scheduled for the procedure. Among 22 patients ultimately receiving transcatheter closure, the procedural success rate was 77.3% (17/22) with a mean time from the initial angiography to the first occluder deployment of 79.95±35.94 minutes. Occlusion devices included ventricular septal defect occluder (n=1, 4.5%), vascular plug (n=3, 13.6%), coils (n=4, 18.2%), and patent ductus arteriosus occluder (n=14, 63.6%). The occluders were deployed via transarterial approach and arteriovenous loop in 6 (27.3%) and 16 patients (72.7%), respectively. No procedural complication was recorded. The postoperative in-hospital length of stay was 4(1-10) days. The incidence of transient ST-T segment change on electrocardiography was 18.1% (N=4).The mean follow-up time was 62.2±45.5 months. The incidence of myocardial infarction and recanalization of the fistula was 4.5% (1/22) and 9.1% (2/22), respectively. The majority (15/22, 68.9%) were in New York Heart Association functional class I.
Conclusions: ranscatheter closure of LCX-CAF is a feasible and effective alternative to surgical repair with comparable outcomes in selected patients. Optimal medical therapy to prevent post-closure myocardial infarction requires further investigation.

Keywords: Congenital coronary artery fistula, Left circumflex coronary artery fistula.

Figure 1



(A-C) LCX to PA fistula; (D-F) LCX to SVC fistula; (G-I) LCX to RA fistula; (J-L) LCX to RV fistula; (M-O) LCX to CS fistula; (P-R) LCX to LV fistula. CS, coronary Sinus; LAD, left anterior descending coronary artery; LCX, left circumflex coronary artery; LV, left ventricle; PA, Pulmonary Artery; RA, right atrium; RCA, right coronary artery; RV, right ventricle; SVC, superior vena cava.

PP-742

Percutaneous occlusion of cardiovascular malformations with the new penumbra coils

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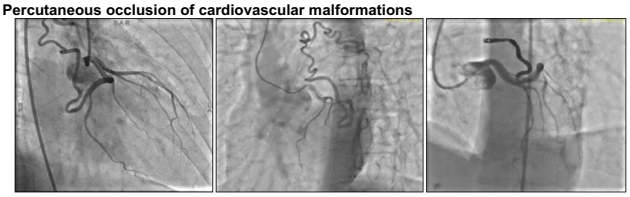
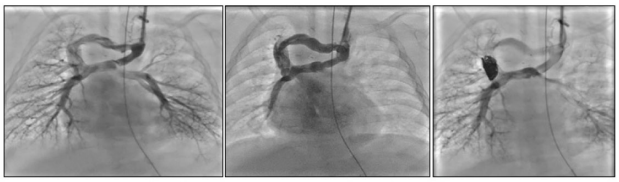
Background and Aim: Percutaneous vessel embolization is usually the preferred treatment method to close congenital or acquired cardiovascular malformations (CVM). Tortuous vessel course, large vessel size and small patients may impede or difficult percutaneous closure. Penumbra have three type of large volume coils (POD, Ruby and Packing), with a wide range of sizes (diameter and length), deliverable through low profile and flexible microcatheters, with a controlled delivery system, which make it particularly interesting for the occlusion of cardiovascular malformations. A case series on different cardiovascular malformations treated with Penumbra Coils in children and adults is presented.
Method: Retrospective analysis of all patients who underwent percutaneous occlusion of VM with Penumbra Coils in a single center. Clinical and angiographic data, procedural details, implanted devices, and complications were assessed. Procedural success was defined as effective device deployment with none or minimal residual flow.
Results: A total of 7 VM were intervened in 6 patients using 11 coils, with median age of 22 years (0.5-62 years). The VM had a mean diameter of 4.5mm (2.1-8.3mm) and included 4 aortopulmonary collaterals, 1 pulmonary arteriovenous fistulae, 1 vertical vein and 1 coronary fistulae. Successful occlusion was achieved in 7 (100%) VM. No clinically relevant complication occurred.

Percutaneous occlusion of cardiovascular malformations



Coronary fistulae occlusion with a ruby coil 4mmx15cm, a POD3 coil 20cm and a packing coil 60cm, in patient with a fistulae from the Cx artery to a subsegmental PA branche.



Vertical vein occlusion with a ruby coil 7mm x 25cm and a packing coil 30cm, after a Glenn surgery in a patient with a complex congenital heart disease.

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Conclusions: To our knowledge, this is the largest series on percutaneous occlusion of CVM with the new Penumbra coils. Percutaneous occlusion was effective and safe. The use of longer and larger volume coils, potentially reduced the procedural time and the total number of devices required. Further studies are required to ascertain the potential impact of Penumbra coils in pediatric and adult cardiac patients with cardiovascular malformations.

Keywords: coil, percutaneous occlusion, vascular malformation

PP-743

Right cardiac catheterization via upper arm venous approach in children and adults with congenital heart diseases

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Background and Aim: The upper extremity venous access is a valid alternative to conventional central access in selected patients referred for Right Heart Catheterization (RHC) due to its low-risk profile and high-success rate. However, data regarding the use of this approach in CHD population are limited. Our purpose is to demonstrate the feasibility of RHC via upper venous approach in children and adults with CHDs.
Method: This was a retrospective, observational, single-center study including 52 patients (11 children and 41 adults) with CHDs underwent RHC via an upper arm vein (basilic, cephalic, or antecubital vein) at our department (Paediatric Cardiology Unit and Adult Congenital Heart Disease Unit, Monaldi Hospital, Naples) between September 2019 and December 2022.
Results: Participant mean age was 43.1±22.3 years (range, 8.4–81.3 years). Mean weight was 67±22.2 kg. Study population included 35 (75%) patients with biventricular anatomy and 13 (25%) with single-ventricle pathology or atrioventricular and/or ventriculoarterial discordance. In this cohort, 50 RHC procedures (96.2%) were successfully performed using an upper arm approach. Right arm access was preferred in 90% of cases. 6-Fr vascular access sheaths were used in 64%, whilst 5-Fr sheaths were used in 34% and 4-Fr in 2% of cases. Concomitant right angiography and coronary angiography were performed respectively in 9 (18%) and in 10 (20%) patients. Access-site switch to femoral access occurred in just one case in which it was required larger sheath for the treatment of Fontan conduit stenosis. Only 1 patient developed a minor adverse event (cephalic vein thrombosis). No irreversible and/or life-threating adverse events were detected.
Conclusions: The upper arm veins are safe and effective to perform a RHC in children and adults with CHD. This approach demonstrates a high percentage of technical success, and few mild complications.

Keywords: Right heart catheterization, Upper arm veins, Pediatric Interventional Cardiology

PP-744

Postoperative catheter thrombaspiration to open an occluded right ventricle-to-pulmonary artery conduit in child

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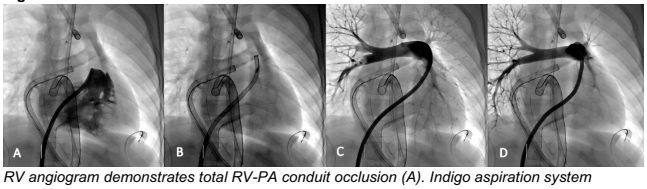
Background and Aim: We report on catheter thrombaspiration to open an occluded right ventricle-to-pulmonary artery (RV-PA) conduit on day 6 postoperative in a 6-year-old girl in the setting of a Ross procedure that was performed for severe native aortic valve insufficiency (AVI) secondary to late diagnosis of bacterial endocarditis.
Method: A 6-year-old girl (18kg/120cm) was referred overseas to our institution for severe native AVI secondary to a late diagnosis of Methicillin-sensitive Staphylococcus Aureus endocarditis. Despite intravenous antibiotics, the patient experienced rapid worsening of the AVI within a week before her referral. She developed refractory congestive heart failure and underwent an urgent Ross procedure with the placement of an RV-PA 18mm Contegra VenPro conduit. At the end of the surgery, the patient had left ventricle (LV) akinesia and required central veno-arterial extracorporeal membrane oxygenation (ECMO) with full assistance and a trans-mitral LV decompression cannula. On postoperative day 6, the right ventricle (RV) was severely dilated despite the support and there was no anterograde conduit flow. Cardiac CT scan suggested complete conduit thrombosis and a head scan showed no active bleeding.
Results: Angiography demonstrated complete conduit occlusion (Fig. 1A). From the femoral vein, we advanced a 12-Fr 115cm long Indigo® CAT12 aspiration catheter from Penumbra® (Alameda, USA) and performed a thrombectomy (Fig. 1B). Subsequent angiography showed a re-canalized conduit and a stiff wire was positioned in the right pulmonary artery (RPA). We performed another set of aspiration thrombectomies followed by balloon angioplasties for smaller sub-occlusive RPA thrombi (Fig. 1C). Exit angiography showed near-total thrombus removal and flow restoration (Fig. 1D). The patient was weaned from ECMO as follow-up ultrasounds showed significant improvement in the LV function and normalized RV function. On postoperative day 10, LV function was good enough to remove the ECMO, replace the conduit, and close the chest. Four days later, total body scan showed no secondary lesion. Discharge brain MRI was unchanged. 3-month follow-up outcomes are good.
Conclusions: The indigo aspiration system is a viable new tool in the armamentarium of pediatric cardiologists to establish the patency of thrombosed vasculatures in children without increasing the systemic risk of bleeding.

Keywords: children, congenital heart disease, mechanical thrombectomy, transcatheter intervention, thrombosis

Fig. 1



RV angiogram demonstrates total RV-PA conduit occlusion (A). Indigo aspiration system advanced within the conduit after thrombectomy (B). Sub-occlusive thrombi in the right pulmonary artery (C). Exit angiography demonstrates near-total thrombus removal and flow restoration (D).

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PP-745

Cerebral embolic protection during transcatheter stent expansion of restrictive extracardiac fontan conduit

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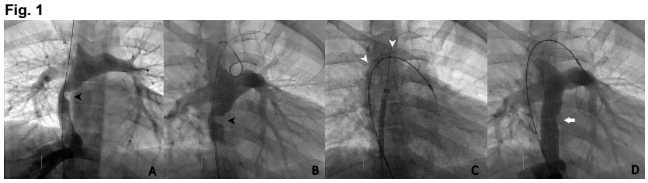
Background and Aim: We report a temporary cerebral protection from paradoxical embolism with TriGUARD-3 (Keystone Heart). to perform a transcatheter stent expansion of an extra-cardiac fenestrated Fontan conduit that was partially obstructed with a large protruding calcified thrombus at the level of the Fenestration.
Method: A 20-year-old female patient (76Kg/164cm) was referred for progressively worsening exertional dyspnea and oxygen desaturation. The patient had a hypoplastic left heart and was palliated with a fenestrated extra-cardiac Fontan (18mm Gore-Tex tube) at the age of five years. She has a history of spontaneous closure of the fenestration shunt during follow-up and the warfarin therapy was switched to daily oral antiplatelet therapy at the age of 14 years for treatment non-compliance. A diagnostic cardiac catheterization showed low central venous pressure and no gradient pressure over the left outflow tract. Angiography showed a large calcified thrombus at the level of the Fontan fenestration, protruding inside the extra-cardiac conduit and reducing the inner diameter by half (Fig.1A). There was an invasive gradient of 2 mmHg across the thrombus. The case was discussed and the patient was scheduled for a conduit stent expansion to relieve the central venous obstacle under cerebral embolic protection.
Results: Hemodynamic measurements and caval angiography were done. The inner diameter of the conduit at the level of the thrombus was 7mm (Fig.1B). The invasive gradient across the thrombus was 3mmHg. Over a stiff wire, the TriGUARD-3 system was advanced sheathless from the left femoral artery into the aortic arch under fluoroscopic guidance. The device was expanded and positioned, for the first attempt to accurately cover the ostia of the innominate, left common carotid, and subclavian arteries (Fig.1C). From the femoral vein, we implanted a 48mm long XXL PTFE-Covered Optimus-CVS® (AndraTec, Germany) using a 20mm AltoSa-XL single balloon catheter (AndraTec, Germany). The stent was re-dilated using an ultra-high-pressure balloon to reduce the stent waist. Exit angiography showed no vascular lesion and good stent apposition to the vascular wall (Fig.1D). The invasive gradient was abolished. The TriGUARD-3 system was easily removed.
Conclusions: Temporary cerebral embolic protection devices can help divert potential paradoxical emboli downstream and allow adequate cerebral blood perfusion.

Keywords: congenital heart disease, embolic protection devices; Fontan circulation; stent interventions.

Fig. 1



First caval angiography showing a large calcified thrombus at the level of the Fontan fenestration, protruding inside the lumen of the conduit and reducing the diameter by half (A). First caval angiography (12 months later) showing an increase in the size of the thrombus with an inner diameter of the conduit of 7 mm at the level of the thrombus (B). TriGUARD 3™ deflection filter device positioned in the aortic arch providing full coverage of all three major branches before the delivery of a 48 mm long XXL PTFE-covered Optimus-CVS® (C). Exit angiography showing no vascular lesion and good stent apposition to the vascular wall (D).

PP-746

Proof of concept: A new solution for low-profile transcatheter implantation of optimus-l stents in small children

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Background and Aim: There is no stent designed or approved for use in infants. We sought to obtain in-vitro and in-vivo data on a new concept conceived to implant Optimus-L stents at infant vessel diameters and offer a potential long-term stent solution.
Method: Nineteen Optimus-L stents were mounted on 8 types of angioplasty balloons with diameters 6, 8, and 10mm using an injection-molded hand-crimper. We evaluated balloon-stent unit (BSU) stability before insertion and advancement through short Terumo introducers with incremental French (Fr)-size and possibility of side-arm contrast injections. Three types of long sheaths were tested. Stents were inflated to balloon nominal diameters and re-expanded to 18 and 23mm. Stent recoil, foreshortening, and fracture were evaluated. In-vivo implantations were performed afterward.
Results: In-vitro: Medtronic-EvercrossTM balloons and modified Terumo-DestinationTM sheaths were the best combination: BSUs were inserted in 6-Fr sheaths with possible injections (for 6 and 8mm balloons), and 7-Fr sheaths without injections (for the 10mm balloon). Retrieving BSUs inside the sheath required one additional Fr-size. Boston-Scientific-SterlingTM and Balton-Lovix balloons, as well as APT-BraidinTM-L guiding sheaths showed unsatisfactory performance. Dilation up to 23mm was possible, stent shortening was <24% at 18mm and <37% at 23mm. Recoil was limited and no stent fractured. In-vivo: Optimus-L stents were used to treat two infants with aortic coarctation and two children with pulmonary artery stenosis using 8mm balloons and low-profile access.
Conclusions: Optimus-L stents can be implanted safely in small patients with a low-profile approach. These stents have the potential to achieve adult size while maintaining structural integrity.

Keywords: Congenital heart disease, children, infants, innovation, stent, transcatheter interventions.

PP-747

Transcatheter management of preterm PDA – can an educational CPD event create awareness?

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Background and Aim: Significant variation exists in the initial management of hemodynamically significant preterm PDA (hs-PDA). Referral for closure thresholds vary depending on neonatologists and individual centres. Current trials are not in favour of early medical treatment. With the development of transcatheter preterm PDA programmes, neonatologists now have another option to consider in the management of babies with hs-PDA.
Method: A preterm PDA CPD event was co-organized between the East and West Midlands Cardiac and Neonatal Networks. Attendees were given a questionnaire on arrival to explore their views around preterm PDA management. Following the end of

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the meeting an electronic survey was sent to all attendees. We specifically looked at the responses from the neonatal community.
Results: A total of 48 pre-meeting responses were collected. Ibuprofen was the preferred initial therapy in 71% (n=34), while 29% (n=14) favoured diuretics. None favoured transcatheter device closure. 54% (n=26) of participants demonstrated awareness of transcatheter device closure as a potential treatment option; however 83% (n=40) felt they were not aware of what the referral criteria for device closure was. There were 43 responses post meeting. 88% (n=38) of respondents reported a favourable impact of the meeting on their views regarding pre-term PDA management. 74% (n=32) favoured transcatheter device closure, 14% (n=6) preferred surgical ligation and 12% (n=5) opted for medical management with ibuprofen.
Conclusions: Transcatheter PDA occlusion is a relatively new therapy within the UK. There is a lack of awareness about the potential benefits of this therapy amongst the neonatal community. Educational programmes can provide the additional information required to increase awareness of the possibility of a transcatheter approach. Further research is required to understand where transcatheter PDA closure should fit in overall treatment protocols for preterm PDA

Keywords: Trans-catheter PDA closure, preterm PDA, neonatology

PP-748

Delayed complete atrioventricular block after transcatheter perimembranous ventricular septal defect closure

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Background and Aim: The soft nitinol KONAR-MF™ ventricular septal defect (VSD) Occluder (MultiFunctional Occluder MFO, Lifetech Scientific) is one of the seldom devices that have obtained the CE mark for transcatheter perimembranous VSD closure. Complete atrioventricular block with the MFO device is rare: the mean rate is less than 1%. Only cases of acute complete atrioventricular block within the few days following device implantation have been reported in patients without preprocedural high-grade atrioventricular block. The aim was to describe here a case of delayed complete atrioventricular block after VSD closure using the MFO device.
Method: After informed written consent from the patient, data were collected about the patient's medical history, the procedure of VSD closure, and outcome.
Results: A 52-year-old man was addressed to the congenital cardiology clinic for exertional dyspnoea. The electrocardiogram showed a complete right bundle branch block with normal PR interval. Transthoracic and transoesophageal echography showed an isolated perimembranous VSD with left atrial dilation, the VSD measured 10 mm on the left side and 6 mm on the right side with a small aneurysm. The VSD was closed percutaneously via a retrograde approach using a 10/8 MFO device without acute complication. The repeated post-procedural electrocardiograms were unchanged and the patient was discharged at 48 hours post-procedure. Follow-up was uneventful until 20 months after the procedure where the patient had fatigue for several weeks followed by episodes of lipothymia. The electrocardiogram showed a complete atrioventricular block with severe bradycardia requiring isoprenaline: a dual chamber pacemaker was implanted.
Conclusions: We report for the first time to our knowledge a case of delayed complete atrioventricular block necessitating the implantation of a pacemaker 20 months after VSD perimembranous closure with the MFO device.

Keywords: Congenital heart disease, case report, high-grade block, left-right shunt, heart failure, arrhythmia

PP-750

Percutaneous pulmonary thrombectomy in an adolescent fontan patient

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Background and Aim: This case report details a percutaneous pulmonary thrombectomy in an 18-year-old female with a Fontan circulation. The patient has a bilateral Glenn and an 18 mm conduit. The fenestration has been device closed and later a stent has been placed in the conduit. A diagnostic catheterization (100 IE/kg heparin IV) one week before admission showed a central venous pressure of 22 mmHg and no signs of thrombo-embolism. Chronic medication was acetylsalicylic acid 80 mg once daily.
The patient presented with acute respiratory distress, tachypnea, and exercise intolerance but was hemodynamically stable. Computed tomography angiography revealed extensive emboli in the right interlobar artery and left lower lobe artery (Figure 1-2).
Method: Under general anesthesia, a 24F sheath (Dryseal, Gore Medical, Newark, Delaware, US) was inserted in the right femoral vein. Pulmonary angiography revealed subtotal occlusion of the right interlobar artery and left lower lobe artery (Figure 3-4).
Results: Aspiration thrombectomy of both pulmonary arteries was performed using 24F and 16F aspiration catheters (FlowTriever, Inari Medical, Irvine, California, US), yielding large amounts of thrombus material (Figure 5) and resulting in subtotal angiographic recanalization (Figure 6-7). Rivaroxaban 15 mg BID was started after continuous heparin IV.
Conclusions: Thromboembolic events are common in Fontan patients, occurring in up to 25% of adult patients. (1) Predisposing factors here are slow flow, increased venous pressure, oral contraception, smoking, lifestyle and recent intervention. Recent guidelines recommend lifelong direct oral anticoagulants over vitamin K antagonists, after a thromboembolic event in Fontan patients. (2) In recent years, catheter-directed pulmonary aspiration thrombectomy has been proposed as a novel therapeutic technique in intermediate and high-risk acute pulmonary embolism and has shown excellent results in terms of safety, immediate effectiveness and reduction of major bleeding. (3, 4) Data on its application in patients with complex congenital cardiac abnormalities is scarce. The use of the more recently introduced extra-large caliber devices in these patients has not been reported before. (5) Our case report demonstrates that percutaneous pulmonary thrombectomy using large caliber devices up to 24F can be safe and effective in young Fontan patients with thromboembolic complications. Endovascular interventions and oral anticoagulation regimens should be tailored to the individual patient.

Keywords: Pulmonary Thrombectomy, Pulmonary Embolism, Fontan

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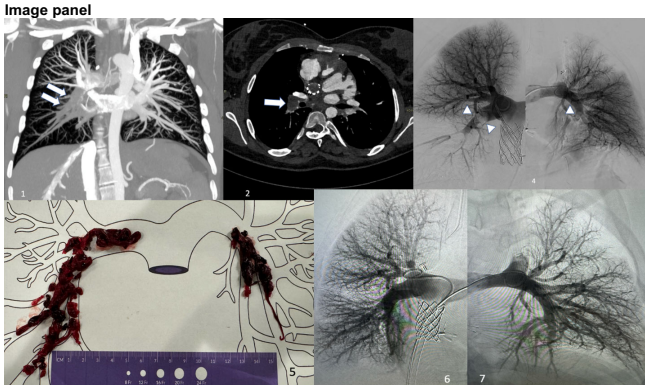
Image panel



PP-752
EARLY HEART BLOCK AFTER TRANSCATHETER CLOSURE OF VENTRICULAR SEPTAL DEFECT

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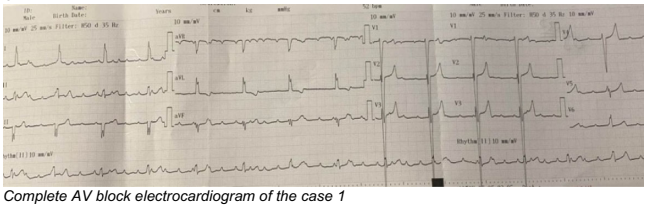
Background and Aim: Left bundle branch block(LBBB) and complete atrioventricular block (CAVB) are rare but important complications after transcatheter closure of PMVSDs that can adversely affect left ventricular function. In our article, 4 cases in which heart block developed early during transcatheter VSD closure are reported.
Method: Case 1:A 2-year-old girl was followed up with a diagnosis of VSD.The VSD was successfully closed with the Lifetech Konar MFO 6-4 device. Electrocardiograms taken during and after the procedure showed no conduction disturbances. Three days after discharge, he was brought to the ER with convulsions. ECG performed revealed CAVB. Since CAVB persisted, a temporary transvenous pacemaker was placed. Dexamethasone 0.6mg/kg/day was started. On the 3rd day of hospitalization, it was observed that he returned to sinus rhythm. Case 2: The ventricular septal defect of a 15-month-old patient was closed. After placement of the device, QRS complex widening was observed on the monitor. The device was withdrawn without release. Dexamethasone was started for the patient with persistent LBBB. In the 1st week of post-procedure follow-up, ECG showed sinus rhythm and no LBBB was observed. Case 3: An 8-month-old patient had closure of the VSD. Dexamethasone was started in the patient whose ECG showed a LBBB.Three weeks later ECG showed the disappearance of the LBBB. Case 4:The VSD of a 14-month-old girl was closed transcatheterly.The LBBB was observed in the ECG taken after the procedure. Dexamethasone was started.The device was surgically removed because the LBBB did not regress on follow-up ECGs.
Results: Atrioventricular block after transcatheter device closure of PMVSDs is due to the proximity of the conduction bundle to the defect. The reported rates of AVB in the literature range from 0% to 5.7%. AVB may generally occur immediately after the procedure or after a while percutaneous closure of the defect.
Conclusions: Young age, low body weight, presence of ventricular septal aneurysm, and choice of large device are important contributing factors for the development of AVB. We believe that low body weight and young age contributed to the development of AVB in our patients.

Keywords: ventricular septal defect, left bundle branch block, complete atrioventricular block, transcatheter closure

CAVB



Complete AV block electrocardiogram of the case 1

PP-754
A treacherous path in the CATH LAB to reach the goal

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Background and Aim: Congenital systemic venous anomalies are known to surprise pediatric cardiologists in catheterization lab as well as in operation theatre if they are undetected or misdiagnosed. Here, we report an unexpected Inferior vena cava (IVC) anomaly complicating Atrial Septal Defect (ASD) device closure in a case otherwise thought to be straight-forward.
Method: A six year old child with 11 mm secundum ASD (left to right shunt) with right sided volume overload with apparently adequate rims (Poor transthoracic acoustic windows) was taken for device closure with Trans esophageal echocardiography (TEE) guidance under general anesthesia. Pre- procedure Echo had shown apparent patent intrahepatic Inferior Vena cava draining into Right atrium.
Right Femoral venous access was first attempted, however there was inability to pass the guide wire. On contrast injection, the drainage into IVC was blocked with drainage via paravertebral plexus. Left femoral side puncture encountered a similar issue with contrast showing hemi-azygous continuation and drainage into right sided superior vena cava.
Results: This landed us in a conundrum and a quick decision of device closure via right internal jugular vein was taken. Through Trans jugular route we could identify that Hepatic segment of IVC was however patent and draining normally into RA, hence masquerading itself from detection on routine echocardiography. It, then took almost 2 hours to place the wire in a pulmonary vein after a lot of attempts and maneuvers. Finally with help of internal mammary artery (IMA) catheter we were able to place the wire in left upper pulmonary vein, just before we were about to abandon the procedure. Subsequently, we succeeded in deploying Amplatzer 16mm septal occluder.
Conclusions: Although rare, the possibility of IVC anomaly exists even if a well patent, normal calibre hepatic segment of IVC is

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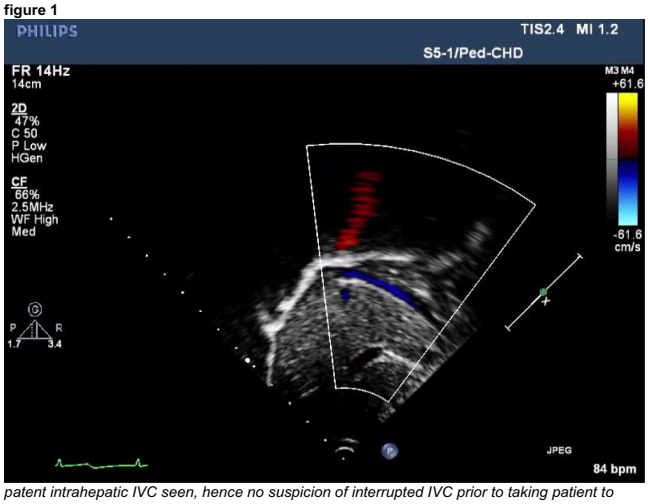
visualized on transthoracic echocardiography. Trans-jugular venous approach for device closure of ASD may not be as easy as it is thought but should be considered in such cases.

Keywords: Atrial septal defect device closure, Interrupted IVC

figure 1



patent intrahepatic IVC seen, hence no suspicion of interrupted IVC prior to taking patient to Cath lab

PP-755

Post-operative complication after percutaneous atrial septal defect closure with gco device in a pediatric population: a multicentric Italian experien

Silvia Teresa Scalera[1], Alessandra Pizzuto[2], Mario Giordano[3], Giampiero Gaio[3], Maria Giovanna Russo[3], Gianfranco Butera[1], Mara Pilati[1], Martino Cheli[5], Maurizio Marasini[5], Roberto Formigari[5], Biagio Castaldi[4], Domenico Sirico[4], Francesco Prati[4], Giovanni Di Salvo[4] and Giuseppe Santoro[2]

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Background and Aim: This multicenter observational study assessed the post-operative complications of the GORE® Cardioform ASD Occluder (WL Gore & Associates, Flagstaff, AZ). This device has theoretical advantages over other commercialized devices due to its softness and anatomical compliance. Our aim was to evaluate short- and medium-term post-operative complications after percutaneous ASD closure with GCO in a pediatric population.
Method: We enrolled 250 patients with ASD submitted to transcatheter closure from January 2020 to October 2023 at four high-volume Italian Pediatric Cardiology centers. Primary endpoint was safety in post-operative follow-up. We have analyzed post procedure complications such as device-related thrombosis, device-related infective endocarditis and tachy/brady-arrhythmias.
Results: Patients' age and weight media were 10,9 years (median 8.9, IQR 5-10,25) and 33,18 kg (median 25 Kg, IQR 19,7-42), respectively. Median ASD diameter was 25,5 mm (IQR 15-22), resulting in QP/QS of 2,4 (median 2,25, IQR 1,3-3,11). After device deployment, three (1,2%) patients developed transient junctional rhythm treated with corticosteroid therapy. One of them needed a short course of Flecainide for atrial ectopic tachycardia. No tachy/brady-arrhythmias were recorded at follow-up. Four patients (1,6%) developed transient atrio-ventricular block. Six patients (2,4%) needed surgical device retrieve; three rescue surgery due to device embolization, one patient underwent to elective surgery for new-onset tricuspid valve regurgitation. Two patients experienced device thrombosis. The patient with early onset underwent surgical management through device removal, while the other patient, diagnosed with thrombosis at the 1-month follow-up, was managed with medical therapy involving anticoagulation, leading to resolution after two months of treatment. In two patients (0,8%), non-hemodynamically significant pericardial effusion was observed, which resolved following corticosteroid therapy.
In summary, post-operative complications were observed in 6.8%, of which only 2.8% required surgery. No device-related infective endocarditis was recorded at follow-up.
Only 2.8% of patients experienced arrhythmic complications, and all resolved without the need for device removal. Device thrombosis and pericardial effusion were observed in both, each in 0.8% of patients.
Conclusions: The transcatheter closure with Gore Cardioform ASD Occluder demonstrates a low rate of post-operative complications, with only a few patients requiring device removal surgery.

Keywords: ASD, GSO, percutaneous, device, post-operative complications.

PP-756

Cardiac catheterization in low weight infants: Experience of a single center

Silvia Teresa Scalera, Mara Pilati, Micol Rebonato, Enrico Piccinelli, Marco Masci, Marco Campanale, Patrizio Moras, Alessandra Toscano, Lorenzo Galletti and Gianfranco Butera

Cardiology, Cardiac Surgery and heart Lung Transplantation; ERN GUARD HEART: Bambino Gesù Hospital and Research Institute, IRCCS, Rome Italy

Background and Aim: In the last 10 years improvements in technical equipments and skills in catheterization laboratories allowed percutaneous treatment of patients with increasingly lower body weight. In literature only few reports describe what kind of interventional procedures are feasible in patients weighing less than 2 Kg and their results.Aim of our study is to describe all the percutaneous procedures performed in our center in patients weighing less than 2 Kg.
Method: All cardiac catheterizations and medical reports of patients undergoing percutaneous procedures with a weight less than 2 Kg, in the period from 2018 to 2023, were retrospectively reviewed.We collected all data including cardiac diagnosis,type of procedure,complication rate and we compare them with the CRISP score.
Results: In our center from 2018 to 2023, 38 patients weighing less than 2 Kg underwent a percutaneous procedure.All the patients were premature, with a median gestational age of 33 weeks (IQR 30-35 weeks), median weight was 1.7 Kg (range min-max 720 g-2 Kg),and median age at catheterization was 14 days (IQR 2-25 days).Diagnosis was:patent ductus arteriosus (39%), critical aortic valve stenosis (13%), truncus arteriosus (5%), critical pulmonary valve stenosis (5%), tetralogy of Fallot with pulmonary atresia (10%) and miscellaneous (38%). Intended procedures were: duct closure (39%), balloon valvuloplasty (23%), ductal stenting (13%), interatrial stenting (5%), pericardiocentesis (5%), percutaneous pulmonary banding (2%) one diagnostic procedure (2%), one Rashkind procedure (2%) and one central venous stenting (2%). Successful procedure rate was 97%,one interatrial stenting failed

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and atrial septostomy was performed instead. Procedural complications occurred in 3 patients (8%):one inferior vena cava dissection,one pericardial effusion and one stent displacement.None of these complications needed surgical treatment. Our population complication rate was slightly lower than that calculated with CRISP score (10.3%). Need for transfusion was registered in 26 patients (68%). At a median follow up of 30 months (IQR 3-26 months)10 patients died for comorbidities related to prematurity, 5 patients underwent another percutaneous procedure and 1 needed a surgical procedure.
Conclusions: In our experience, cardiac catheterization in children weighing less than 2 Kg is usually interventional, has a high procedural success rate with a complication rate lower than that predicted with CRISP score.

Keywords: low weight infants, CRISP score

PP-757

Developing a CMR-guided cardiac catheterization program: Challenges and initial experience

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Background and Aim: Congenital Heart Disease (CHD) affects a significant number of newborns. Long-term survival to adulthood is common with current medical and surgical treatments. Current radiographic techniques hinder effective interventions due to poor soft tissue visualization, prolonged procedures and increased radiation exposure. Traditional methods for assessing hemodynamics, like thermodilution and Fick's principle face inaccuracies. In contrast CMR's velocity-encoded phase contrast accurately measure stroke volume, informing diagnoses and treatment decisions.
This study aims to validate the efficacy of calculating cardiac output and vascular resistance by integrating invasive hemodynamic values with MRI-derived flows. We seek to establish the safety and feasibility of combining cardiac catheterization and CMR to minimize radiation exposure in CHD.
Method: A cardiac catheterization was performed in a suite equipped with a 1.5 T CMR scanner. A CMR-compatible monitor was used to mirror the scanner display for in-room catheter visualization. A single lumen balloon wedge-pressure end-hole catheter, a pigtail catheter and the Emery Glide Straight tip MR Wire were used for all cardiac catheterizations. Cardiac and pulmonary index were computed using both Fick's method and MRI.
Results: Fifteen patients underwent MRI-guided cardiac catheterization between May 2022 and November 2023. The diagnosis were 6 univentricular hearts and 9 primitive cardiomyopathies. The median age was 10 years (IQR 7-19,5y) and median body weight was 40 Kg(IQR27,5-57 Kg). Median cardiac index was 2,65 L/min/m2 (IQR 2,45-3,02) by CMR flow vs. 3 L/min/m2 (IQR 2,7-3,9; p=0,7) by Fick methos. Median PVR was 1,9 WUmq (IQR1,45-2,47) by CMR flow vs. 1,9WUmq (IQR1,6-2,1; p=0,04) by. Fick method. The procedural success rate was 93,3%, with a median total procedural time of 122 minutes. No complications occurred.
Conclusions: This study contributes to the ongoing validation of hemodynamics measurement acquired through CMR. The utilization of CMR evaluations proves to be instrumental in enhancing the reproducibility of hemodynamic assessments within CHD population. The determination of pulmonary vascular resistance (PVR) remains crucial for risk stratification in CHD, impacting intervention decisions. The demonstrated reliability of CMR measurement in this context suggests promising potential for improving the management of congenital heart disease.

Keywords: CMR-cardiac catheterization, congenital, PVR

PP-758

Percutaneous pulmonary artery banding: An alternative to surgery in high-risk patients

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Background and Aim: Pulmonary artery banding procedure is a palliative approach for patients with pulmonary high flow and/or pulmonary hypertension. In low weight infants, surgical treatment is considered high-risk in terms of mortality and morbidity.In literature some reports described a percutaneous approach with the use of microvascular plug (MVP).Aim of our study is to describe the use of MVP for percutaneous pulmonary banding in two infants weighing less than 2 Kg.
Method: Characteristics of two patients with complex congenital heart disease undergoing percutaneous pulmonary banding were collected.
Results: Patient 1 had a diagnosis of truncus arteriosus,with ventricular septal defect and a pseudoaneurysm of the right ventricle. After collegial discussion, a percutaneous approach was chosen, involving the implantation of microvascular plugs (MVP, Medtronic) in the right and left pulmonary arteries (PA).Procedure, performed at a weight of 1.1Kg,consisted in implantation of two devices (MVP7Q-MVP5Q) in the left PA and one device in right PA(MVP8Q).No complications occurred.One month later at the weight of 1.6Kg,because of the persistence of right pulmonary overflow, the 9 mm right device was percutaneously retrieved, and a surgical right PA banding was performed, leading to an improvement in infant's condition. This allowed the attainment of an optimal weight (2,5Kg) for corrective surgery, during which one device was easily removed while that positioned distally in the left PA was left in site. One month after surgical correction, cardiac catheterization showed severe reduction of the flow in the left lower lobe through the MVP5Q.A successful dilation and opening of the device was performed by using 6x20mm Mustang balloons leading to a perfectly non-obstructed PA flow.
Patient 2 had a diagnosis of hemi-truncus causing a left pulmonary high flow state.Surgery was deemed not feasible because of the low weight (2Kg) and comorbidity so we perfomed a percutaneous MVP implantation(MVP7Q) in the left PA with optimal result (doppler-gradient of 60 mmHg and good distal flow) and a rapid

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clinical improvement of the infant.During follow-up, she developed severe diffuse bronchial-tracheal-malacia and she was considered for palliative care and died 6 months later.
Conclusions: Percutaneous banding with MVP is a feasible procedure in selective cases.Larger experience and longer follow-up is needed.

Keywords: Pulmonary banding, MVP, low-weight

PP-759
Emergency re-opening of completely closed ductus venosus - case report of an 890g premature newborn with total anomalous pulmonary venous connection
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Background and Aim: In patients with infracardiac TAPVC pulmonary venous blood is collected in a vertical vein passing into the abdomen. In the majority of cases the vertical vein drains into the portal vein. Patency of ductus venosus (DV) is essential then to bypass the liver. Closure of the DV results in severe obstruction of pulmonary venous blood flow.
Method: We present an 890g premature baby (28th week of gestation) with infracardiac TAPVC following spontaneous closure of DV. As surgical correction was no option because of the prematurity, emergency stenting of DV was attempted.
Results: At the age of 15 days the child was referred from an external hospital with infracardiac TAPVC and evidence of rapidly progressive obstruction of the DV. On admission Doppler-ultrasound revealed complete closure of the DV. Residual patency had been documented immediately prior to referral. Following percutaneous access via the right jugular vein using a 4 Fr sheath the DV was probed with a 2.7F micro catheter under sonographic guidance. After placement of a 0.018" wire via the portal vein to the pulmonary veins a bare metal coronary stent (3.5mm/13mm) was successfully implanted. Because of in-stent stenosis the stent was redilated 5 weeks later with a 6mm balloon. Due to proximal obstruction of the stent by liver tissue, the stent was extended using a second stent (bare metal, 6mm/15mm). At the age of 13 weeks (weight 2.1 kg) surgical repair with connection of the pulmonary vein confluence to the left atrium was performed successfully. The DV was closed interventionally 2 weeks later using a 6mm vascular plug.
Conclusions: In extremely low birth-weight infants with infracardiac TAPVC stenting of DV is a feasible palliative approach to postpone surgical repair. To the best of our knowledge, our case describes the smallest infant treated by DV stenting. Access via the right internal jugular vein offers the best route for interventional procedures involving the DV even in premature patients. All 4 interventions in our patient were performed via this route without complications. Following surgical repair, interventional closure of the DV is necessary to prevent portosystemic shunting.

Keywords: TAPVC, stenting, ductus venosus, extremely low birth-weight infant

PP-760
Aortic coarctation stenting – two decades of experience
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Background and Aim: Coarctation of the aorta (COA) is a congenital cardiac anomaly accounting for 4% to 7% of all congenital heart disease. Intravascular stent therapy for COA has gained acceptance as a primary option for treating native and recurrent COA in adolescents and adults.
Method: Retrospective analysis included 27 patients with aortic coarctation stenting performed at Mother and Child Health Institute of Serbia between 2000 and 2023. Before the COA stenting, computed tomography (Figure 1A) or cardiac nuclear magnetic resonance was performed.
Results: The study included 19 males (13.4±4.4 age; body weight 51.8±15.1 kg). Previous aortic coarctation operations and balloon angioplasty were performed in 13% and 43.5% of patients. Echocardiography findings before the procedure point out an aortic coarctation pressure gradient of 60 (IQR 53.5 – 68.5) and aortic diameter on a diaphragm of 14 mm (IQR 12.2 – 15.0 mm). Near aortic interruption has been seen in % of patients (Figure 1B,C). Manometric examinations before the procedure showed an average peak-to-peak gradient of 38.5±13.9 mmHg. Cheatham platinum (CP) stents were used in 17/27 patients (Figure 1D,E,F), while covered CP stents were used in 37%. Manometric measurements after stenting pointed out a significant decrease in peak-to-peak gradient (38.6±18.9 vs. 9.5±11.9 mmHg; $p < 0.001$). Complications were not observed during the intervention. The follow-up period was 34 mounts (IQR 9.5–71).
Stent re-dilatation required 10 patients. Patients who required stent re-dilation were younger than patients who did not require re-dilatation (12.2, IQR 9–13.4 vs. 14, IQR 11.5–16 years; $p = 0.04$). Additionally, this cohort had a narrower median aortic diameter on a diaphragm compared to the rest of the patients in the group (12.6±0.9 vs. 14.4±1.4 mm; $p = 0,03$). Re-dilatation was performed after a median of 30 mounts (IQR 16.5 – 38.5). In one patient, stent in-stent was placed (CP stent – Z-MED balloon 22 mm) (Figure 1G,H,I).

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Conclusions: COA stenting is a safe and successful procedure to treat native and recurrent COA in adolescents and adults. The need for re-dilatation is more common in young children and those with a smaller diameter of the aorta on the diaphragm.

Keywords: aortic coarctation, stenting, stent re-dilatation

Figure 1.

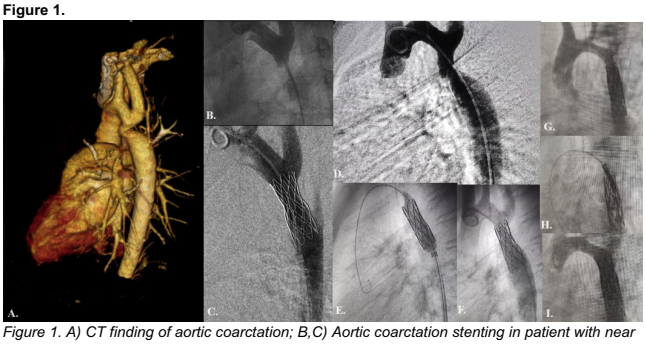

Figure 1. A) CT finding of aortic coarctation; B,C) Aortic coarctation stenting in patient with near aortic coarctation; D,E,F) Aortic coarctation stenting with covered CP stent; G,H,I) Stent in-stent technique in the treatment of aortic coarctation.

PP-761
Congenital descending aorta to right atrial fistula: Transcatheter embolization of a very rare anomaly with PFM coils

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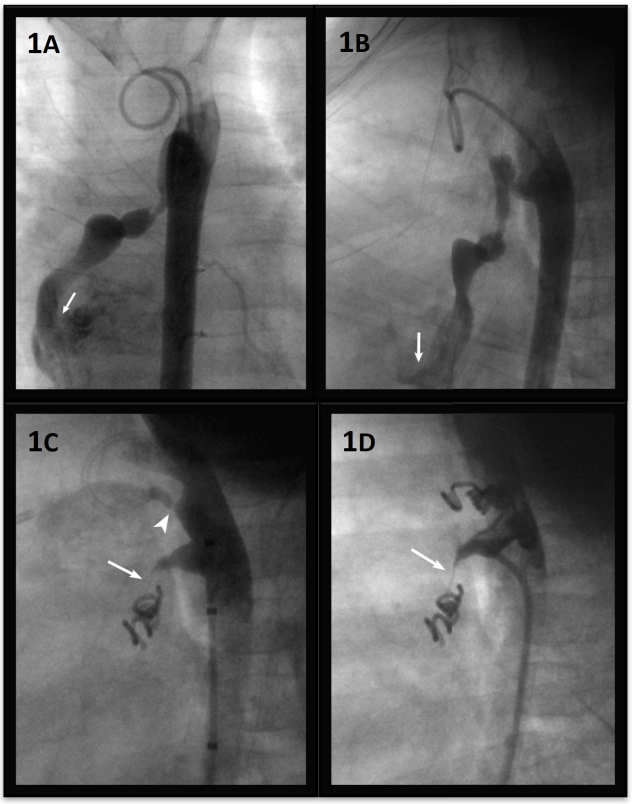
Background and Aim: Communications between the aorta and the right and left atria are uncommon anomalies and usually involve the ascending aorta either in the form of congenital coronary cameral fistulae or acquired connections associated with aortic dissection and endocarditis. Fistulous malformation from the descending aorta is very rare and has only been described in 3previous reports, connecting to the right atrium, left atrium or the superior vena cava, azygos vein and the innominate vein.
Method: We present a 7-month-old 5.6kg girl with trisomy-21 presented to our hospital with tachypnea and failure to thrive. On echocardiography, she had a moderate 10mm secundum atrial septal defect, a small 4mm high muscular ventricular septal defect, a small PDA, severe right ventricular dilation and high-velocity continuous flow entering the RA with 70mmHg gradient. An abnormal tortuous vessel was depicted originating from the descending aorta below the isthmus, while Doppler interrogation of the tricuspid regurgitation and the flow through the ventricular septal defect indicated systemic pulmonary artery pressures.
Results: Angiography depicted the continuous flow vessel with multiple stenotic and dilated parts originating from the descending aorta just below the isthmus and, after a long tortuous course, entering the right atrium (1A-B). After entering the fistula from the right atrial opening, a 5x4mm Nit-Occlud coil system was used in order to achieve a complete occlusion without complications (1C). The PDA was addressed at a next session after patient's short term post intervention significant improvement and satisfactory weight gain (1D). Due to the elevated, albeit improved, pulmonary artery pressures, the presence of trisomy 21 which may accelerate pulmonary vascular disease and the patient's small size rendering interventional closure of her septal defects difficult, the patient few months later underwent surgical closure of her intracardiac defects with a benign postoperative course. Her 3-year clinical and echocardiographic follow-up, demonstrated no pathology.
Conclusions: In summary, this is the first literature report of a descending aorta to right atrial fistula in conjunction with atrial and ventricular septal defects and PDA in a trisomy 21 infant, which caused rapid increase of pulmonary vascular resistance and was successfully occluded by transcatheter placement of Nit-Occlud coil system.

Keywords: Aorta to right atrial fistula, transcatheter occlusion

Figure 1



Frontal (1A) and lateral (1B) projection of descending aortogram depicting a very tortuous vessel with many stenotic and dilated portions originating just below the isthmus and entering the right atrium (arrow). (1C) Lateral projection of descending aortogram 4 months after the fistula closure showing complete occlusion of the fistula by the placed Nit-Occlud coil system with a very narrow tract (arrow) from the ampulla of the fistula to the coil. The arrowhead points to the patent ductus arteriosus. (1D) Lateral projection of descending aortogram after the patent ductus arteriosus closure depicts the Nit-Occlud coil system in the occluded fistula with the narrow tract leading to it and the placed Nit-Occlud coil system above it occluding the patent ductus arteriosus.

PP-762
Interventional treatment options for neonatal aortic thrombosis

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Background and Aim: Neonatal aortic thrombosis, a rare and life-threatening condition, lacks standardized treatment guidelines, especially in cases with end organ failure. We present a case of a 10-day-old neonate with major aortic thrombosis successfully treated using local recanalization and rtPA. Here, we share our

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experience with off-label thrombectomy devices applicable in this patient group.
Method: Complex interventional thrombectomy and local rtPA.
Results: A previously healthy 10-day-old neonate presented with severe dehydration, acute systolic heart failure, marked metabolic acidosis and absent lower extremity pulses. Echocardiography revealed no coarctation but indicated spontaneous major aortic thrombosis with diminished pulsatile flow to the iliac arteries. Due to the patient's critical overall condition and advanced end organ failure, we decided to proceed with interventional thrombectomy. Angiography confirmed complete thrombotic obstruction of the abdominal aorta distal to the superior mesenteric artery with diminished renal perfusion extending into both iliac arteries. During the procedure, we used the Amplatzer Piccolo PDA occluder and an Amplatzer Vascular Plug II (AVP II) in combination with a long 5F sheath from the right iliac artery. However, the Piccolo PDA occluder was not practical for thrombus removal in our case due to mechanical instability. Thrombectomy was completed using AVP II (6/8 mm). After aspiration of thrombotic material, local rtPA administration and balloon angioplasty of the vessels, angiography revealed residual thrombotic material in both iliac arteries. Thus, we initiated one course of systemic rtPA therapy, resulting in mild puncture side bleeding managed conservatively. The patient's overall clinical condition improved immediately post-recanalization. Anticoagulation was continued with heparin and aspirin. The patient was discharged on day 14 with recovered renal function. Partial thrombosis of the right iliac artery with adequate peripheral perfusion was still noted. A comprehensive coagulopathy evaluation will follow.
Conclusions: Interventional thrombectomy is a safe, feasible alternative to surgery for neonatal aortic thrombosis with end organ failure. Given small vessel sizes, on-label thrombectomy devices are not practical in this patient group. Despite being mentioned in an earlier case report, the Amplatzer Piccolo PDA occluder's mechanical instability limited its utility. In our case, off-label use of AVP II proved more effective in a case with more organized thrombotic material.

Figure 1

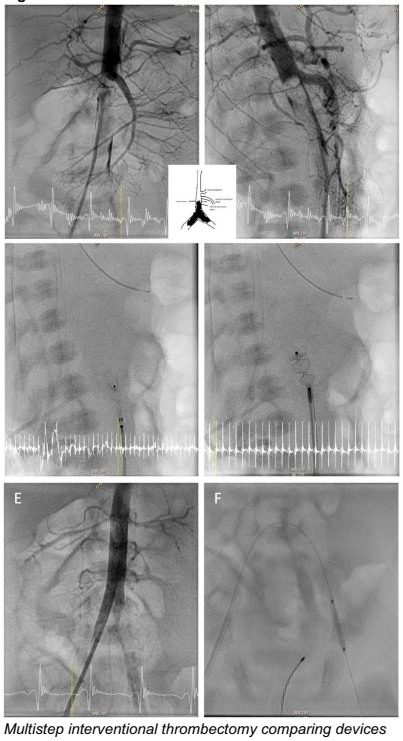

Fig 1 A+B: Pre-intervention angiography with complete thrombotic obstruction of the abdominal aorta distal to the superior mesenteric artery.

Fig 1 C: 1st attempt to thrombectomy using the Amplatzer Piccolo PDA occlude (Abbott, Illinois, USA), note device deformation at thrombus site.

Fig 1 D: 2nd attempt to thrombectomy using the Amplatzer Vascular Plug II (St Jude Medical, Minnesota, USA), device shape retained.

Fig 1 E+F: Balloon angioplasty addressing residual thrombotic material in the iliac artery in crossover technique.

Multistep interventional thrombectomy comparing devices

Keywords: Neonatal aortic thrombosis, Interventions in newborn, Embolectomy device, Angioplasty

PP-763

Who is behind the scene ? — coarctation of aorta presenting as dilated cardiomyopathy: An unusual presentation of CO-A

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Background and Aim: Coarctation of aorta is a congenital cardiac malformation, characterized by the narrowing of the aorta extends from discrete isthmus obstruction to tubular hypoplasia of the entire aortic arch. Symptoms depends upon the severity of coarctation, timing of progression and adequacy of collateral circulation. Coarctation in infancy presented with congestive heart failure. We observed during infancy and early childhood presented as dilated cardiomyopathy. Our report highlights unusual presentation of coarctation of aorta as DCM.
We presented that before label any patient as idiopathic dilated cardiomyopathy, we must exclude all possible specific causes of dilated cardiomyopathy because such type of cardiomyopathies are curable and very rewarding like our patient.
Method: it is an prospective observational study.
Results: A 3 year-old boy weighing 10kg admitted with the complaints of respiratory distress since 1month of age along with poor feeding, diaphoresis and not gaining weight. With this he was diagnosed as a case of DCM at his 3moth of age. But he lost follow up. He presented with tachycardia, tachypnea, Blood pressure was 110/60 mmHg in upper limb and 90/50 mmHg in lower limb. Femoral pulses were weaker with radio femoral delay. Bilateral basal crepitations and liver was palpable, 4 cm. Precordium examination revealed hyper dynamic precordium with shifted apex beat at left 5th intercostals space, Gallop rhythm was present and grade 2/6 systolic ejection murmur heard in the left 2nd intercostals area. CXR revealed cardiomegaly with pulmonary edema. Echocardiogram showed markedly dilated left ventricle, left atria with EF 22% with mild AR, mild TR. Severe coarctation of aorta with GDT of 74 mmHg. Co-A segment was 2.6 mm. Stabilizing the patient, Cardiac catheterization done and showed that pre Co-A pressure was 98/61/73 mmHg and post Co-A pressure was75/57/63 mmHg. Both Aortic valve and coronary artery were normal. So, DCM presumably resulting from the isolated coarctation of aorta. After successful Coarctoplasty pre –Co-A GDT was 88/55/73 mmHg and post GDT was 84/57/63 mmHg.LV function improved gradually with EF 32%.
Conclusions: With these, we presented that before we label any patient as idiopathic dilated cardiomyopathy, we must exclude all possible specific causes of dilated cardiomyopathy.

Keywords: Coarctation of Aorta, DCM

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PP-764

Use of devices during pediatric cardiac catheterization does not cause higher complication rates, but sheath size does

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Background and Aim: Complication rates in pediatric cardiac catheterization vary dependent on the intervention carried out. This study assesses if the of location of the intervention (intra- or extra-pericardial) and if the use of any device has an influence on complication rates.
Method: In this retrospective cohort study we compared the data of 1042 cardiac catheterizations in children with structural heart disease between 2010 and 2018. Major and minor complications were analyzed.
Results: 475 were diagnostic and 567 were interventional procedures. 97 (9,3%) complications were recorded in all procedures. There was no significant difference between intrapericardial interventions and extrapericardial interventions regarding complications, p=0,476). The use of devices showed a lower complication rate compared to interventions without device use (p=0,027). Larger sheath size related to body size is positively correlated with higher complication rates (OR 1,12 (1,00-1,25); p=0,044).
Conclusions: Neither the location of the intervention nor the use of devices were correlated with higher complication rates. A relatively large sheath size is a risk factor for complications.

Keywords: pediatric cardiac catheterization, complications, sheath size, devices

Neurodevelopment and psychosocial care

PP-765

Patients undergoing aortic ARCH surgery show different brain oxygen saturation patterns during surgery

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Background and Aim: During aortic arch surgery, the aortic clamp may influence cerebral perfusion and oxygenation. Little is known about the actual brain oxygenation (BO) during surgery and the resulting neurodevelopmental consequences.
We aim to analyze BO patterns measured by intraoperative near-infrared spectroscopy (NIRS) in a cohort of newborns undergoing aortic arch repair and evaluate their impact on the mid-term neurodevelopmental outcome.
Method: Newborn patients who underwent aortic arch surgery between 2017-2023 were included. Patients with hypoplastic left heart syndrome and with known neurological abnormalities were excluded. Arch hypoplasia (AH) was defined as z-score –2 at transverse arch measured by CT-scan. Cerebral oxygen saturation was measured intraoperatively using NIRS. Some somatic and brain damage biomarkers were analysed perioperatively. Neurodevelopmental outcome was assessed at 2 years using Bayley´s and Vineland tests.
Results: We included 33 patients (19 male), 20 with AH (transverse arch mean z-score –3). Patients had simple coarctation (CoA) (4), CoA+AH (12), CoA+ventricular septal defect (10), complex CoA (with other congenital heart disease) (3), and interrupted aortic arch (IAA) (4). Mean age at surgery was 12 days, mean weight was 3227gm. Eight patients were corrected via sternotomy and cardiopulmonary bypass (CPB), and 25 by thoracotomy. The average aortic clamp time was 12 min. In terms of intraoperative neuromonitorization, BO was lower in patients under CPB, especially those with IAA. No differences were found among groups of CoA or among those with and without AH. Patients without CPB and those with uncomplicated CoA had lower lactate levels after surgery. Fifteen patients had neurodevelopmental tests at 2 years, and only one experienced moderate compromise. While there were no statistically significant differences in Bayley´s score, patients with simple CoA performed better. However, due to our small sample size, we could not demonstrate important differences in neurodevelopment among patients with simple CoA and the other groups, or in those with HA, and also in those with or without CPB. Finally, we found no correlation between BO during surgery and neurodevelopmental outcome.
Conclusions: In our cohort, patients with IAA and those who required CPB have lower BO saturations intraoperatively, but this was not correlated with neurodevelopmental scores.

Keywords: Aortic arch, neurodevelopment, brain oxygenation

PP-766

Listen properly: What did chd patients and their relatives report about the onset of the corona pandemic in Germany? A nationwide analysis

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Background and Aim: Patients with CHD and their families are already impacted by the CHD in their daily lives due to the often severe congenital disease. When the Corona pandemic broke out in early 2020, those affected were confronted with a new threat of a global pandemic that was completely unknown to people in Europe, but especially in Germany. In order to gain a more detailed insight into the issues that chronically ill people and their relatives face in such an exceptional situation, a nationwide online survey was conducted to shed some light on the situation and to be better prepared in future comparable situations.
Method: The survey was conducted online by the German National Register for Congenital Heart Defects. Study participants were invited to participate by e-mail. A total of 3,558 patients were included in the statistical analyses (53% female).
Results: Survey participants were given the opportunity at the end of the questionnaire to describe their perceptions and experiences of the Corona pandemic. Of the 3,558 study participants, 565 (15.9%) provided such additional qualitative information (56.8% female). In comparison, this opportunity to express opinions was used primarily when a moderate or complex CHD was present [no qualitative responses: simple CHD (27%), moderate CHD (36.6%), complex CHD (29.1%), other CHD (7.3%); qualitative responses: simple CHD (19.6%), moderate CHD (34.9%),

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complex CHD (37.9%), other CHD (7.6%)]. The 565 study participants most frequently (30.4%) reported insufficient information about Corona in the context of CHD, with worries (24.1%) the second most commonly reported, closely followed by explicitly expressed fears related to Corona (23.2%).
Conclusions: Lack of information, concerns, and fears were the most frequently mentioned issues related to the Corona pandemic. On the one hand, this may be explained by the actual lack of information at the beginning of the Corona pandemic; on the other hand, a good and regular exchange between doctor and patient should prevent uncertainties, worries and fears, especially among chronically seriously ill patients, even in crisis situations. To do this better in the future, strategies for patient communication in times of crisis such as a global pandemic should be developed to increase patient safety.

Keywords: CHD, online survey, COVID-19, Corona pandemic in Germany

PP-767

What do chd patients and their relatives report after one year of corona pandemic in germany? A nationwide analysis of qualitative responses

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Background and Aim: Patients with CHD and their families are already impacted by the CHD in their daily lives. Even one year after the start of the Corona pandemic, those affected faced major challenges from the global pandemic. In order to gain a more detailed insight into the issues that chronically ill people face in such an exceptional situation, a nationwide online survey was conducted to shed some light on the situation and to be better prepared in future comparable situations.
Method: The survey was conducted online by the German National Register for Congenital Heart Defects (NRCHD). Study participants were invited to participate by e-mail. A total of 3,179 patients were included in the statistical analyses (52.6% female).
Results: Participants were given the opportunity at the end of the questionnaire to describe their perceptions and experiences of the Corona pandemic. Of the 3,179 study participants, 615 (19.3%) provided such additional qualitative information (51.4% female). In comparison, this opportunity to express opinions was used primarily when a moderate or complex CHD was present
[no qualitative responses: simple CHD (14.4%), moderate CHD (40.1%), complex CHD (36.5%), other CHD (9.1%); qualitative responses: simple CHD (14.6%), moderate CHD (35.3%), complex CHD (43.1%), other CHD (7%)]. Most frequently (26%), the 615 study participants reported insufficient information about Corona in relation to CHD, vaccinations (24.1%) were the second most frequently mentioned topic, closely followed by the topic of the often perceived insufficient prioritisation of CHD patients in everyday (medical) life during the Corona pandemic (23.4%).
Conclusions: Lack of information, vaccination, and prioritization were the most frequently mentioned issues related to the Corona pandemic. While concerns and fears were the most frequently mentioned topics in a survey of NRCHD patients at the beginning of the Corona pandemic, this changed to protective measures and the handling of particularly vulnerable patients by the health system and society. Clear plans, rules and explanations should be a central part of future pandemic response. In this context, the provision and clear communication of protection options for particularly vulnerable persons such as the chronically ill still seems to be expandable in order to increase patient safety and maintain compliance.

Keywords: CHD, online survey, COVID-19, corona pandemic in Germany

PP-769

Psychiatric disorders in children and adolescents with fontan circulation in denmark

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Background and Aim: Long-term co-morbidities including neuropsychological co-morbidities are common in children with congenital heart disease. Miles et al found in a registry study, that children and adolescents born with a CHD have a greater risk for psychiatric co-morbidities including anxiety disorder, autism spectrum disorder, and attention hyperactivity disorder. As the registries only include patients with psychiatric diagnoses made by a psychiatrist working in the public service, the results may potentially only reflect the tip of the iceberg. Many children and adolescents with somatic diseases such as epilepsy and mental health difficulties do not receive sufficient help for their mental health problems. We do not know if patients with CHD and mental health problems receive sufficient help.
Method: This is a questionnaire study of psychiatric comorbidities and received help in children and adolescents (age 5-17y) with complex CHD (Fontan circulation). Children, adolescents, and their families answer the Development and Well-Being Assessment (DAWBA) questionnaire and a questionnaire about receiving help at home. The questionnaires are then rated by three independent raters and controlled by two specialists in children and adolescent psychiatry and a consensus psychiatric diagnosis is made. The results will be compared to the Danish background population.
We will evaluate the mental help aid that the children and adolescents with both CHD and a psychiatric diagnosis have received.
Results: Results are pending. 22 of 65 Fontan-operated patients have given informed consent to participate in the study so far. Of these 22 participants, 15 have answered the psychiatric questionnaire. A psychiatric diagnosis was found in 9 of the 15 participants who answered the psychiatric questionnaire.
Conclusions: Pending on the results this study will answer the question if children and adolescents with a complex CHD have a larger burden of mental health problems than the background

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population and if children and adolescents with both a CHD and a mental health problem are receiving adequate mental help aid.

Keywords: Congenital Heart Defect, children, adolescents, mental health, psychiatry disorders, screening

PP-770

Emotion regulation strategies in youngsters (8-17 years) with a congenital heart disease (CHD) in comparison to healthy controls

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Background and Aim: Because of their medical vulnerability, youngsters with a congenital heart disease (CHD) may experience more overwhelming emotions while growing up than their healthy peers. To date it remains unclear whether these youngsters differ from their healthy peers with regard to the strategies they use to regulate their emotions. This study examines whether youngsters with CHD use more maladaptive and less adaptive emotion regulation strategies (ERS) compared with healthy controls.
Method: A sample of 8 to 17-year-old youngsters who were born with a CHD (percutaneous and/or invasive surgically corrected) was recruited (N = 203; 54.2% boys and 45.8% girls) and matched with healthy controls (N = 229; 52.4% boys and 47.6% girls) with regard to their age, gender and level of education. All participants completed self-report questionnaires about the use of emotion regulation strategies (FEEL-KJ). If the youngsters did not complete their self-report FEEL-KJ, the parents' answers on the parent report FEEL-KJ were consulted.
Results: In line with the hypothesis, a multivariate difference between both groups was found on the use of Maladaptive ERS ($p \leq .05$). Youngsters with CHD (M = 16.53, SD = 4.92) reported more us of 'Self-Devaluation' compared with the control sample (M = 15.43, SD = 5.16; p =.03). In contrast with the hypothesis, no multivariate difference was found between both groups on the use of Adaptive (p =.30) or External ERS (p =.53). The other four Maladaptive ERS (Giving Up, Withdrawal, Rumination and Aggressive Actions) showed no difference between these two groups. Results also show that both groups did not differ in the use of any of the Adaptive ERS (Problem Solving, Distraction, Forgetting, Acceptance, Humor Enhancement, Cognitive Problem Solving and Revaluation). The remaining three strategies (Social Support, Expression and Emotional Control) were also not significantly different in both groups.
Conclusions: Youth with CHD differ from their healthy peers with regard to their increased use of self-devaluation as ERS. Future research should further unravel emotional processes in youth with CHD. Pediatric patients may also benefit from emotion regulation training in order to strengthen this sample's wellbeing and to cognitive restructure these self-devaluative thoughts.

Keywords: emotion regulation, youngsters, CHD, matched healthy controls

PP-771

Pediatric post-pump chorea: Clinical features and outcomes

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Background and Aim: Post-pump chorea is a rare and poorly described complication of heart surgery, particularly with regard to its therapeutic management and evolution.
Method: Descriptive monocentric retrospective study including all children with post-pump chorea over a 10 year period (2014-2023). Clinical presentation, radiological findings, and outcomes were analyzed based on clinical, biological, and cerebral imaging data.
Results: Over 10 years, 7059 pediatric cardiac surgeries with cardiopulmonary bypass (CBP) were performed at Necker hospital, and 11 patients experienced post-pump chorea (0.15%), including 5 boys (45%). Median age at the diagnosis of post-pump chorea was 5.7 years [0.45-9.9]. One patient was premature, and none had genetic syndroms. Median BMI was low (14.8 [11.9-17.4]). Six patients had cyanotic heart disease. Six patients (55%) had previously undergone surgery with CBP. Median CBP duration and length of stay in the intensive care unit were typical, at 132 minutes [64-362] and 6 days [1-186], respectively. The interval between surgery and symptom onset was 20 days =[4-64], with a median duration of 44 days [3-181]. Abnormal movements primarily affected the face (100%) and were often bilateral (82%), involving the upper limbs (91%) and lower limbs (82%). Initial cerebral MRI did not show recent ischemic lesions. Six patients (55%) received medical treatment: tetrabenazine (n=2), intravenous immunoglobulins (n=1), corticosteroids (n=1), L-Dopa (n=1), cyamemazine (n=1). Chorea persisted beyond 6 months in only 1 out of 11 patients.
Conclusions: Post-pump chorea is extremely rare and can occur several months after cardiac surgery. Therefore, it is essential to recognize it. It does not appear to be directly related to surgical injury or pre- or peri-operative severity criteria. The prognosis is generally favorable within 6 months following diagnosis

Keywords: Chorea, neurological complications, cardiac surgery, cardiopulmonary bypass

PP-773

Educational experiences of children with congenital heart disease

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Background and Aim: A considerable body of international evidence exists demonstrating the consistently poor educational outcomes faced by children with Congenital Heart Disease (CHD). Children tend to lag further behind as they get older and these effects last longer term into outcomes in adulthood. It is therefore vitally important that adequate support is provided at an early stage in order to facilitate better educational outcomes for this cohort. The aim of this study was to identify the challenges facing teachers

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and parents of children with CHD in getting the best educational outcomes in school and to explore how these challenges might be overcome in order to improve the educational experience for this population.
Method: Co-productive work was conducted with three groups: (i) parents of children with CHD (ii) primary school teachers/ classroom assistants who had experience of teaching children with CHD and (iii) children and young people aged 11-17 with CHD. Focus groups/ one-to-one interviews were conducted with each group to explore the lived experience, views, and ideas and to establish the current level of need.
Results: Main challenges for teachers and classroom assistants of children with congenital heart defects included absenteeism as well as a lack of information around CHD and how it affects the individual child. Parents reported feeling anxious and scared around leaving the child at school and had concerns as to how their child would cope academically as well as in a social, emotional, and physical sense. Building a strong relationship and having frequent communication between the teacher/ parent/ child was considered key in alleviating anxiety and promoting a supportive environment.
Conclusions: Teachers of children with CHD require further support to include tailored information pertaining to the individual child updated on a regular basis. Parents require frequent communication with teachers as to how their child is coping. This information will be utilized in the next phase of the research to develop interventions to support teachers and families and to improve the overall educational experience for children and young people with CHD.

Keywords: congenital heart defect, congenital heart disease, educational challenges, educational outcomes, academic support

PP-775

Mothers, fathers, and the care work of children with congenital heart disease

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Background and Aim: Patients with congenital heart defects (CHD) are chronically ill and usually need lifelong medical and psychosocial care/support. This makes comprehensive information provision and communication necessary for the patients themselves, but also for their families. It has proven effective to involve not only the patients but also their closest relatives in the treatment process. Our study focuses on the role of the father in families with children with CHD. Among other things, we want to clarify the extent to which the care work of fathers and mothers differs and the burden is equally distributed or primarily one parent takes over these tasks.
Method: The survey was conducted online by the German National Register for Congenital Heart Defects (NRCHD). Study participants were invited to participate by e-mail. A total of 1,109 parents were included in the statistical analyses (57.9% mothers).
Results: Of the parents surveyed, 103 (9.3%) reported being single parents (mothers: 13.6%; fathers: 3.4%). The question about who mainly took care of the child when he or she was in the hospital was answered by the parents surveyed as follows: The mother (69.3%), the father (3.3%), both equal (26%), someone else/don't know (1.4%). The doctor visits and/or routine examinations in the hospital/heart center with the child are also mostly done by the mothers according to the parents: the mother (62.6%), the father (7.6%), both equal (29.5%), someone else/don't know (0.4%). According to the parents' assessment, the mother (51%) is most familiar with the child's CHD, previous medical interventions, medications, etc. (father 7.2%; both equal 41.5%; someone else/ don't know 0.3%).
Conclusions: The results clearly show that care work is largely done by mothers. It is a major and important task for the future to distribute this care between both parents and to involve the fathers more in the daily treatment routine, because care and support by both parents is important for good psychosocial development and an optimal medical outcome.

Keywords: CHD, online survey, parents, psychosocial care

PP-776

School enrollment of children with congenital heart disease in general

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Background and Aim: Outcomes for congenital heart disease have improved dramatically, with the majority reaching school age. We have previously found that extremely low birth weight children with congenital heart disease required learning support more frequently. However, the school enrollment among children with congenital heart disease in general is not clear. Objective To investigate the school enrollment among children with congenital heart disease in general.
Method: Subjects were cases born between 2010 and 2016, diagnosed with congenital heart disease, and followed through school age in our hospital. From the medical records, we investigated cardiac disease, treatment, and the school attendance.
Results: 453 cases of congenital heart disease were included in this study. Chromosomal abnormality was included in 59 patients (12.8%). Surgical treatment (surgery and catheterization) was performed in 273 patients (60.4%). The enrollment status was 351 (77.5%) in regular class, 100 (22.1%) in learning support (38 in special classes, 62 in special needs schools), and 2 were unknown. The frequency of requiring learning support was lower than the 47% previously reported for very low birth weight infants, but higher than the 3.1% reported for the general population in Japan. Of the cases requiring study support, 59 cases had chromosomal abnormalities and 41 had uncomplicated chromosomal abnormalities (uncomplicated group). All cases of chromosomal abnormalities required learning support. A history of NICU admission was 32 (78.0%) in the uncomplicated group and 103 (29.3%) in the regular class group, with a higher rate in the uncomplicated group ($P<0.05$). Simple heart disease was significantly less common in uncomplicated group than in the regular class group (48.8% vs. 72.6% $P<0.05$), and surgical treatment was performed at a higher rate in uncomplicated cases (70.7% vs. 29.3% $P<0.05$). The reasons for choosing learning support for the uncomplicated group included mental retardation (28 patients), hearing loss (4 patients, 9.8%), and developmental disability (10 patients, 24.4%).
Conclusions: More cases of congenital heart disease in general required learning support than in healthy population. Cases requiring learning support tended to have neonatal onset, complex cardiac disease, and surgical treatment compared to cases in the regular class.

Keywords: School Enrollment, Congenital Heart Disease

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Nursing and Allied Health Professionals

PP-777
Improving the nursing handover of pediatric cardiology patients who will be transferred from the pediatric intensive care unit to the pediatric unit

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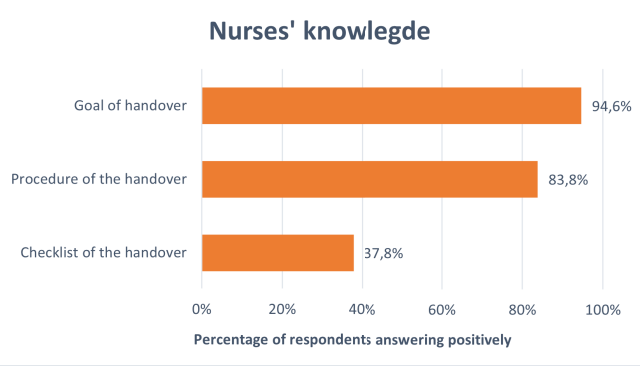
Background and Aim: Following cardiac surgery, pediatric patients are initially admitted to the pediatric intensive care unit (PICU) before being transferred to the pediatric unit (PU). A thorough transfer is essential to guarantee continuity and quality of care. Therefore, we improved the nursing handover from the PICU to the PU emphasizing psychosocial care aspects.
Method: In February 2022, nurses from the PICU and the PU started a quality improvement project involving the nursing handover. A preparatory face-to-face handover has been developed based on team experiences and literature. A pilot started including mainly pediatric cardiac surgery patients in June 2022. This pilot was evaluated within the working group and an improved handover was implemented in January 2023. The handover was recently evaluated using a survey with open-ended and closed questions asking for demographics and experiences with the improved handover. Close-end questions were analyzed descriptively. For the open-ended questions content analysis was used.
Results: Approximately three times a month, a transfer of a complex patient from the PICU to the PU occurs. Based on the pilot, the criteria and checklist were adjusted. The improved handover includes a daily inventory to determine which complex patients will be transferred in the upcoming days. Twice a week the handover is planned a day before the actual transfer, consisting an oral handover emphasizing psycho-social care aspects and bedside acquaintance.
Thirty-seven nurses (19%) from PICU (n=4) and PU (n=33) responded to the survey. Forty percent of these nurses had more than five years of experience in pediatric nursing. Of the respondents, twenty-five percent had experience with the improved handover and the subsequent transfer. Seventy-eight percent indicated a better transfer due to the improved handover. Figure 1 shows nurses' knowledge about the improved handover.
In the open answers, nurses explained that they are better prepared for the transfer with a positive effect on the confidence of the concerned families. The improved handover is experienced as more time-consuming, and timing of the transfer can be difficult to estimate.

Figure 1. Nurses' knowledge about the goal, procedure, and checklist of the improved handover.



Conclusions: A preparatory face-to-face nursing handover contributes to a better transfer of pediatric patients from PICU to PU. Regular evaluation provides us valuable insights for further improvement.

Keywords: transfer, nursing handover, pediatric, parents, cardiac surgery

PP-778
Little hearts at home

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Background and Aim: Healthcare globally is looking to digitisation to help tackle quadruple priorities: improving patient experience, improving overall population health, reducing costs, and improving the experience for health workers. Progress is inevitably restricted by limitations in resources and funding. Therefore, Alder Hey Innovation Centre and Alder Hey Heart Centre have developed a clinically validated paediatric RPM (remote patient monitoring) platform, Little Hearts at Home®™. (LHAH).
LHAH transforms existing post-operative pathways from a reactive approach to a proactive and preventative model. The platform aims to facilitate a safe early discharge, reduce emergency admissions, and reduce unnecessary readmissions.
Method: LHAH provides babies born with severe heart defects, such as single ventricle anatomy, with cross-organisational RPM, connecting patient, parents, community care providers, critical care teams, and clinical staff. Funded by NHS England, over the last 12 months, a team of paediatric cardiac specialists supported by an innovation team embedded within an NHS organisation, have used agile methodology and iterative testing to develop and implement LHAH successfully across the Northwest Congenital Heart Disease Network.
Results: We have successfully onboarded >200 community nurses across the North West Congenital Heart Disease Network and currently monitoring 31 active patients on the platform. For example, following a 6- months stay from birth at Alder Hey, patient X was able to go home to the Lake District (160-mile round trip) six weeks earlier than normal, in time for Christmas, with clinicians confident in monitoring their condition through LHAH platform. The early discharge has been estimated to have saved the Trust £101,000 in resources and allowed for another critically ill infant to utilise the bed space and clinical capacity reducing the waiting list.
Conclusions: The intent is using agile project management approach to establish LHAH as a paediatric focused remote patient monitoring platform, Alder Hey hosted platform, further developing and extending functionality into other specialties for Alder hey and expanding into other health care providers.

Keywords: Little, Hearts, at, Home

PP-779
Adolescents With Chd And Neurodevelopmental Disorders – How To Enable Them In Their Self-management

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Background and Aim: Congenital heart disease (CHD) is the most common birth defect, with a global prevalence of 8.2 per 1000 newborns. Also shown is a 44% higher prevalence of

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neurodevelopmental disorders in CHD compared to the general population. At present in our pediatric cardiac ward, we are confronted with adolescents with CHD and neurodevelopmental disorders. One of them is a 16-year-old male patient with hypoplastic left heart syndrome, failing Fontan and developmental amnesia. Mainly his short-term memory is affected. During the last two years, there have been many hospitalisations due to various factors, such as inadequate medication and symptom management. Due to his condition, it was difficult to maintain his autonomy and at the same time enable him to take part of his process of treatment.
Method: The first action was to identify the main factors limiting his self-management in an interprofessional roundtable discussion with him and his family. Furthermore, a literature search and a reference request to developmental pediatricians was made to identify adequate tools that can help him in his daily life.
Results: Adolescents with CHD and developmental amnesia need more guidance in their treatment plan. In order to determine these conditions, it is important to check their self-management during Nursing rounds and to define achievable goals together. Also we introduced a whiteboard to help him memorize and keeping track of medication, therapies and so on. This also included instructions and timestamps for the day. For medications we set alarms on his phone. It is very important to have regular conversations with the patient and his family to detect changes in an early stage.
Conclusions: An individualized treatment plan is needed for effective care of patients with developmental amnesia. This is optimized through nursing rounds and interprofessional case discussions to review the measures. Through the above interventions, the patient receives maximum support to participate in his treatment process. He has benefited most from the primary care. Through continuity of care, we were able to establish a routine that supported him in maintaining his autonomy and take a proactive part in his self-management.

Keywords: Congenital Heart Disease, Adolescents, Self-Management, Neurodevelopmental Disease, Developmental Amnesia

PP-780

Diagnostic yield of arrhythmogenic right ventricular cardiomyopathy in paediatric relatives of probands with negative or non-diagnostic autopsy

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Background and Aim: Cardiac evaluation at a specialist centre is indicated for first-degree relatives of individuals who have suffered a sudden cardiac death (SCD). For those in whom post-mortem does not reveal the cause of death, autopsy may find no abnormalities (Sudden Arrhythmic Death Syndrome - SADS) or findings of uncertain significance, termed 'non-diagnostic'. Autopsy findings are used to guide appropriate phenotypic screening of at-risk relatives. Recent studies have explored 'concealed cardiomyopathy' as a cause of autopsy-inconclusive SCD including Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC), an inherited cardiomyopathy characterised by fibrofatty replacement of the myocardium diagnosed using Task Force Criteria (TFC). This study aimed to describe the incidence of ARVC TFC abnormalities in childhood first-degree relatives of SCD individuals with SADS or non-diagnostic autopsies.
Method: A retrospective single tertiary referral centre cohort of children (≤18 years) with a first-degree relative with SCD without a pre-mortem cardiac diagnosis and negative or non-diagnostic autopsy from 2005 to 2023 was formed. Cardiac investigations were reviewed in relation to the ARVC Task Force Criteria (TFC) and data collected for patients meeting at least one ARVC TFC at any point during follow-up.
Results: Of 361 patients identified, 38 (10.5%) from 34 families (14.7%) fulfilled ≥1 criteria from the ARVC TFC. Of these, probands were mostly a parent (n=30, 88.2%) and male (n=23, 67.6%), with a mean age of 34.09 years (SD±10.14). The proportion of SADS and non-diagnostic autopsy in the proband was equal (n=17, 50%). True SADS autopsies were significantly more likely to have undergone cardiac specialist post-mortem [p=0.03]. The commonly fulfilled criteria was a positive Signal Averaged ECG in at least one vector (n=22, 57.9%). Six patients (15.7%) fulfilled criteria for a diagnosis of ARVC. Seven patients (18.4%) had documented Ventricular Tachycardia during follow-up.
Conclusions: This study shows one in ten paediatric relatives of SCD probands with negative or non-diagnostic post-mortems fulfilled ≥1 ARVC TFC criteria with a small number meeting diagnostic criteria for ARVC. This suggests these patients should be comprehensively screened for both inherited arrhythmia and heart muscle disease and highlights the importance of a specialist cardiac post-mortem and histology in cases of SCD to guide family screening.

Keywords: Sudden Cardiac Death, SADS, Family Screening, Arrhythmogenic Right Ventricular Cardiomyopathy

Results

ARVC TFC 2010 Categories		All Patients n (%)	SADS Post-Mortem n (%)	Non-diagnostic Post-Mortem n (%)	*p value*
	MRI	(*n*=30)	(*n*=13)	(*n*=17)	
I. Global or regional dysfunction or structural alterations	*Dilatation and Dysfunction*	3(10)	0	3(17.6)	0.074
	ECG	(*n*=38)	(*n*=19)	(*n*=19)	
III. Repolarisation abnormalities	*T-Wave Inversion in V1 and V2 at >14yo*	6 (15.8)	2 (10.5)	4 (21.1)	0.660
IV. Depolarisation/ Conduction abnormalities	*Positive SAECG*	22 (57.9)	12 (63.2)	10 (52.6)	0.511
	Terminal Activation Delay	4 (10.5)	3 (15.8)	1 (5.3)	0.604
	Ambulatory Monitoring	(*n*=35)	(*n*=18)	(*n*=17)	
V. Arrhythmias	*VT at any point during follow-up*	7 (20)	3 (16.6)	4 (23.5)	0.676
	>500 VEs in 24 hours	3 (8.5)	1 (5.5)	2 (11.7)	0.560

Table 1. Breakdown of ARVC Task Force Criteria fulfilled by paediatric patients and comparison between SADS and Non-Diagnostic post-mortem in probands

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PP-781

Family outcomes in families of children with congenital heart disease, down syndrome and both conditions from ecuador, spain and the united states

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Background and Aim: Families of children with chronic health conditions such as Congenital Heart Disease (CHD) and Down syndrome (DS) face many ongoing challenges. While some families have difficulty adapting to these challenges, others adapt well and some even thrive. The aim of this international study was to examine five family outcomes in families of children with CHD, DS, and both conditions from Ecuador, Spain and the United States (USA).
Method: Parents completed a packet of questionnaires that included the Family Outcomes Survey (FOS). This measure includes 5 scales designed to assess the following five family outcomes: 1) families understand their children's strengths, abilities and special needs, (2) families know their rights and advocate effectively for their children, (3) families help their children develop and learn, (4) families have support systems, and (5) families access desired services, programs, and activities in their communities). If a mean score for a FOS scale is 4 or higher, the indicator has been met. Clinicians and group leaders of support groups and foundations in Ecuador, Spain and USA shared an Invitation to Participate with eligible families. The survey was available in Spanish and English.
Results: 560 parents completed the packet of questionnaires (141 Ecuador, 273 Spain, and 146 USA). One-way analysis of variance F tests were used to compare means for the 5 FOS scales across the 9 groups and there were significant differences for all 5 FOS scales. Mean scores were lowest on the FOS scale designed to assess a family's ability to know their rights and advocate for their child (mean scores were lower than 4 for 6 of the 9 groups). Four groups (Ecuador DS, Ecuador Both, USA DS, USA Both) had mean scores lower than 4 for 2 FOS scales.
Conclusions: Adapting to the ongoing challenges associated with living with CHD and DS can be difficult for families, especially if the affected child has both conditions or the condition affects the individual's cognitive ability. However, some families are able to adapt successfully. More research is needed to understand how social determinants of health, such as where a family lives, can influence family outcomes.

Keywords: Congenital Heart Disease, Down Syndrome, Family

PP-782

Protocol For The Management Of The Safeguard Device In Nursing Units: Enhancing Patient Safety And Care Quality

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Background and Aim: The SafeGuard device is a pressure-assisted tool designed to facilitate the achievement and maintenance of haemostasis in femoral vascular accesses and to reduce compression time. The lack of knowledge of this device among nursing staff in post-anaesthetic recovery rooms, intensive care units, and wards can compromise the quality of care and patient safety. The primary objective was to equip nursing staff in various units with the necessary knowledge for efficient management of SafeGuard, as well as to act effectively when complications may arise.
Method: A protocol for the management of SafeGuard was designed, disseminated through posters in key areas of care (Anaesthetic Resuscitation, Post-Anaesthetic Resuscitation Unit, Paediatric Intensive Care Unit, and Ward) and complemented with informative talks. Effectiveness was assessed through a satisfaction survey.
Results: After the implementation of the protocol, a significant reduction in the number of calls to the haemodynamic nursing staff was observed. Additionally, the surveyed staff reported a decrease in anxiety levels when dealing with incidents involving the device. An increase in the correct autonomous intervention by nursing staff in response to incidents was recorded.
Conclusions: The introduction of a specific protocol for the management of SafeGuard in nursing units is an effective strategy to improve patient safety and the quality of care. Adequate knowledge of the device by the nursing staff reduces dependence on the haemodynamic team and decreases anxiety related to its use. These results underline the importance of specific training for staff in device management.

Keywords: SafeGuard Device, Haemostasis, Nursing Training, Patient Safety, Care Quality, Vascular Access

PP-783

The importance of pediatric palliative care on a cardiac intensive care unit – a case reporT

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Background and Aim: Roughly one percent of children are born with congenital heart disease (CHD). 25% of cases in the USA are categorized as critical CHD. These need heart surgery and care in the cardiac intensive care unit (CICU). This exposes the children to heightened risks of morbidity and mortality induced by multiple surgical interventions and extended CICU stays. We aimed to evaluate the psychological burden of parents from children with CHD and the supportive role of Pediatric Palliative Care Team by literature review.
Method: The results are based on a combination of literature review and direct observations of a three-month-old girl undergoing a five-month CICU stay.
Results: The technologically advanced CICU environment can be unfamiliar and daunting for parents, leading to shifts in parental roles and experiences of separation. This elevates the susceptibility to mental health disorders such as depression, anxiety, and acute stress disorders. Literature indicates a significant correlation between the severity of parental depression symptoms and the complexity of surgical procedures in their offspring. Maternal depression is associated with diminished bonding between mother and child. Prolonged CICU stays and repeated operations amplify the risk of anxiety, with some anxiety symptoms persisting over years. The critical nature of CHD, coupled with the challenges faced by parents, diminishes the overall quality of life for the entire family. Parents exhibit elevated stress levels, driven by uncertainties and concerns about the child's health. During the five-month CICU stay of our patient we identified an exhausted and anxious family due to the uncertainties and the complications they have already occurred.

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Conclusions: One proposed strategy to alleviate parental stress and improve the compromised quality of life in families with children with a critical CHD is the integration of the pediatric palliative care (PPC) team. Beyond managing patient symptoms, PPC teams offer substantial support to families by addressing the psychosocial and spiritual needs while ensuring continuity across various teams and wards. To enhance the early integration of PPC in children with CHD, the development of a comprehensive conceptual framework is recommended.

Keywords: pediatric palliative care, cardiac intensive care unit, CICU, PPC, psychological burden in parents

PP-784

Are infants with congenital heart disease (CHD) on parenteral nutrition at greater risk of intestinal failure associated liver disease (IFALD)?

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Background and Aim: Parenteral nutrition (PN) is commonly provided to infants with congenital heart disease (CHD) due to feeding delays associated with clinical status. IFALD is well-documented with liver cholestasis affecting up to 15.7% of all infants with short-term PN (≤1month), 60.9% in long-term PN (≥2months). CHD can be associated with liver dysfunction which could hinder hepatocytes from secreting bile, resulting in cholestasis. The aim was to see if there was a specific association with IFALD in infants with CHD whilst on PN.
Method: A retrospective review was conducted looking at patients <1 year of age with CHD on PN for >14 days between 2017-2023. The enteral feeding status was noted and baseline and peak bilirubin levels, with percentage of conjugated bilirubin were reviewed.
Results: 46 episodes of PN were noted. 59% of PN were in the neonatal period (≤28 days), of this 96% had a bilirubin peak within 14 days of PN, these were not deemed as IFALD, but likely neonatal jaundice. 4% (n=1) had a bilirubin peak at day 35 of PN and high percentage conjugated bilirubin, reflecting a case of cholestasis. 41% of PN episodes were in infants (>28 days). In this cohort 84% had no rise in bilirubin against baseline or remained in reference range. 16% (n=3) had a bilirubin peak after 14 days, 2 patients had no conjugated bilirubin screen, whilst one had a high percentage of conjugated bilirubin. In 82% of episodes enteral feeds were started within 14 days of starting PN.
Conclusions: There was a high frequency of possible neonatal jaundice in the cohort making definite conclusions hard to determine. Discounting these cases, only 4% of PN episodes lasting >14 days developed elevated total bilirubin and/or high percentage of conjugated bilirubin suggesting a possible link to IFALD. Compared to the frequency of PN cholestasis in the literature (15.7-60.9%) this is low, suggesting CHD may not be associated with a greater risk of IFLAD. An additional consideration is a high level of patients commenced on enteral feeds within the first 14 days of PN, supporting evidence that enteral feeding is a protective factor against IFALD.

Keywords: Congenital Heart Disease (CHD), Parenteral Nutrition (PN), Intestinal Failure Associated Liver Disease (IFALD)

Paediatric Cardiovascular Intensive Care

PP-785

Newborn cardiovascular recovery from birth assessed by pulse oximeter screening

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Background and Aim: Pulse oximeter (PO) screening is used to monitor arterial blood saturation. The PO derived peripheral perfusion index (PPI) is computed by a ratio of the pulsatile and the non-pulsatile part of the returning light thus providing information on hemodynamic stability. We assess whether the PO screening could give clinicians relevant information on the wellbeing of the baby in addition to screening the newborns for severe congenital heart defects (CHD).
Method: We report data of 697 infants born in Oulu university hospital between October 2nd and December 2nd in the year 2006. PO measurement (Masimo SET Radical) was performed before delivery room discharge, (mean 102.4 (SD 29.1) min after birth). Information on saturation (SaO2 %) and PPI was collected.
Results: Saturation measurement was obtained from 637 newborns, and PPI information from 72% of the newborns. The mean SaO2 was 98.6 % in 55 (9%) preterm, 98,3% in 547 term, and 97.7% in 35 (5%) post term born infants. The PPI values varied between 0.26 and 7.13 with the mean of 1.83 (1.04 SD). Female (49%) and male infants had similar mean SaO2% values (98.4 % vs. 98.3 %), but their mean PPI results differed significantly (1.66 vs. 1.96, p = 0.002). The infants that were later transferred to the NICU, had lower SaO2 % values (97.2 vs 98.4, p=0.048), but there was no difference in their PPI-values (1.91 (0.90) vs 1.80 (1.04), p=0.69). None of the study participants had severe CHD. Echocardiography was performed on 33 (5%) newborns. There were nine infants with ventricular septal defects and four with patent ductus arteriosus, with no abnormality on SaO2% or PPI values.
Conclusions: We investigated neonatal cardiovascular recovery using PPI and SaO2% values after childbirth on the neonates referred to mother and baby unit. The infants that needed care at NICU during the first days of life, had lower saturation values after birth. As a novel finding, this study suggests that when using the PPI as an indicator of the hemodynamic stability, the cardiovascular recovery on the male infants is superior (p=0.002).

Keywords: Saturation, Peripheral perfusion index, Hemodynamics,

PP-786

Assessment of psychosocial problems and needs of pediatric patients on a ventricular assist device and their impact on care, including the families

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Background and Aim: There are currently fewer than 20 pediatric patients in Germany depending on an extracorporeal VAD. For months to years, they wait for transplantation or recovery. Due to the increased complication rate associated with such a VAD, the patients must remain in the clinical environment for the entire time. For the families, it´s associated with drastic cuts in family life and high psychosocial stress levels. This results in special requirements for the responsible nursing staff. The aim is to assess the psychosocial problems and needs of parents and children who spend a long time in a clinical setting with a VAD. The results will be compared with the nursing standards of the affected wards, which will be evaluated and dedicated care plans will adapted if necessary. In this way, future patients and families should be better supported and psychosocial stress can be counteracted at an early stage.
Method: In order to improve our nursing standards, we plan a qualitative study in which patients, who have been dependent on a VAD, and their parents will be interviewed about the challenges during their time in hospital. We will include all 15 patients cared for during the last 4 years in our tertiary care center. Pediatric patients should be only interviewed from the age of 8 years or older.
Results: We hope that the results based on empirical values and the planned qualified survey will be able to identify the needs of patients and their families. In addition, we will analyze the relationship to interpersonal interactions with nursing care and the organization of everyday life in the clinical environment as to medical support.
Conclusions: The evaluation of the questionnaires should reveal potential psychosocial problems and needs that differ from those of other patients with shorter inpatient stays; these patients require a different type of nursing care from the staff in everyday clinical practice. By sensitizing the nursing staff in caring for VAD patients and the families concerned, excessive stress and long-term trauma can possibly be avoided. As this project is still in preparation stage, we look forward to exchanging ideas with other centers and invite potential participants.

Keywords: Pediatric patients, extracorporeal ventricular assist device (VAD), psychosocial burdens, long-term hospitalization, qualitative adaptation of medical care.

PP-787

Unclear severe mitral regurgitation in the course of systemic infammatory response, as a result of mistreatment?

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Background and Aim: Mitral regurgitation in children is most commonly a consequence of defective atrioventricular valve formation during the early embriological development. Less frequently it is caused by infection, but also various rheumatic heart disease and genetic connective tissue disorders. Rarely, MR can be a consequence of a systemic immune mediated inflammatory process.
Method: Case presentation.
Results: We report the case of a 6-year-old girl who was transferred to our center due to cardiac decompensation of unidentified etiology. Her past medical and family history was unremarkable. She first presented to a pediatric emergency department in a neighboring country one week earlier with signs of respiratory insufficiency with pleural effusion following one month of cough and malaise. Despite thoracocentesis and mechanical ventilation, her respiratory function continued to deteriorate, and she started to develop the signs of multiorgan failure, so continuous inotropic and vasoactive support was introduced. At the admission to our center echocardiography was performed showing severe regurgitation of all heart valves with preserved ejection fraction. Progressive de-escalation of inotropes was initiated along with afterload reduction therapy, which resulted in gradual right heart and cardiac output recovery. Due to excessive water retention and oliguria, CRRT was performed for 2 days. Magnetic resonance of the heart showed severe dilatation of all cavities due to regurgitation, mostly mitral. Lung MSCT showed ground glass opacity. She had a non-nephrotic range proteinuria and severe hypocomplementemia (with slightly increased CRP. ANCA, ANA and ASO titre were negative. She received 2g/kg of methylprednisolone and 10g of IVIG, which lead to recovery of all valves, except mitral, while 3D echocardiography revealed possible anterior mitral chordae rupture with prolapse. After 2 months of hospital treatment she was finally discharged with grade II-III mitral regurgitation, necessitating pharmacological stabilisation. During the follow further mitral regurgitation was noted up to grade I, with no signs of immune mediated diseases.
Conclusions: While systemic inflammatory response s in children commonly occurs with various etiology, the possible affection of heart valves during its course is unpresented. Regardless of etiology, our case demonstrates that the improvement of hemodynamic stability during intensive cardiologic management leads to permanent amelioration of heart associated symptoms.

Keywords: Mitral regurgitation, Heart Valve, Systemic inflammatory response

PP-789

Stroke volume measurements by echocardiography in children

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Background and Aim: Stroke volume (SV) and cardiac output (CO) monitoring is a cornerstone of hemodynamic assessment. Physicians are likely insufficiently capable of recognizing a low CO when using clinical examinations. An echocardiography assessment in the apical/parasternal view is recommended position for CO and cardiac index (CI) measurement. The apical/parasternal position is not always readily obtainable for measurement. To our knowledge, there are no data comparing transthoracic echocardiography parasternal (TTE-P) and transthoracic echocardiography jugular (TTE-J) SV measurements in pediatric patients. This study compared TTE-P and TTE-J SV measurements in children. We hypothesized that the results of the TTE-J measurement would not differ from the TTE-P measurements.
Method: A single-center prospective observational study was conducted in 37 spontaneously ventilating, hemodynamically stable children (7.55±4.65yrs, girls made up 57% of the study patients) who were admitted to the university hospital. None of the enrolled patients had aortic valve disease. A pediatric cardiologist obtained 3 sets of velocity time integral (VTI) within a period of 3 to 5 minutes in both apical and jugular views. Aortic valve diameter (AVD) was measured in the parasternal long-axis view. A

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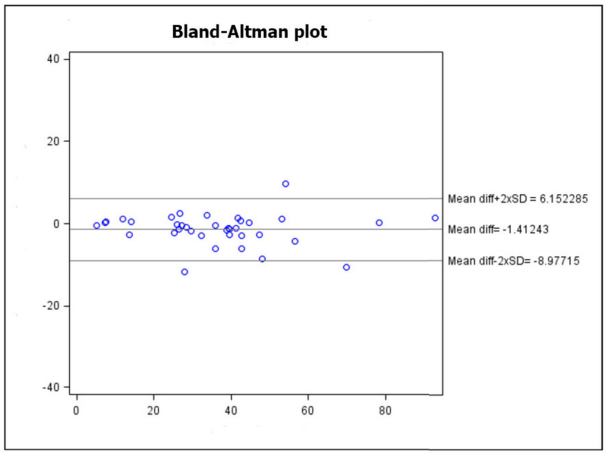
research assistant recorded VTI, SV, and AVD measurements while the investigator was blinded to these results. The SV of the left ventricle was calculated as a product of aortic valve cross-sectional area multiplied by VTI. For reported results comparing paired SV measurements were obtained by Bland-Altman analysis (bias and limits of agreement) and percent difference analyses was calculated.
Results: Both approaches were applicable in 94.7% of patients. The intraobserver variability TTE-J and TTE-P were 6.5± 4.3% and 7.8 ± 5.8%, respectively. The measured SV using TTE-J and TTE-P were 38.91 ± 19.98mL, and 37.58 ± 19.81mL, respectively. The bias (and limits of agreement) for SV comparing TTE-P to TTE-J was −1.41 (−8.98–6.15). The percentage error in SV measurements with TTE-P was calculated 21% relative to TTE-J.
Conclusions: Our findings demonstrate that the methods are interchangeable and might be used for CO assessment in children. Both methods have low level of intraobserver variability in children. Further studies will be required to determine the accuracy under changing hemodynamic conditions.

Keywords: stroke volume, cardiac output, parasternal, jugular, echocardiography



Bland-Altman plot for SV between TTE-J and TTE-P.

PP-790
Ultrasound for diaphragmatic dysfunction in postoperative cardiac children

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Background and Aim: Diaphragmatic Dysfunction is a common cause of failed extubation and prolonged mechanical ventilation after pediatric cardiac surgery in up to 14%. This study aims to evaluate the role of critical care bedside Ultrasound performed by intensivist to diagnose diaphragmatic dysfunction and the need for plication after pediatric cardiac surgery.
Method: Retrospective cohort study on prospectively collected data for post-operative children admitted to PCICU during 2020. Diaphragmatic dysfunction was suspected based on difficulties in weaning from positive pressure ventilation or Chest X-Ray findings. Ultrasound studies were performed by PCICU intensivist and confirmed by qualified radiologist.
Results: Out of 344 post-operative patients, 32 needed diaphragm ultrasound for suspected dysfunction. Ultrasound confirmed diaphragmatic dysfunction in17/32 (53%) patients with an average age and weight of (10.8±3.8) months and (6±1) Kg respectively. The incidence rate of diaphragmatic dysfunction was (4.9%) in relation to the whole population. Diaphragmatic plication was needed in 9/17 cases (53%), with rate of 2.6% in post-operative cardiac children. Mean plication day was (15.1±1.3) after surgery. All patients who underwent plication were under 4 months of age. Post plication they were discharged with mean Pediatric CICU and hospital stay of (19±3.5) and (42±8) days respectively.
Conclusions: Critical care ultrasound assessment of diaphragmatic movement is a useful and practical bedside tool that can be performed by a trained pediatric (CICU) intensivist. It may help in early detection and management of diaphragmatic dysfunction post pediatric cardiac surgery which may have potential positive effect on morbidity and outcome.

Keywords: Ultrasound, diaphragm, paediatric cardiac surgery, phrenic nerve injury, mechanical ventilation

Algorithm for ultrasound-enhanced management of diaphragmatic dysfunction (DD) after paediatric cardiac surgery.

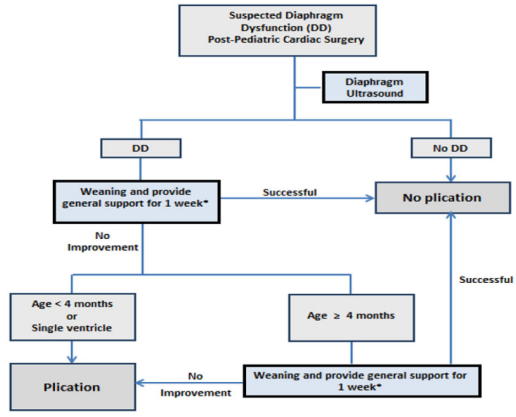

Figure 2.
Algorithm for ultrasound-enhanced management of diaphragmatic dysfunction (DD) after paediatric cardiac surgery. *Diaphragm ultrasound will be repeated at the end of the weekly weaning trial from positive pressure respiratory support to confirm DD.

*Algorithm for ultrasound-enhanced management of diaphragmatic dysfunction (DD) after paediatric cardiac surgery. *Diaphragm ultrasound will be repeated at the end of the weekly weaning trial from positive pressure respiratory support to confirm DD.*

PP-791
Ultrasound-guided post-pyloric feeding tube insertion in peri-operative cardiac infants

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Background and Aim: Delivery of enteral nutrition in critical infants post-paediatric cardiac surgery is sometimes hampered, necessitating direct feeding into the small intestine. This study is highlighting the role of ultrasound-guided post-pyloric feeding tube insertion performed by the paediatric cardiac ICU intensivist in critically ill infants.

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Method: We carried out a prospective pilot observational experimental study in peri-operative cardiac infants with feeding intolerance between 2019 and 2021. Feeding tube insertion depends on a combination of ultrasound and gastric insufflation with air-saline mixture. Insertion was confirmed by bedside abdominal X-ray.
Results: Out of 500 peri-operative cardiac infants, 15 needed post-pyloric feeding tube insertion in median 15 postoperative day. All were under 6 months of age with average weight of 3 ± 0.2 kg. Median Risk Adjustment for Congenital Heart Surgery Categories was 4. Median insertion time was 15 minutes. No complications have been reported. First pass success rate was 87%, while a second successful insertion attempt was needed in 2 cases (13%). Target daily calorie intake was achieved within average of 3.5 ± 0.4 days. Mean post pyloric feeding tube stay was 20 ± 3 days. Out of 15 infants, 3 patients died, 1 patient needed gastrostomy tube, and 11 patients were discharged home on oral feeds.
Conclusions: Ultrasound-guided post-pyloric feeding tube insertion using gastric insufflation with air-saline mixture in peri-operative cardiac infants with feeding intolerance is a useful and practical bedside tool, and it can be performed by a trained paediatric cardiac ICU intensivist. It may have potential positive effects on morbidity and outcome.

Keywords: Ultrasound, post-pyloric feeding tube, paediatric cardiac surgery

Pictures illustrating ultrasound (US) probe positionand orientation to visualise the insertion of feeding tube within the pylorusin infant with feeding intolerance peri-operatively

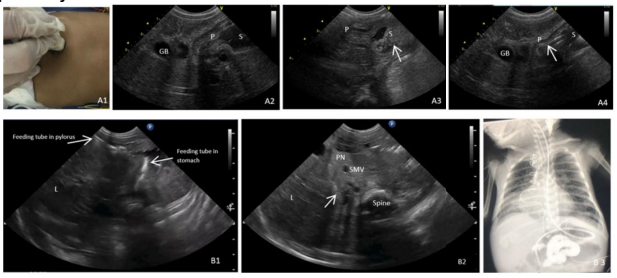

Figure 2. A1-4: Pictures illustrating ultrasound (US) probe positionand orientation to visualise the insertion of feeding tube within the pylorusin infant with feeding intolerance peri-operatively; A1: The US probe is placedin the longitudinal plain toward left shoulder, 2 cm below the costal marginright to sibxiphoid area; A2: US visualisation of the pylorus; A3: US visualisationof dynamic fogging in stomach using air-saline mixture (arrow); A4: USvisualisation of the feeding tube passirg the pylorus as a double line artefact (arrow). GB indicates gallbladder; P: pylorus; S: stomach. B1-3 Picturesillustrating (US and X-ray) to visualise the post-pyloric feeding tube ninfant with feeding intolerance peri-operatively. B1: feeding tube passingpylorus in trans-liver US window; B2: US visualisation of post-pylorus tubeinsertion in distal second segment of duodenum (arrow); C: Abdominal X-ray withcontrast confirms post-pyloric feeding tube in distal second segment ofduodenum without intestinal perforation. GB indicates gallbladder; L: liver; PN: pancreas; SMV: superior mesenteric vein.

PP-792
Bedside ultrasonography screening for congenital renal anomalies in children with congenital heart diseases undergoing cardiac repair

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Background and Aim: Ultrasound (US) assessment of renal anomalies in children requiring pediatric cardiac surgery is not a standard practice. This study is highlighting the role of bedside US performed by intensivist to detect occult renal anomalies associated with congenital heart disease (CHD).
Method: A cross sectional study for 100 consecutive children with CHD admitted to Pediatric Cardiac Intensive Care Unit (PCICU) in 2019. US of kidneys screening was performed by trained pediatric cardiac intensivists to ascertain the presence of both kidneys in renal fossae without gross anomalies and to investigate if early detection of occult kidney anomaly would have any impact on outcome.
Results: After screening of 100 consecutive children with CHD with renal US, we identified in 94 cases (94%) normal right and left kidney in the standard sonographer shape within the renal fossae. In 6 cases further investigation revealed ectopic kidney in 3 patients (50%), solitary functional kidney in 2 patients (33%) and bilateral grade IV hydronephrosis in one patient (17%). Urinary tract infection developed peri-operatively in 66% of the cases with kidney anomalies with statistical significance compared to patients with normal renal US (P: 0.0011).
No significant renal impairment was noted in these patients post-surgery. We observed no specific association between the type of renal anomaly and specific CHD.
Conclusions: Routine renal US in children with CHD demonstrated prevalence of associated congenital renal anomalies in 6% of children undergoing cardiac surgery. The presence of occult renal anomalies was associated with higher UTI risk. Performing routine renal US as a standard practice in children with CHD is justifiable

Keywords: Renal ultrasound, Congenital renal anomalies, Congenital heart disease

Renal US views

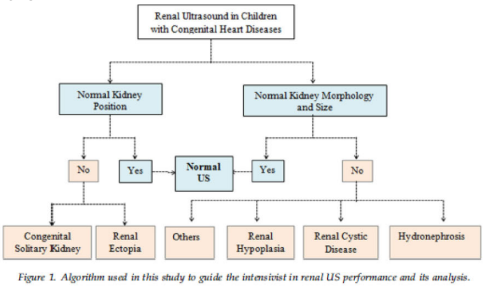

Figure 1. Algorithm used in this study to guide the intensivist in renal US performance and its analysis.

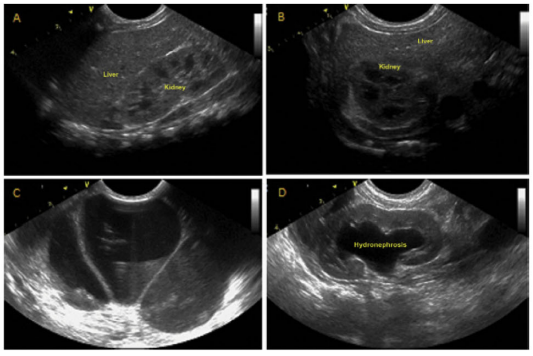

Figure 2. Renal US views of our cases: A) Standard sonographer shape of the right kidney in long-axis view obtained through the liver demonstrating echogenicity of the normal renal parenchyma, which is approximately equal to the echogenicity of the normal liver. The renal sinus is hyper echoic compared to the renal parenchyma. B) Transverse (short axis) view of the normal right kidney. C) Multi-cystic dysplastic kidney. The left kidney is totally replaced by cysts. D) Significant hydronephrosis (marked dilation of the calices and renal pelvis).

Figure 1. Algorithm used in this study to guide the intensivist in renal US performance and its analysis. Figure 2. Renal US views of our cases

PP-793
Impact of pre-operative intensive care on outcomes in patients with ventricular septal defects in an LMIC

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Background and Aim: Children with congenital heart disease (CHD) necessitate specialized and resource-intensive care and cardiac surgeries for better outcomes. It is more likely in a developing country due to delayed diagnosis and post-diagnosis neglect, followed by presentation frequently in compromised state. We evaluated impact of a recently established dedicated pre-operative Pediatric Cardiac Intensive Care Unit (PCICU) on outcomes of children with ventricular septal defects (VSD).
Method: This ambispective study spanned over a period of 5 years, encompassing retrospective (Nov 1, 2018 - Oct 5, 2021) and prospective (Oct 5, 2021 - Oct 31, 2023) data collection. Clinical profile, critical care needs and outcome of children with VSD presenting in critically ill state in the Pediatric emergency and subsequently transferred to this unit was compared before and after establishment of full-fledged critical care facility. Impact of starting a formal intensive care services was assessed on clinical characteristics, critical care needs, pre-operative and post-operative outcome.
Results: Out of 1098 patients with CHD admitted, 230 (25.9%) had VSD (M:F; 143:87). Median (IQR) age at diagnosis in pre-PCICU and post-PCICU were 143 (84;256) days and 120 (60;180) days respectively.Respiratory distress was the predominant presentation (79.5%). Among them, 64.7% were severe underweight, 32.1% were severely stunted, and 42.9% had microcephaly. Comparison of clinical profile of patients between these two periods is presented in the table
Conclusions: Admission to specialized PC-ICU for pre-op stabilization is associated with increased utilization of critical care interventions. This has the potential to lead to a more efficient utilization of resources and lowered hospital costs for children in need of congenital cardiac surgery.

Keywords: Ventricular septal defect, pediatric cardiac intensive care unit, congenital cardiac surgery

Table: Comparison of Clinical characteristics, Critical care needs and Outcomes of patients with Ventricular septal defects

Characteristics		Pre-PCICU (n=143)	Post-PCICU (n=87)	p-value
Gender	Male Female	88 55	55 32	0.79
Age (days) [Median (IQR)]		143 (84;256)	120 (60;180)	0.08
Age Groups	First Week	1	1	0.70
	Post 7-days Neonates	9	10	
	Post-neonatal Infants	113	67	
	Post-infancy	20	9	
Pre-operative NIRS, n (%)		107 (74.8%)	75 (86.2%)	0.66
Pre-operative NIRS duration among survivors (days) [Median (IQR)]		2 (1;15)	13 (6.5;18)	0.09
Need of Ventilation, n (%)		47 (32.8%)	36 (41.3%)	0.19
Pre-operative Ventilation Duration among survivors (days) [Median (IQR)]		11 (3;41)	12 (7.5;18.5)	0.000
Need of Inotropes, n (%)		48 (33.6%)	26 (29.8%)	0.56
Inotrope duration among survivors (days) [Median (IQR)]		4 (2;5)	3 (2;5.5)	0.82
Maximum Vasoactive Inotropic score [Median (IQR)]		20 (10;76)	50 (20;70)	0.08
Pre-Operative ICU Stay among pre-op survivors(days) [Median (IQR)]		22 (11;39)	27 (17.5;35.5)	0.09
Pre-Operative ICU Stay among those who underwent surgery (days) [Median (IQR)]		22.5 (15.5;26.2)	17 (11;30)	0.53
Pre-operative Outcome	Discharge	107 (74.8%)	69 (48.3%)	0.11
	Deaths, n (%)	20 (13.9%)	14 (16.1%)	
	LAMA, n (%)	14 (9.7%)	4 (4.6%)	
	Transfer for surgery among survivors	34 (23.7%)	25 (17.5%)	0.68
Post-operative Outcome among those went for surgery (n=59)	Discharge	27 (79.4%)	20 (80%)	0.68
	Death	3 (8.8%)	4 (16%)	
Total Hospital Stay who got operated and survived (days) [Median (IQR)]		45 (34.2;56.7)	48.5 (28.7;59)	0.8

Comparison of Clinical characteristics, Critical care needs and Outcomes of patients with Ventricular septal defects

PP-794

Evolution of nt-probnp before and after MIS-C diagnosis and treatment

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Background and Aim: In multisystem inflammatory syndrome in children (MIS-C or PIMS temporally associated with SARS-CoV-2), N-terminal pro-B-type natriuretic peptide (NT-proBNP) is used for positive and differential diagnosis but contrasting cut-offs have been suggested. Moreover, the usefulness of NT-proBNP to assess treatment efficacy is unclear. The aims of the present study were to compare NT-proBNP values according to the time of measurement and to associate NT-proBNP with clinical and biological inflammatory markers after treatment.
Method: A single-centre cohort observational study included patients with MIS-C, according to the World Health Organization definition, from May 2020 to April 2023. The timing and level of NT-proBNP samples available for each patient were collected retrospectively, compared, and correlated to the duration of fever and C-reactive protein (CRP) level.
Results: Thirty-seven children (19 (51%) males, median age (interquartile range) 8.8 (5.6-10.8) years, 14 (38%) with shock, 33 (89%) with a documented positive SARS-CoV-2 test) were included. Before diagnosis, NT-proBNP values were significantly higher at 6 days from first symptoms than at 3 days (32933 (7773-61592) versus 1994 (1291-4190) pg/mL respectively, p=0.031). From diagnosis and treatment, the NT-proBNP decrease at 2 days with the 50% cut-off had a 100% negative predictive value to exclude persistent fever only in children with high NT-proBNP at diagnosis > 11000 pg/mL (95% confidence interval (69-100%), p=0.036, n=16). In all children, median NT-proBNP at 3 days post-diagnosis was significantly lower than at diagnosis (2584 (1828-4088) pg/mL versus 8558 (1920-16836), p=0.028). The percentage of NT-proBNP and CRP decrease at 4 days correlated positively (-58% (-84%-+2%) versus -77% (-82%–56%) respectively, r=0.75, p=0.026, n=9).
Conclusions: NT-proBNP increased with time from first symptom to diagnosis suggesting different thresholds for MIS-C diagnosis depending on the time of illness. After diagnosis and treatment, a rapid decline in NT-proBNP was associated with an elevated NT-proBNP level at diagnosis > 11000 pg/mL, a resolution of fever within 48 hours, and a rapid decline in CRP.

Keywords: Brain natriuretic peptide, paediatric, heart failure, diagnostic delay, COVID-19, treatment resistance

PP-795

Pediatric delirium: An underestimated disorder after cardiac surgery on the cardiac intensive care unit (CICU)

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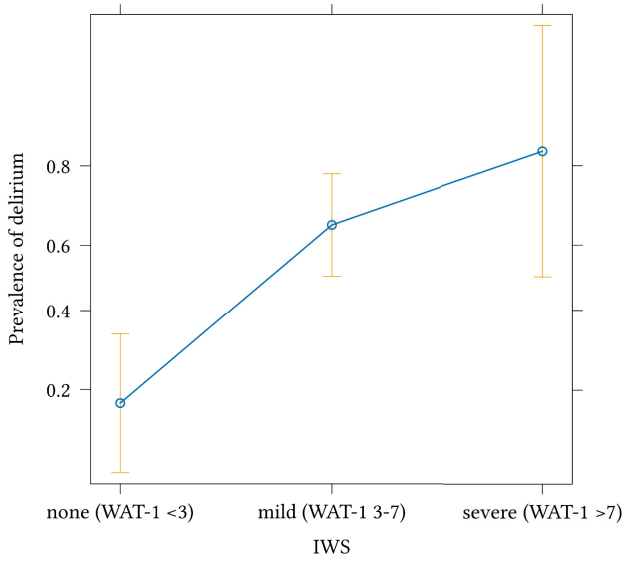
Background and Aim: Delirium is defined as an acute neuropsychological disorder, whose appearance is associated with various, known risk factors in the adult population that are also very common in pediatric patients, especially after cardiac surgery. Compared to older screening instruments, the Cornell assessment of pediatric delirium (CAPD) now allows screening of toddlers and even neonates, representing the vast majority of CICU patients. With evolving focus on neurodevelopmental outcome for children after cardiac surgery and the existing knowledge about neuropsychological impact of postoperative delirium in adults, prevalence and risk factors in children need to be evaluated to enable diagnosis and prevention, facilitate treatment options and improve neurodevelopmental outcome.
Method: The preliminary endpoint of this retrospective, single-center study was to determine the delirium prevalence using the CAPD and differentiating symptoms to the iatrogenic withdrawal syndrome (IWS) using the withdrawal Assessment Tool-1 (WAT-1). Screening was performed eight hourly on all postoperative CICU-patients over a period of six months. Multivariate logistic regression analysis of independent variables including severity of cardiac disease (RACHS-1), severity of IWS (mild: WAT-1: 3-7, severe: WAT-1 >7) cardiopulmonary-bypass-time, hypoxia (SaO2 <92% for consecutive 6 hours), metabolic derailment (blood glucose >250mg/dl) and low cardiac output (LCO) (serum lactate >2mmol/l) was performed.
Results: Delirium and IWS scoring were conducted in 74.7% of 150 patients. The overall prevalence of delirium was 34.6% (CAPD >9). IWS was present in 42.3% (WAT-1 > 3), whereas 33.3% having mild and 9.3% severe manifestation. IWS was the only significant independent variable for delirium with an Odds ratio (OR) of 9.3 (p<0.001) for mild and 29.8 (p<0.001) for severe manifestation. Hypoxia (OR 1,8), LCO (OR 1,6) and metabolic derailment (OR 1,2) seemed to be associated with delirium, but results were not significant. Surprisingly cardiopulmonary-bypass-time (OR 1.0) was not a significant predictor.
Conclusions: Pediatric delirium and IWS have a high prevalence in children after cardiac surgery and screening tools should be integrated into standard care. Withdrawal as a significant risk factor for delirium requires better sedation and weaning strategies to improve prevention of delirium. Overall, further research is needed on postoperative management and neurodevelopmental outcome for children after cardiac surgery who suffer from delirium.

Keywords: Pediatric delirium, Iatrogenic Withdrawal Syndrome, Neurodevelopmental Outcome, Pediatric Intensive Care Unit

Delirium prevalance and severity of Iatrogenic Withdrawln Syndrome (IWS)



Pediatric patients after cardiac surgery with mild IWS had a 9,1-fold higher risk (95%CI 3,2-28,9, p= < 0,001) and with severe IWS a 22,6-fold higher risk (95%CI 4,4-178,7, p= < 0,001) of developing a delirium.

PP-796

Postoperative mediastinitis in children: clinical features, outcomes and mortality risk factors

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Background and Aim: Mediastinitis is a rare complication of congenital heart surgery that increases mortality and morbidity. However very few studies focused on the description of mediastinitis and its outcomes in pediatric patients.
Objectives: To describe the clinical features and outcomes of post operative mediastinitis in children and identify risk factors for mortality.
Method: We retrospectively reviewed all children with mediastinitis over a period of 10 years (2013-2023). Mediastinitis was defined by a purulent discharge of the mediastinum requiring surgery or organisms cultured from discharge of the mediastinal area. We assessed clinical and microbiological features. The outcomes were mortality and ICU length of stay.
Results: The incidence of mediastinitis was 0.74% with 57 cases out of 7665 cardiopulmonary bypasses. Median age at surgery was 12 days with 67% patients younger than 3 months old and a median weight of 3.5kg. 60% had delayed sternal closure. Median delay between surgery and diagnosis was 8 days (range: 2-39). The most frequent germs were staphylococcus (46%), gram negative bacteria (35%) and fungi (9%). Median delay between surgical revision and diagnosis was 2 days (range: 0-55). All patients had surgical debridement: 46 (81%) in the operating room and 11 (19%) in the ICU. Median duration of antibiotic therapy was 44 days. Median duration of stay was 21 days (range: 10-113) in the ICU and 35 days (range: 11-188) until hospital discharge. Outcomes in the ICU included a median of 8 days of assisted ventilation and 7.5 days of inotropic support. 9 patients needed veno-arterial ECMO.
Mortality rate was 27% with 12 out of 15 deaths occurring in the ICU. Mortality risk factors were surgical revision in the ICU compared to the operating room (OR 4.9; CI95% 1.3 to 19.9), delayed sternal closure superior to 3 days (OR 5.0; IC 95% 1.3 to 16.5) and fungal mediastinitis (OR:14.9; IC95% 2.0 to 185.4).
Conclusions: Mediastinitis is a severe complication leading to high mortality and morbidity in children. Mortality risk factors are: surgical revision in the ICU, prolonged delayed sternal closure and fungus infection.

Keywords: mediastinitis, infection, cardiac ICU, cardiac surgery

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PP-797

Endothelial glycocalyx degradation and circulating endothelin-1 levels in children undergoing cardiac surgery with cardiopulmonary bypass

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Background and Aim: Pediatric cardiac surgery with cardiopulmonary bypass (CPB) induces a proinflammatory response and alterations in the microcirculation that are associated with postoperative complications. The mechanism responsible for endothelial injury during surgery and how it affects the perioperative outcomes is still unclear.The goal of this study is to characterize the inflammatory response and endothelial dysfunction after cardiac surgery with CPB and to correlate the degree of endothelial glycocalyx (EG) shedding, endothelin-1 (ET-1) levels and cytokine production with postoperative outcomes.

Method: Plasma and serum samples from 27 children (median age 4.5 years) undergoing cardiac surgery with CPB were obtained pre-operatively (T0), 30 min after CPB (T1), 6h after pediatric intensive care admission (T2), on postoperative day 1 (T3), and day 2 (T4). Syndecan-1 (SD-1), ET-1 and inflammatory cytokines (IL-6, IL-10, IL-8, and IL-1ra) were measured using ELISA/ Luminex kits. Eleven healthy age-matched children were used as control group.

Results: Preoperative levels of SD-1, ET-1, and cytokines were in the normal range in all children with the exception of children with cyanotic CHD who had higher SD-1 levels compared to the control group (Figure 1A). SD-1, ET-1 and cytokine levels increased significantly during surgery and peaked at T1 for both SD-1 and inflammatory cytokines and at T2 for ET-1 (Figure 1B). By T4, SD-1, ET-1 and cytokine levels had decreased to baseline levels except for children with cyanotic CHD who had persistent elevated SD-1 and ET-1 levels.

SD-1 and ET-1 levels were only weakly correlated to each other. They were however positively correlated with PICU length of stay and maximal vasoactive-inotropic score, while ET-1 levels were also correlated with duration of mechanical ventilation (Figure 1C).

Conclusions: Cardiac surgery with CPB caused endothelial injury and EG shedding that were associated with postoperative morbidity. Children with cyanotic CHD presented the highest levels of ET-1 and SD-1 suggesting a possible increased susceptibility for endothelial dysfunction in this group of CHD.

Keywords: cardiac surgery, cardiopulmonary bypass, endothelial injury, glycocalyx degradation, inflammation

Fig 1

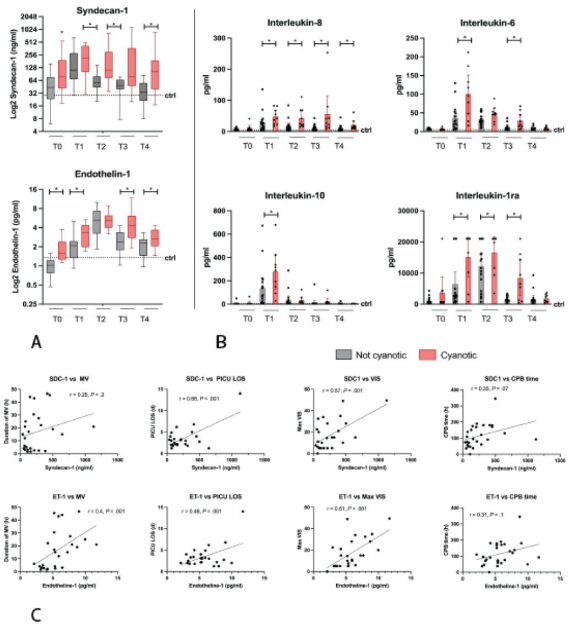

Figure 1. A): levels of Syndecan-1 and endothelin-1 in plasma from cyanotic (n=9) and non-cyanotic (n=18) patients at different timepoints. Pre-operatively (T0), 30 min after CPB (T1), 6h after pediatric intensive care admission (T2), on postoperative day 1 (T3), and day 2 (T4). Data are expressed in median with interquartile range. B): levels of inflammatory cytokines. Data are expressed as mean with SD. Data from control children are reported as a dotted line. C) Relation between syndecan-1 and endothelin-1 plasmatic levels and duration of mechanical ventilation, pediatric intensive care length of stay, maximal vasoactive-inotropic score, and duration of cardiopulmonary bypass

PP-798

Intrapericardial streptokinase: a penultimate solution in non purulent - fibrinous pericarditis

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Background and Aim: Post operative pericardial effusion, leading to the development of fibrinous pericarditis occurs in 9-14% of pediatric patients post cardiac surgery. Standard management includes anti-inflammatory drugs and may require insertion of pericardial drain. Loculated effusion may be difficult to treat. Intrapericardial streptokinase has been used in purulent pericarditis with variable success. Here we present a case of non purulent fibrinous pericarditis treated successfully with intrapericardial streptokinase.

Method: An 11 year old female patient, status post Fontan completion, was incidentally detected to have moderate pericardial effusion on 5 weeks post operative check up. Echocardiography showed 25mm pericardial effusion with strands, towards Right ventricle side, towards conduit and posteriorly, mild flow acceleration in right pulmonary veins, well functioning Fontan circuit, normal ventricular function, not in tamponade. CT scan confirmed our findings with mild encystment of fluid seen in left prevertebral retroatrial region, indenting the left atrium and the pulmonary veins.

Results: Patient was admitted, started on medications and decision to insert intrapericardial drain taken. Intrapericardial pigtail placed drained 160ml serous fluid, however subsequent echocardiography showed 20mm pericardial effusion with multiple loculations

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and fibrinous strands. Hematological and pericardial fluid investigations were not suggestive of infection. In view of persistent collection and no pigtail output, intrapericardial Streptokinase therapy was given for three consecutive days. Following therapy, 150ml fluid drained through the pigtail. No effect on systemic coagulation and complications related to local fibrinolysis were observed. Post procedure echocardiography revealed 3mm pericardial effusion on right ventricle side, well functioning Fontan circuit, no flow acceleration in pulmonary veins.
Conclusions: Cardiac surgery related pericardial effusion is often encountered in pediatric outpatient setting. Presence of loculated and fibrinous pericarditis complicates management and requires prompt intervention to prevent constrictive pericarditis. Trial of intrapericardial streptokinase should be considered in stable patients prior to surgical intervention.

Keywords: Intrapericardial streptokinase, Non-purulent fibrinous pericarditis, post cardiac surgery, post-pericardiectomy syndrome

PP-799

Impact of establishment of pre-operative intensive care unit on outcome of total anomalous pulmonary venous connections: An ambispective analysis

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Background and Aim: Neonates with obstructive TAPVC may present with cyanosis, metabolic acidosis, respiratory failure, and shock requiring pre-operative stabilization in critical care setting. Improvement in perioperative management has markedly changed the outcomes of TAPVC in developing country. Data on clinical profile and outcome of these TAPVC patients are scanty, especially from developing countries. We assessed impact of a newly commissioned dedicated pre-operative Pediatric cardiac intensive care unit (PCICU) on clinical profile, critical care needs and outcome among critically ill TAPVC patients.
Method: A new 10-bed unit was commissioned to manage pre-operative children with heart disease, who present in critically ill state in a Pediatric Emergency of a tertiary care teaching hospital in an LMCI on Nov 2018. Initially it worked in the ward setting with 2 monitors and 1 ventilator. The patients got shifted to the fully-functional formal ICU (with 10 monitors and ICU beds, 5 ventilators and 4 high-flow cannula systems) on Oct 2021. Medical records of admitted children with congenital or acquired heart disease were reviewed retrospectively before shifting to the formal PCICU (Nov 2018–Sept 2021) and prospectively after shifting (Oct 2021-Oct 2023). Clinical characteristics, critical care needs and outcome were compared between these two periods.
Results: Total of 1098 patients with congenital heart disease were admitted during the study period. Complete information could be retrieved form medical records for 1073 patients. [ward setting (Nov 2018-Sept, 2021), n=533; intensive care (Oct 2021-Oct 2023) n=354] and these included non cardiac (70), immediate post op(19), post op follow up (57) and acquired heart diseases (40). Seventy-one patients were admitted with diagnosis of TAPVC [Ward setting, n=45, ICU n=26]. These children were diagnosed at postnatal age of 75 (28;150) days and presented to hospital at 88(29.5;164.25) days. Respiratory distress (69.8%) was the main presenting complaint of these children 52 (73.2%) were Unobstructed TAPVC while 19 (26.7%) were obstructed TAPVC. Supracardiac type was the most common type of TAPVC(n=42, 59.1%).
Conclusions: Identification and presentation to the tertiary care centre and subsequent surgery is significantly delayed in our setting. A significant number of these patients needs intensive care pre-operatively. TAPVC has good outcome on timely critical care. Appropriate pre-operative care is a boon to children with late presentation.

Keywords: TAPVC, PCICU, Critical care needs, Outcome

Table 1

Characteristics		Ward Setting (n=45)	ICU (n=26)	p-value
Gender	Male	34	20	0.896
	Female	11	6	
	Ambigious	0	0	
Age (days) [Median (IQR)]		78(35:180)	90(29:150)	0.873
Age Groups	First Week	5	4	0.927
	Post 7-days Neonates	6	4	
	Post Neonatal Infants	32	16	
	Post-infancy	2	2	
Pre-operative NIRS, n (%)		24(53.3%)	17(65.3%)	0.322
Pre-operative NIRS duration among survivors (days) [Median (IQR)]		5(3.5:14)	9(5:12.5)	0.437
Need of Ventilation, n (%)		17(37.7%)	15(57.6%)	0.104
Pre-operative Ventilation Duration among survivors (days) [Median (IQR)]		7(3:14.5)	4.5(2.75:16.25)	0.662
Need of Inotropes, n (%)		16(35.5%)	13(50%)	0.233
Inotrope duration among survivors (days) [Median (IQR)]		5.5(1.75:12.25)	5(1.5:7)	0.686
Vasoactive Inotropic Score [Median (IQR)]		15(5.5:62.75)	30(15: 71.25)	0.343
Pre-Operative ICU Stay among preop survivors(days) [Median (IQR)]		7.5(6:14)	10(6.25:13)	0.630
Pre-Operative ICU Stay among those who underwent surgery (days) [Median (IQR)]		11.5(6:19)	11(6.75:16)	0.698
Pre-operative Outcome	Discharge	23	11	0.106
	Deaths, n (%)	13(28.8%)	14(53.8%)0	
	LAMA, n (%)	9(20%)	1(0.03%)	
	Transfer for surgery among survivors	16(57.1%)	12(42.8%)	0.167
Post-operative Outcome among those went for surgery (n=298)	Discharge	3	7	0.167
	Death	16	5	
Total Hospital Stay who got operated and survived (days) [Median (IQR)]		36(28:52)	33(26:35)	0.157

Image represents the observations of our study in tabular format

PP-800

UHL's anomaly presenting as severe right ventricular dysfunction without classic signs of right heart failure

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Background and Aim: Right-sided heart failure (HF) is a rare condition in the pediatric population, often posing diagnostic challenges and typically requiring surgical intervention targeted at the underlying cause.
Method: Case report.
Results: Case description: A two-month-old infant was referred with progressively worsening central cyanosis over a period of

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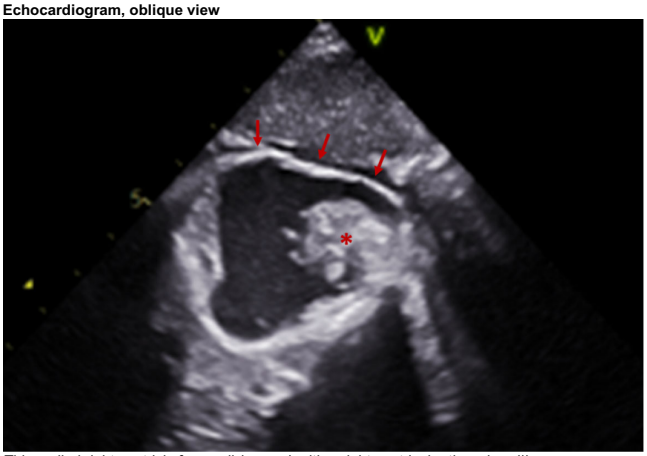
several weeks, unresponsive to oxygen therapy and no other significant findings. An echocardiogram conducted via telemedicine showed severe right ventricular failure, moderate left ventricular dysfunction and an atrial septal defect (ASD) with a right-to-left shunt. Lung pathology was excluded. Despite the age, prostaglandin E1 was transiently initiated, but discontinued due to intolerance, namely frequent apneas and bradycardia. The patient was admitted to an Intensive Care Unit and an echocardiogram revealed a non-contractile thin right ventricle's anterior wall (approximately 1mm), severe right ventricular dysfunction, an apical thrombus in the right ventricle, and moderate left ventricular dysfunction. An angio-CT was performed, ruling out anomalous pulmonary venous return. Given the findings in the context of severe right ventricular dysfunction, Uhl's anomaly was considered as a possible diagnosis. Genetic testing for cardiomyopathies and extensive viral screening yielded negative results. The condition progressively worsened with hypotension and increased lactate levels. Nitric oxide therapy was started and showed transient improvement by reducing right ventricular afterload. An urgent bidirectional Glenn shunt was carried out, including a myocardial biopsy, which confirmed the diagnosis of Uhl's anomaly. Post operatively he was placed on ECMO support. Weaning was complicated by severe sepsis and disseminated intravascular coagulation, from which he demised.

Conclusions: Uhl's anomaly, with fewer than 100 reported cases, entails the replacement of myocardium with fibrous tissue, resulting in right-sided heart failure. This case aims to discuss strategies to maintain appropriate left ventricular preload in the face of severe right ventricular dysfunction due to Uhl's anomaly and the absence of typical signs of right-sided heart failure – swelling and shortness of breath – attributed to the presence of ASD.

Keywords: Right heart failure, Uhl's anomaly

Echocardiogram, oblique view



Thin-walled right ventricle free wall (arrows) with a right ventricular thrombus (*).

PP-802

Impact of provision of pre-operative pediatric cardiac critical care on outcome of coarctation of aorta: an ambispective analysis

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Background and Aim: Critical Coarctation of aorta (CoA) presents in the neonatal period as duct-dependent lesion, while less severe ones may present later with respiratory distress and/or shock with or without pneumonia requiring pre-operative stabilization in critical care setting. Data on clinical profile and outcome of these sick CoA patients are scanty, especially from developing countries. We assessed impact of a newly commissioned pre-operative Pediatric Cardiac Intensive Care Unit (PCICU) on clinical profile, critical care needs and outcome among critically ill CoA patients.

Method: Medical records of children with CoA were reviewed before (Nov 1, 2018–Oct 5,2021) (retrospectively) and after commissioning PCICU (Oct 6, 2021 – Oct 31, 2023) (prospectively). Clinical characteristics, their critical care needs and outcome were compared between these two periods.

Results: Total of 1098 patients with congenital or acquired heart disease were admitted during the study period. Complete information could be retrieved form medical records for 1073 patients. Seventy-four patients had CoA [ward setting (Nov, 2018-Sept, 2021), n=42; intensive care (Oct, 2021-Oct, 2023), n=32] and these included immediate post op (1), post op follow-up (5). Median age at diagnosis was 30(IQR-15.5-59.2) days, (male: female;2.6:1). Respiratory distress was the common presenting symptom. Forty-seven (69.1%) patients presented with respiratory distress, while fifteen (22.1%) patients presented with shock. Isolated CoA was the commonest presentation (n=48; 70.6%), while it was associated with dextro-transposition of great arteries and double outlet right ventricle (n=4; 20%) each, followed by ventricular septal defect and total anomalous pulmonary venous connection (n=3; 15%) each. Seventeen patients (25%) had duct-dependent circulation, and needed Prostaglandin E1 infusion. Clinical profile and outcome of patients are shown in the table.

Conclusions: Establishment of a specialized unit for preoperative care of children with CoA has a crucial role with good outcomes in terms of reduction of inotrope days, pre-op ICU stay among those who went for surgery and total hospital duration. There is

Table: Comparison of Clinical Characteristics, Critical Care Needs and Outcome of patients admitted during the two study periods

Characteristics (n=68) (excluding immediate and post-op follow up)	Pre-shifting (n=37)	Post-shifting (n=31)	p-value
Age (days) [Median (IQR)]	43(16.5,90)	30(21,90)	0.510*
Age Groups, n (%)			
First Week	5(13.5%)	3(9.7%)	0.569*
Post 7-days Neonates	10(27%)	14(45.1%)	
Post Neonatal Infants	18(48.6%)	12(38.7%)	
Post-infancy	4(10.8%)	2(6.4%)	
Gender (M: F)	3.6:1	1.8:1	0.205*
Pre-operative Non-invasive mechanical ventilation (NIMV), n (%)	30(81.1%)	25(80.6%)	0.964*
Pre-operative NIMV duration among survivors (days) [Median (IQR)]	1(1.0,1.0)	9.5(3.5,27.5)	0.347#
Need of Ventilation, n (%)	18(48.6%)	17(54.8%)	0.611*
Pre-operative Ventilation Duration among survivors (days) [Median (IQR)]	26.5(3,50)	5(1.5,17.5)	0.281#
Need of Inotropes, n (%)	17(45.9%)	13(41.9%)	0.740*
Inotrope duration among survivors (days) [Median (IQR)]	3(3.0,3.0)	2.5(1.25,5.25)	0.240#
Maximum Vasoactive Inotropic Score [Median (IQR)]	30(10,40)	5(3(26,80)	0.180#
Need of PGE1, n (%)	9(24.3%)	8(25.8%)	0.466*
PGE1 duration among survivors (days) [Median (IQR)]	5(1,14)	36(24.5,60)	0.143#
Pre-Operative ICU Stay among preop survivors(days) [Median (IQR)]	26(3,49)	13.5(4,41)	0.260#
Pre-Operative ICU Stay among those who underwent surgery (days) [Median (IQR)]	23(13.5,30)	11(7.5,22.5)	0.190#
Pre-operative Outcome			
Discharge, n (%)	21(56.7%)	17(54.8%)	0.631*
Deaths, n (%)	13(35.1%)	13(41.9%)	
Leaving against medical advice, n (%)	3(8.1%)	1(3.2%)	
Transfer for surgery	16(43.2%)	17(54.8%)	0.341*
Post-operative Outcome among those went for surgery (n=33)			
Discharge, n (%)	12(32.4%)	11(35.5%)	0.570*
Deaths, n (%)	4(10.8%)	6(19.3%)	
Total Hospital Stay who got operated and survived (days) [Median (IQR)]	47(42.5,73.5)	30(22.5,60)	0.243#

*Chi-square test had been applied; #Mann-Whitney U test has been applied

Image shows the observation of our study results in terms of critical care needs and outcome of children with coarctation of aorta

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increase in pre-operative non-invasive and decrease in invasive respiratory support suggestive of better care. Pre-operative discharges have increased; however, this could not change the proportion of pre-operative deaths.

Keywords: Coarctation of aorta, Pediatric Cardiac Intensive Care Unit, Critical care needs, Outcome

PP-803

Impact of the dominant ventricle on perioperative outcomes after fontan surgery: a single-center retrospective observational study

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Background and Aim: Increased perioperative pleural effusion and prolonged hospital stay have been reported in patients whose dominant ventricle (DV) is right ventricle. However, we feel that there is a discrepancy with actual clinical course. Therefore, we aimed to clarify the impact of DV on the perioperative outcomes after Fontan surgery.
Method: We enrolled the 96 patients who underwent Fontan surgery from 2013 to 2023 in Hokkaido University. All patients were divided into 3 groups: left ventricle group [L], right ventricle group [R], and undetermined group [U] by defining a scarred ventricle as a ventricle with a preoperative catheterization end-diastolic volume ratio of less than 50% of normal. To clarify the clinical impact of DV, the ANOVA analysis was performed for following parameters during perioperative period such as pleural fluid volume, mean CVP [[(5 to 9mmHg time (hr)*5 + (10 to 15)*10+(15 to 20)*15 + (20<)*20]/total ICU time (hr)], duration of intubation, ICU stay, and hospital stay, Vasoactive-inotropic score (VIS), and several laboratory data such as Base Excess (BE), lactate(Lac) immediately after surgery, and peak serum CK levels during ICU stay.
Results: The three groups consisted of 46 L (48%), 39 R (41%), and 11 U (11%), respectively. The mean value (% or ± SD) [P] of 3 groups were as follow: pleural fluid volume (mL/kg) L:155 ±172, R:145±139, U:213±225 [>0.05], mean CVP(mmHg) L:9.6±1.8, R:10.1±2.3, U:10.7±2.1 [0.05], extubation within 6 hours of ICU admission L:32(70), R:18(46), U:5(45) [0.02], ICU stay(d) L:3.7±1.8, R:4.5±2.7, U:4.1±2.7 [0.04], hospital stay(d) L:22.2±8.5, R:29.3±22.4, U:22.6±6.3 [0.05], VIS(intraoperative/postoperative day 1/postoperative day 2) L:14.2±6.5/11.7±6.0/7.2±3.9, R:17.7±11.9/15.1±10.8/7.0 ±5.6, U:13.6±9.8/10.3±8.3/5.7±4.0 [0.03/0.03/>0.05], BE(mEq/L) L:-3.9±2.4, R:-4.9±2.4, U:-3.3±4.2 [>0.05], Lac(mmol/L) L:2.0±0.9, R:2.2±1.0, U:2.0±1.0 [>0.05], peak CK(U/L) L:677±603, R:610±823, U:527±226 [>0.05], respectively. Although early extubation and ICU discharge were more common in group L, the differences in DV did not affect pleural fluid volume, mean CVP, laboratory data, and hospital days. Inotropes use was higher in R until the first postoperative day, but was adjustable to the same level as the other groups the next day as a result of management to maintain vital data.
Conclusions: While the impact of DV on long-term outcomes is known, the impact on the Fontan perioperative outcomes was limited.

Keywords: dominant ventricle, perioperative outcomes

PP-804

Disruption of the intestinal epithelial barrier is associated with elevated endotoxin exposure and systemic inflammation in fontan patients

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Background and Aim: Disruption of the intestinal epithelial barrier (IEB), the so-called "leaky gut", has been implicated in cardiovascular disease. Whether the IEB is disrupted in Fontan patients remains to be determined. The aims of this study were to assess IEB integrity and identify risk factors for barrier dysfunction in Fontan patients.
Method: This multicenter cross-sectional study included 68 Fontan patients (mean age 11.9 ± 7.5 years; female 38.2%; protein-losing enteropathy (PLE) 20.6%). Fontan patients with favorable hemodynamics (n = 16, 23.5%) were classified as the low risk (LR) group. Serum biomarkers of IEB disruption (intestinal fatty-acid binding protein 2 (FABP2)), endotoxin exposure and antimicrobial immune response (endotoxin-core antibodies (EndoCAb), lipopolysaccharide-binding protein (LBP)), and systemic inflammation (IL-6) were measured. In addition, fecal markers of gut inflammation (calprotectin) and enteric protein-loss (alpha-1 antitrypsin) were obtained in 39 patients.
Results: LR patients had significantly lower FABP2 levels (0.8 (0.5 – 1.6) ng/mL vs 2.0 (1.1 – 4.1) ng/mL, p = 0.006), and NTproBNP z-scores (0.3 (-0.5 – 1.2) vs 1.2 (0.4 – 2.1), p = 0.013) compared with high-risk patients. Patients with PLE or a systemic right ventricle (SRV) had the highest FABP2 levels, suggesting disruption of IEB. FABP2 levels correlated negatively with serum IgG levels (r = -0.310, p = 0.034). NTproBNP z-scores were significantly increased in patients with a SRV (p = 0.012). Interestingly, we found a significant inverse correlation between NTproBNP z-scores and EndoCab IgM (r = -0.297, p = 0.048), indicating increased endotoxin exposure in patients with unfavorable hemodynamics. IL-6 was significantly associated with LBP (r = 0.336, p = 0.039), EndoCab IgA (r = -0.378, p = 0.019), and gut inflammation (r = 0.539, p = 0.002) suggesting that augmented endotoxin exposure, probably due to gut inflammation and IEB disruption, is associated with more severe systemic inflammation. We also found significant correlations between enteric protein-loss, endotoxin exposure and higher IL-6 levels.
Conclusions: Disruption of the IEB is associated with endotoxin exposure and an altered antimicrobial immune response with

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subsequent systemic inflammation in high-risk Fontan patients. Approaches to improve intestinal barrier function may provide a new therapeutic strategy in these patients.

Keywords: Fontan, protein-losing enteropathy, heart failure, inflammation, gut permeability, endotoxemia

PP-805

Atrioventricular valve systolic to diastolic ratio is associated with exercise capacity in fontan patients

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Background and Aim: Echocardiographic assessment of the systemic ventricular function in Fontan patients remains challenging. The atrioventricular valve systolic to diastolic duration ratio (AVVSDr) seems a promising echocardiographic measure of global systemic ventricular function, incorporating both diastolic and systolic function. However, associations between AVVSDr and measures of cardiopulmonary exercise testing (CPET) have not been studied. This study explored associations between AVVSDr and CPET measures in Fontan patients.
Method: A retrospective chart review was performed to identify Fontan patients (aged > 10 years, clinically stable, with a systemic left or right ventricle (SLV or SRV)) who underwent echocardiography and CPET for inclusion in a pilot study. We hypothesized that AVVSDr may be associated with VO2max and VO2max% pred.The AVVSDr was obtained from the Doppler flow signals across the AV-vale in the apical 4-chamber views.
Results: Twenty Fontan patients (30.0% female; age 14.5 ± 1.9 years) were included. Ten patients had a SRV (50.0%), and 9/20 patients were in NYHA class ≥ II (45.0%). The median AVVSDr was 0.86 (0.67 – 1.17), and was significantly higher in patients with a SRV (SLV: 0.67 (0.53 – 0.96) vs SRV 1.01 (0.83 – 1.38), p = 0.019). Of notice, we observed a significant relationship between AVVSDr and heart rate (HR) (r = 0.524, p = 0.018). The AVVSDr corrected for HR was significantly different between dominant ventricle subgroups (p = 0.041) and was higher in those in NYHA class ≥ II (p = 0.025). CPET measures were not significantly different between dominant ventricle subgroups, but VO2max and VO2max%pred were lower in NYHA class ≥ II patients (p = 0.006 and p = 0.013, respectively). We showed significant inverse correlations between AVVSDr, VO2max (r = -0.504, p = 0.024), and VO2max%pred (r = -0.605, p = 0.005), even after correction for HR. Moreover, these associations do not seem to be dependent on ventricular morphology (SRV of SLV) or NYHA functional class (class I vs ≥ II).
Conclusions: We found strong associations between AVVSDr and exercise capacity in Fontan patients, suggesting that AVVSDr is a promising echocardiographic marker of global systemic ventricular function.

Keywords: Fontan, heart failure, ventricular function, exercise capacity

Pulmonary hypertension, heart failure and transplantation

PP-806

Q fever infection: A rare cause of dilated cardiomyopathy in a 2-year-old boy

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Background and Aim: Dilated cardiomyopathy (DCM) is the predominant form of pediatric cardiomyopathies. Etiologies are multiple, with most of cases being idiopathic. In evaluating a child with DCM, it is crucial to eliminate potentially reversible secondary causes.
Method: We report the case of a previously healthy 3-year-old boy, living in an urban area. He was admitted for prolonged dyspnea and asthenia.
Results: No significant family history was found, including early childhood deaths or familial cardiomyopathy. Signs of « Flu-like » illness and fever was noted 20 days ago with a non-specific skin rash on both lower limbs. At admission, the patient had pyrexia of 38.5 °C, tachycardia (130 beats/min), with tachypnea, and normal blood pressure. Physical examination was unremarkable. A chest radiograph showed cardiomegaly. The electrocardiogram indicated sinus tachycardia with signs of atrial hypertrophy. Laboratory tests results showed a normal leukocyte count, thrombocytosis (platelet count of 787.000 cells/L) and increased serum level troponin T up to 0.35 ng/mL (normal< 0.04ng/mL).The initial echocardiography revealed an increase in left ventricular (LV) end-diastolic dimension measured at 43mm, a global hypokinesia and a grade 2 mitral insufficiency. It also shows a reduced ejection fraction of the LV (LVEF) mesered 25-30%. As part of the etiological assessment, a metabolic disorder has been eliminated. Serological tests for herpesvirus, cytomegalovirus, Epstein–Barr virus, ParvoB19, Toxoplasmosis, rickettsiosis and Salmonella Typhi were negative. The blood level of L-carnitine was within the normal range. Q fever was diagnosed on the basis of a positive serologic test of Coxiella Burnetii (phase I IgG titre up to 3400). The patient was treated with Rifampicin (20mg/kg/Daily) and cotrimoxazol (30mg/kg/daily) in addition to the heart failure treatment which resulted in a improvement in cardiac function, and the LVEF was improved to 40% within one month.
Conclusions: The unusual myocardial localization of Q fever pathogen can be severe, emphasizing the importance of conducting Coxiella serology in endemic countries when faced with any case of DCM.

Keywords: Dilated,cardiomyopathy, q fever,cociella burnetti

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PP-807

pulmonary hypertension in patients affected by shone complex: A 15-year retrospective follow-up study

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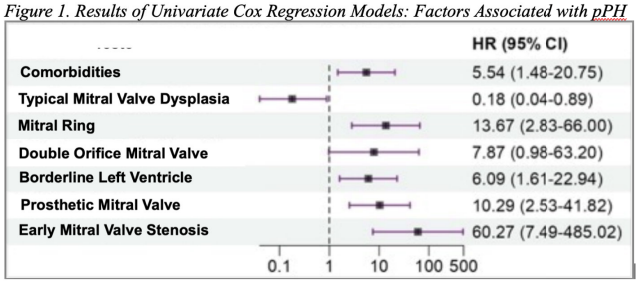
Background and Aim: Children with Shone Complex (SC) often require multiple surgeries and procedures, with their prognosis not fully outlined in existing literature. Pulmonary Hypertension (PH) is a mortality risk factor, especially with increased Pulmonary Vascular Resistance (PVR), leading to combined PH (cPH). Considering the clinical importance of PH, we decided to better analyze patients diagnosed with SC in our care to evaluate the factors associated to the risk of development of cPH.
Method: We examined 104 patients with left heart obstructions and abnormal mitral valves (MV) monitored at our center from April 1981 to July 2023. Retrospective analysis covered clinical, echocardiographic, hemodynamic, and operative data. In line with current literature, post-capillary PH (pcPH) were confirmed if mPAP were >20 mmHg along with a PCWP >15 mmHg in the absence of elevated PVRI. Conversely, under the same conditions but with a PVRI >3 WU/m2, the diagnosis of cPH were established. The primary focus was the development of cPH. Subsequently, the population was stratified by condition severity, functional, and anatomical features.
Results: Among patients who had cardiac catheterization (n=24, 23%), 16 (66%) showed PH, with 11 (46%) having pcPH and 5 (20%) immediately demonstrating cPH. Four (44.5%) up to the patients with pcPH, developed late cPH. The presence of mitral ring (cHR 6.5;p=<0.001), borderline left ventricle (cHR 8; p=<0.001) and mitral stenosis (cHR 5.4;p=<0.03) was associated with a higher risk for pcPH. Early mitral stenosis (developed within the first year of life) was an independent risk factor for pcPH (aHR12.1;p=0.008) and significantly increased the risk of cPH al well (cHR 29.7;p=<0.001). Identified risk factor for cPH are summarized in figure 1. Anatomically, BLV diagnosis (aHR6.1;p=0.008) and the presence of a mitral ring anomaly (aHR13.7; p=0.001) were independent risk factors for cPH.
Conclusions: Our findings suggest that the presence of a mitral ring and a borderline left ventricle may be potential indicators of the severity of the condition in patients with SC, possibly associated to a predisposition to cPH. In order to improve the management of this patients and the perinatal counseling it may be useful to investigate the genetic background of this predisposition.

Keywords: Pulmonary Hypertension, Shone Complex

Risk factor for Combined Pulmonary Hypertension

Figure 1. Results of Univariate Cox Regression Models: Factors Associated with pPH



Results of Univariate Cox Regression Models: Factors Associated with pPH

PP-808

Combined pulmonary hypertension in patients with multiple left heart obstruction in shone complex

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Background and Aim: The management of patients with multiple left heart obstructions in Shone Complex (SC) becomes more challenging when complicated by Combined Pulmonary Hypertension (cPH). Moreover, it has been observed that some children develop cPH early, even in the absence of chronic exposure to increased left atrial pressures. Considering the clinical significance of cPH, we decided to reevaluate our patients with SC complicated by cPH to try to identify risk factors for early elevation of Pulmonary Vascular Resistance (PVR).
Method: We examined 104 patients with left heart chamber obstructions and abnormal mitral valves at our center from April 1981 to July 2023. Retrospective analysis covered clinical, echocardiographic, hemodynamic, and operative data. The primary focus was the development of cPH. In line with current literature, cPH were confirmed if mPAP were >20 mmHg along with a PCWP >15 mmHg and PVRI >3 WU/m2. Patient who showed cPH with a previous diagnosis of post-capillary PH (pcPH) was considered to have a late cPH (l-cPH), conversely they were classified as affected by early cPH (e-cPH).
Results: Among patients who had cardiac catheterization (n=24, 23%), 9 (37.5%) showed cPH with an average time of onset of 6.2 ± 4.6 years. Ten-year probability of cPH-free survival was significantly reduced in patients with mitral stenosis (MS) developed within the first 12 months of life (63.0% vs. 98.7%; p=<0.001) and in those with a borderline left ventricle (BLV) compared to patients without these conditions (83.6% vs. 95.5%; p=0.0025). Five (55.6%) patients demonstrated e-cPH and 4 (44.4%) developed this condition chronically (l-cPH). In the bivariate analysis (Table 1), within the two subgroups, e-cPH appears to be associated, approaching statistical significance, with the presence of a ring-type mitral anomaly.
Conclusions: The development of cPH in patients with SC appears to be more common in patients with early MS and BLV. The presence of a ring-type mitral anomaly is associated with e-cPH; however, further studies are needed to substantiate this finding.

Keywords: Pulmonary Hypertension, Shone Complex

Bivariate Analysis between Patients with e-cPH and l-cPH

Figure 1. Bivariate Analysis between Patients with e-cPH and l-cPH

	e-cPH (n=5)	l-cPH (n=4)	p
Comorbidity	2(50.0%)	2 (50.0%%)	0.764
Typical Mitral Displasia	1 (50.0%)	1 (50.0%)	0.858
Mitral Ring	5 (71.4%)	2 (28.6%)	0.073
Borderline Left Ventricle	3 (60.0%)	2 (40.0%)	0.764
Mitral Valve Stenosis	5 (62.5%)	3 (37.5%)	0.236
Aortic Stenosis	2 (50.0%)	2 (50.0%)	0.764

Bivariate Analysis between Patients with e-cPH and l-cPH

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PP-809

Therapies for pulmonary hypertension associated with bronchopulmonary dysplasia: A systematic review

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Background and Aim: Bronchopulmonary dysplasia (BPD) is the most common chronic lung disease associated with prematurity. Pulmonary hypertension affects an important proportion of infants with BPD and is associated with worse outcomes. Despite this, evidence and consensus is lacking on treatments for infants with this condition.
Method: Two independent researchers performed a systematic review of the literature using the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidance. The review focused on publications between January 2010 and June 2023 written in English that addressed targeted treatment of pulmonary hypertension in infants with BPD and reported on markers of pulmonary hypertension or clinical outcomes.
Results: The search initially identified 489 articles, of which 14 were included in the review. The most commonly used therapy that targeted pulmonary hypertension was sildenafil. Table 1 shows reported outcomes for infants receiving therapy for pulmonary hypertension associated with BPD.
Conclusions: Therapies for pulmonary hypertension in infants with BPD are associated with an improvement in clinical or echocardiographic markers of pulmonary hypertension in 35-88% patients. However, mortality in this population remains high (9-33%) and the quality of evidence is low, with most studies being retrospective and patients acting as their own controls. Therefore, further research is needed to determine the effects, safety and optimal duration of standalone and combination therapies for pulmonary hypertension in infants with BPD.

Keywords: pulmonary hypertension treatment, bronchopulmonary dysplasia

Reported outcomes for children receiving therapy for pulmonary hypertension associated with bronchopulmonary dysplasia.

	Cohen	Del Cerro	Dillon	Hernandez	Jeremiasen	Kadmon	Migdal	Nees	Nyp	Piastra	Qasim	Sallmon	Tan	Trottier-Boucher
Year Published	2019	2014	2023	2020	2022	2016	2021	2020	2012	2011	2018	2022	2015	2015
Design	R	R	R/P	R	R	R	R/P	R	R	R	R	R/P	R	R
Study size	135	22	19	15	74	20	20	101	21	10	3	15	22	23
Average GA (weeks)	Not Included	27 (24-30)	24 (23-25)	Not Included	22–27 (73%) 28–36 (27%)	25 (23-31)	27.7 (± 4.26)	25 (I24-27)	27 (23 - 33)	29.5*	26*	24.6 (24.4-24.9)	25.6 (± 1.3)	26
Average BW (g)	Not Included	740 (490-1450)	598 (572 – 735)	Not Included	727 (635 - 841)	Not Included	735 (675 - 920)	646 (540 - 790)	641 (460 - 1900)	No: Included	543*	650 (550 - 695)	631 (± 181)	710
Age at Start of Therapy	4 months (1 week - 6.9 years)	Not Included	40.4 Weeks	250 days (112 - 540)	7 months (5-10.25)	0.4 years (0-2)	1.8-25 months	Not Included	167 days (82 - 307)	6.0months (2.75-9.50)	37+5*	Not Included	169 days (I126 - 219)	106
Duration of Therapy	1.75 years (2 weeks - 5.7 years)	Not Included	Not Included	258.5 hours (77 - 1263)	4.6 months (1.9-6.8)	Not Included	Not Included	Not Included	Not Included	5.5days (2.7-8.5)	4 weeks	Not Included	Not Included	71 days
Experimental Intervention	Sildenafil Monotherapy(90%) Combination therapy (10%)	Sildenafil (100%) Bosentan (32%) Inhaled iloprost (23%) Subcutaneous treprostinil (18%) Combination Therapy (41%)	Sildenafil	ECMO	Sildenafil (93%) Bosentan (12%) Iloprost (9%)	Sildenafil (60%) Sildenafil+ Bosentan (25%) Bosentan (5%)	Sildenafil (85%), Bosentan (5%) Sildenafil + Bosentan (10%)	Sildenafil (98%) Bosentan (13%) Inhaled iloprost (35%) IV Epoprostenol (12%). SC Treprostinil (9%)	Sildenafil	Sildenafil	Sildenafil	Sildenafil (73%) Sildenafil +Macitentan (27%)	Sildenafil	Sildenafil
Side Effects	Airway spasm with desaturation (1%)	Sildenafil: abdominal distension (5%), erections (9%), worsening of GERD (9%) Iloprost: hemoptysis (60%) Treprostinil: severe local reaction (25%) Bosentan: elevation in liver enzymes (14%)	ROP (11%)	Inotropes use (33%) Cannulation site bleeding (28%) RRT (27%)	Not Included	Bosentan: raised liver enzymes (33%)	Not Included	Sildenafil: increased respiratory requirement or frequency of desaturations (4%), hypotension (1%), irritability (1%), airway spasm (1%) Iloprost: increased respiratory requirement or frequency of desaturations (11%), hypotension (3%), pulmonary oedema (3%)	Hypotension (5%)	Nct Included	None Observed	None Observed	None Observed	Transient hypotension (43%) Increased oxygen requirement (4%)
Study duration	2004-2015	March 2006 - December 2011	January 2018 - November 2020 (Retrospective) December 2020 - January 2022 (Prospective)	1982 - 2018	2007-2017	2008-2014	2009-2015 (Retrospective) 2016-2020 (Prospective)	2005 - 2016	2005 – 2009	2007-2009	Not Included	January 2016 - December 2018	2004-2012	January 2009 - May 2013
Follow up	3.7 Years (6 months - 10 years)	35 Months (21-91)	14 weeks	Not Included	Not Included	2 years (0.3 - 5.5)	15.9 Months (2-53.1)	5.1 years (0.38 - 12.65)	12 Months	1 Month	Not Included	12 Months	1 Year	Not Included
Mortality (%)	26	24	21	13	9	0	30	33	19	No: Included	Not Included	Not Included	27	22
Proportion of patients showing an improvement in PH (%)	45	50	42	93	Not Reported	50	50	77	70	70	67	60	67	57

R = Retrospective, R/P = Retrospective and Prospective, ROP = Retinopathy of Prematurity, RRT = Renal Replacement Therapy.

PP-811

Impact of collaboration to improve outcomes after heart transplantation in a small country: the slovak experience

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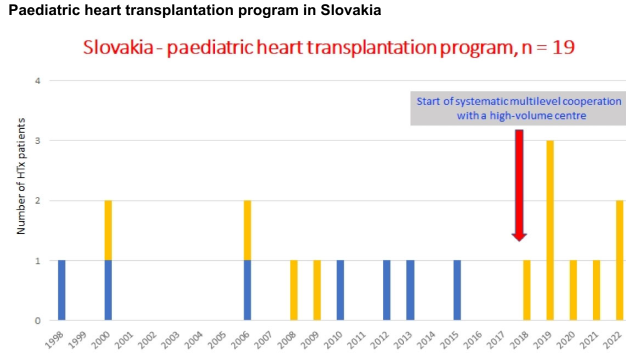
Background and Aim: Worldwide, there are only a few countries with population less than 6 million where a comprehensive paediatric heart transplantation (HTx) program is available. Slovakia, with a population of 5.4 million has one paediatric cardiac surgical program located in Bratislava. The first paediatric HTx was performed in 1998 with only 10 additional HTx during the subsequent 20 years. We sought to analyse how systematic collaboration with a high-volume transplant centre inclusive of initial HTx expert site visit to Bratislava, adoption of transplantation protocols, and consultations on individual patients including video conference sessions influenced early outcomes.
Method: A retrospective review of modes, frequency, and content topics of consultations during 5-year period was performed. Comparison of paediatric HTx outcomes in Bratislava before (1998-2017, era I) and after (2018-2022, era II) implementation of multilevel collaboration with the high-volume HTx centre was also performed.
Results: Since 1998, 19/34 children listed for HTx in Bratislava have undergone HTx (11 in era I, 8 in era II; Figure 1). During era II, consultations for individual patients with the high- volume center consisted of 15 video conference sessions, 30 e-mail interactions, and 4 urgent telephone calls. The main topics of individual consultations were transplant candidacy (n=12), pre-transplant care and mechanical support (n=12), evaluation of donor suitability (n=2), acute post-transplant care (n=3), immunosuppression management (n=8) and chronic post-transplant care (n=12). Waiting list mortality decreased from 48% in era I to 11% (p=0.06) in era II. Patients and procedural characteristics were comparable between the 2 eras. Three patients had pretransplant mechanical circulatory support during each period. The incidence of rejection within the first year after HTx decreased from 64% in era I to 13% in era II (p=0.03). Seven patients died after HTx during era I and none during era II, including survivors from era I.
Conclusions: Implementation of a systematic multilevel cooperation with a high-volume HTx centre including utilisation of video conference was associated with trends to higher transplant activity, reduced waiting list mortality and improved early outcomes in a national low-volume paediatric HTx centre. Such collaboration may serve as a model for other countries starting new paediatric HTx programs.

Keywords: paediatric heart transplantation, outcomes, telemedicine

Paediatric heart transplantation program in Slovakia



PP-812

Paediatric heart transplantation in congenital heart disease – single centre experience over 10 years

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Background and Aim: Children with congenital heart disease (CHD) with failing cardiac function following palliative or corrective cardiac surgery are increasing in prevalence and have become a common indicator for heart transplantation over the last decade. We report our single centre 10-year experience. This retrospective study aims to identify pre- and post-transplant factors associated with morbidity and mortality in children with CHD, who underwent cardiac transplantation.
Method: We conducted a retrospective analysis of paediatric patients (aged 16 years or less) with CHD who underwent heart transplantation at Newcastle Freeman Hospital between January 1st 2013 and December 31st 2023.
Results: A total of 71 orthotopic heart transplants were performed on 66 patients with CHD. 36 were males (54.5%), and 30 were females (45.4%). Notably, 5 patients (7.57%) required re-transplantation. The mean age of this cohort was 10.6 years (SD 4.8; range 5 months-18 years), and the mean age at the time of heart transplant was 6.5 years (SD 4.6; range 5 months-17 years). The average weight at the time of transplant was 35.2kg (SD19.6; range 3.6-96kg). There was significant heterogeneity in pre-transplant diagnoses. The majority of these patients had single ventricle physiology (50; 74.2%) and 54% (27/50) of this cohort had Hypoplastic Left Heart Syndrome (HLHS). In pre-transplant assessments, the average number of sternotomies was 2.88 (range 1-5). 15% (10/66) of patients had baseline renal function impairment (eGFR <30 ml/min/1.73m^2), and 33% (22/66) had gastrointestinal comorbidities, including 11% (7/66) requiring feeding through gastrostomy. Post-transplant, the mean Paediatric Intensive Care Unit (PICU) stay was 14 days, and 13% of patients required V-A ECMO support. Complications during an 8.6-year follow-up included 3.5% experiencing rejection, 85% dealing with infections necessitating IV antibiotics, 11% developing coronary vasculopathy, and 7% facing Post-Transplant Lymphoproliferative Disorder (PTLD).
Conclusions: In this high-risk CHD cohort, we observed a higher mortality rate than those with paediatric cardiomyopathy. The mean survival time for the CHD group was 4.86 years (SD 3.64). These findings highlight the complexities and challenges associated with heart transplantation in paediatric patients with CHD, emphasising the need for continued research and refined clinical strategies to improve outcomes in this vulnerable population.

Keywords: Paediatric Heart transplantation, congenital heart disease, End-stage Heart failure.

PP-814

Mortality and heart disease in children at a tertiary center

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Background and Aim: Mortality from heart disease is frequently reported in registries, but rarely with causes of death and often with very limited clinical characteristics.
To describe the distribution of causes of death in children with heart disease over a period of 11 years in a single institution (2010-2020), and to estimate the evolution of mortality rates over the same period.
Method: Each death between the ages of 0 and 18 years was prospectively collected. Clinical and demographic data on circumstances and cause of death were retrospectively extracted from the computerized medical record. Mortality rates are presented by type of heart disease and immediate cause of death. Changes in mortality rates by underlying cause during the study period were analyzed.
Results: 929 patients died during the study period, including 186 (20%) newborns receiving compassionate primary care, 699 (75.2%) congenital heart defect (CHD), 140 (15.1%) cardiomyopathies (CM), 40 (4.3%) pulmonary arterial hypertension (PAH), 20 (2.2%) isolated arrhythmias or conduction disorders. Among them, 366 (39.4%) were born prematurely, including 281 (40%) CHD and 69 (49%) CM. Syndromic patients or those with genetic mutations represented 289 (31.1%) patients, of whom 212 (30%) had CHD, 44 (31%) CM, and 21(53%) PAH. After excluding children in compassionate care, age at death was younger than one year for 479/743 (64%) patients, and mortality related to CHD remained the prominent cause until the age of 12 years. Approximately 50% of the cardiac mortality occurred within 30 postoperative days for CHD with 431(58%) of children having complex CHD. Heart failure was by far the most common cause of death in these children (39%), followed by PAH/pulmonary hypertension crisis (12%), sepsis (11%), sudden death (9%), respiratory failure (8%), neurological impairment (6%), arrhythmia or conduction disorder (4%). There were no significant changes in mortality rates overall or by population subtype during the study period.
Conclusions: Mortality in this center is led by death from heart failure and PAH crisis. Death occurs preferentially postoperatively in neonates, premature infants and infants with complex CHD. Efforts to manage heart failure and pulmonary hypertension are critical to reducing cardiac mortality in children.

Keywords: mortality, heart failure, pulmonary hypertension, preterm birth

PP-815

Pulmonary veno-occlusive disease exhibiting a pulmonary vasodilator-tolerant phenotype in a patient with a novel EIF2AK4 gene variant

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Background and Aim: Pulmonary Veno-Occlusive Disease (PVOD) is a rare and life-threatening condition. It is characterized by selective pulmonary venule remodeling, often exhibiting symptoms similar to PAH. Diagnosis relies on pulmonary function tests, arterial blood gas analysis, non-contrast chest computed tomography (CT) and genetic testing.
Method: Confirmation of heritable PVOD/pulmonary capillary hemangiomatosis (PCH) relies on the detection of biallelic EIF2AK4 mutations. Drug-induced pulmonary edema when exposed to standard PAH therapies is a phenomenon indicative of PVOD or PCH.
Results: We report a case of a 17-year-old girl with an incidental diagnosis of PAH presenting with history of frequent syncope and resting oxygen saturation of 93%. Echocardiography identified moderate to severe tricuspid regurgitation with preserved right ventricle function with right ventricle pressure estimate of 90

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mmHg. NTproBNP was elevated to 3730 ng/l. Due to these findings and the inability to complete the standard 6-minute walking test, the patient was stratified to WHO functional class III. Pulmonary function tests indicated severely diminished diffusing capacity for carbon monoxide (22%). CT angiography confirmed features indicative of PVOD. Consequently, a dual pulmonary hypertension therapy was initiated (sildenafil, bosentan) with sequential titration. While symptoms improved (syncope disappeared, NTproBNP dropped to 995 ng/l), functional class remained unchanged. Cardiac catheterization revealed negative vasoreactivity testing, pulmonary vascular resistance 18Wu/m2, and mean pulmonary artery pressure 44mHg on ambient air. The therapeutic regimen was cautiously expanded to include subcutaneous infusion of treprostinil with normalization of NTproBNP and without onset of pulmonary edema. Genetic analysis detected a novel biallelic mutation in EIF2AK4 (c.1021C>T, p.Gln341Ter). The patient demonstrated a pulmonary vasodilator tolerant phenotype of PVOD. Due to partial improvement of clinical state with persistent exertional hypoxemia, we kept the patient on triple PAH therapy and started preparing her for lung transplantation.
Conclusions: To our knowledge, this is the first report of a pediatric PVOD case with a novel biallelic EIF2AK4 gene mutation. It highlights the variable clinical response of PVOD patients to pulmonary vasodilator therapy. In the absence of pulmonary edema, specific PAH therapy may aid in stabilizing the patient as a bridge to lung transplantation.

Keywords: Pulmonary Veno-Occlusive Disease (PVOD), Pulmonary Arterial Hypertension (PAH), EIF2AK4 mutation

PP-817

Potts shunt as a palliative treatment option for suprasystemic idiopathic pulmonary arterial hypertension: an in-silico modelling study

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Background and Aim: The Potts Shunt (PS) is proposed as palliative treatment option for critically ill pediatric patients suffering from suprasystemic Pulmonary Arterial Hypertension (PAH) and right ventricular (RV) failure. It is an extracardiac shunt, that connects the Left Pulmonary Artery to the Descending Aorta, to decompress the overloaded RV. Different variants have been created from anastomoses and grafted conduits, and recently incorporating valves to prevent backflow for infrasystemic pulmonary pressure, without consensus. The aim of this study is to assess the patient-specific hemodynamics following the integration of the shunt, while investigating the differences across the various potential shunt modifications, by implementing multi-fidelity computational models.
Method: A hemodynamics virtual twin of a 13-year-old patient treated at Necker Children's Hospital was built. The patient presented suprasystemic pulmonary pressure, as well as a hypertrophic RV, and was treated with a conduit-based shunt. The present multi-fidelity model is capable of visualizing the flow in the shunt region capturing the blood flow dynamics of the system, and predicting its effect on the rest of the body, including the heart, the lower and upper body circulation.
Results: The PS resulted in a near-equalization of the aortic and pulmonary artery pressures for an implanted shunt diameter of 7.6mm, in the patient as in-silico. The model further predicted that the RV working volumes, EDV and ESV decrease, leading to RV decompression. Yet, the stroke volume (SV) rises, thus actually increasing RV workload. Increasing the shunt diameter results in a drop of Qp/Qs), with a higher shunt flow, but also inducing backflow from the systemic to the pulmonary circulation, indicating the necessity of a unidirectional valve. By comparing four different cases (valved/non-valved anastomosis and conduit), our model demonstrates preserved oxygen saturation in the upper body, with larger diameters reducing lower body saturation. All of the variants lead to acceptable hemodynamics indices.
Conclusions: This in-silico model efficiently captures pre and post operative patient-specific hemodynamics, offering valuable insights into the physiology of this disease. With further validation, it can serve as a predictive tool for clinical decision-making regarding the optimal shunt design selection for suprasystemic PAH.

Keywords: pulmonary arterial hypertension, Potts shunt, virtual human twin, in-silico medicine, intervention planning

PP-818

Unconventional ventricular assist device support in complex congenital heart defects as a bridge to heart transplant

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Background and Aim: Long-term mechanical circulatory support (LT-MCS) is an important treatment modality for patients with severe heart failure (HF). Ventricular assist device (VAD) in single ventricle physiology or congenital heart defects (CHD) with intracardiac shunts presents a technical challenge in implantation and management.
Method: Retrospective chart review of clinical and surgical data of patients with VAD in a tertiary care children's hospital.
Results: Case 1. 11-year-old girl with tricuspid atresia and pulmonary stenosis. She underwent a systemic-to-pulmonary shunt in neonatal period, a bidirectional Glenn procedure at 10 months of age, a proximal right pulmonary artery stenting at 2 years of age, and a Glenn takedown (because of elevated pulmonary artery pressures and hypoplastic pulmonary branches) and right systemic-to-pulmonary shunt at 10 years of age. Given persisting severe hypoxemia and NYHA functional class IV she was eligible for heart transplant (HTx). The concomitant implantation of VAD as subpulmonary MCS and the reconstruction of central pulmonary arteries and ascending aorta with Dacron grafts were performed. She was successfully transplanted on day 72.
Case 2. 5-year-old girl with multiple ventricular septal defects (VSDs) or "Swiss cheese VSDs". She underwent pulmonary artery banding at 3 months of age. Heart surgery was discarded due to its complexity. At 5 years of age, she developed biventricular diastolic dysfunction, so a hybrid approach for closure of VSDs was tried without success. Given the hemodynamic and respiratory support (and kidney and hepatic failure), she was included on the HTx list. Unconventional VAD was implanted with tricuspid and pulmonary valve closure and removal of septal devices (conversion to single ventricle physiology). Cannulas were placed in right atrium-pulmonary artery and left ventricle-aorta. She was transplanted in stable condition on day 109.
Conclusions: Adequate LT-MCS as a bridge-to-transplant in complex CHD is essential for the improvement and recovery of other organ function and contributes to the success of HTx.

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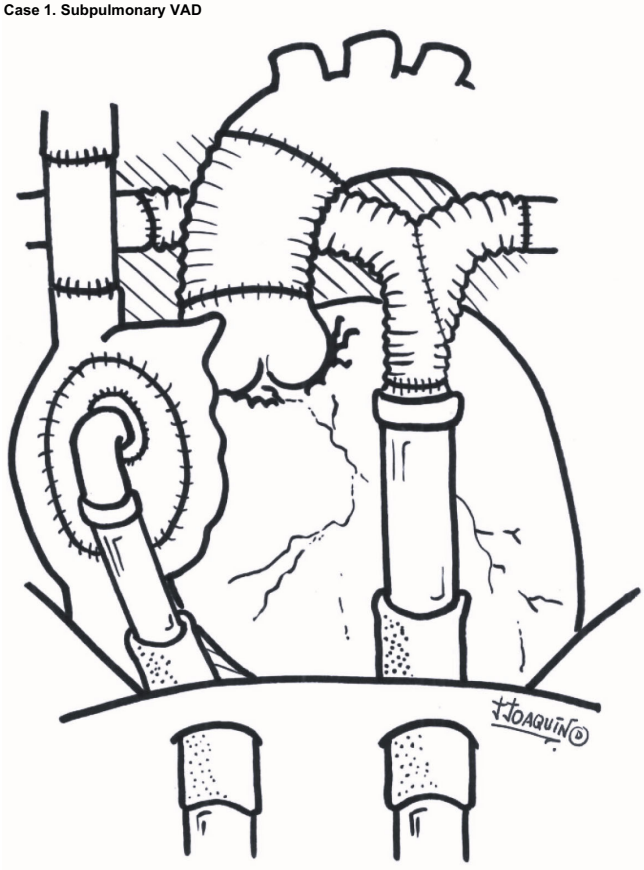
The combined implantation of VAD and the reconstruction of the pulmonary artery and aorta might simplify and reduce the ischemic time of the upcoming HTx.
In the context of multiple VSDs and biventricular HF, a possible alternative for VAD success is a conversion to single ventricle physiology.

Keywords: Ventricular assist device, heart transplant, single ventricle, ventricular septal defects

Case 1. Subpulmonary VAD



PP-819
Heart rate variability as a prognostic marker in pediatric pulmonary arterial hypertension- a prospective observational study

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Background and Aim: Risk stratification in adults patients with PAH is standardized modality to assess morbidity and mortality. However similar risk stratification in pediatric patients with PAH is challenging.The current modalities of risk stratification are either invasive or subjective and may not be performed in across the pediatric age group. Hence there is a pressing need to identify a noninvasive objective parameter to assess risk in pediatric PAH
Method: Prospective Study period Jan 2022 – sept 2023. All prevalent paediatric patients with IPAH who were stable on dual combination therapy were included in the study. Patients were divided into three risk categories using the EPVD risk calculator. Standard deviation of normal-to-normal intervals (SDNN), Standard deviation of mean values for normal to normal intervals (SDANN) and square root of the mean square differences of successive RR intervals (RMSSD) were measured using 4 hours of holter monitor. On follow up patients were assessed for disease progression (increase NT-pro-BNP, decrease 6MWTD >10%, deterioration FC by 1, death or listing for lung transplant/ Potts shunt. Mean values of HRV were compared across groups.
Results: 46 (28F), mean age 8.6 +/- 4.9 years, weight 22 +/- 13 kg and BSA 0.8 +/- 0.3. 15,17 and 14 were in high, intermediate and low risk category respectively. Patients in high risk category had lower values of HRV [SDNN (60.2 ±31.7 vs 79.5 ± 24.01 vs 80.6 ±28.4ms, p= 0.019), SDANN (49.5± 26.4 vs 62.2 ± 18.2 vs 63.4±27.1ms, p= 0.08), RMSDD (20.8 ±11.5 vs 33.4 ±18.3ms vs 32.4 ±14.4 ms, p= 0.05)] as compared intermediate and low risk. Patients (10) who had clinical worsening had lower values of SDNN (62.4 ±21.9 vs 78.6 ± 32.3ms, p= 0.02), SDANN (50.3± 18.09 vs 62.05 ± 27.9 ms, p= 0.05) and RMSDD (22.7 ±13.5 vs 31.6 ±15.8ms, p= 0.015)]. SDNN of 61.7 ms, SDANN of 49.7 msec and RNSDD of 20.1 msec were able to predict disease progression with good sensitivity and specificity
Conclusions: HRV is significantly lower in high risk category and those who demonstrated disease progression and could be used as a non-invasive, objective parameter to predict disease progression.

Keywords: Risk stratification, Heart rate variability

PP-820
Clinical outcomes and transition patterns of pediatric pulmonary hypertension patients upon reaching adulthood in poland

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Background and Aim: The care of children with pulmonary hypertension (PH) in Poland is centralized in 8 medical pediatric centers. Upon reaching 18 years of life, patients are transferred to one of the 23 centers for adults with PH. This study aims to assess key clinical

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parameters and outcomes in patients with PH before and after transfer to adult care.
Method: We analyzed data from the Polish multicentre registry (BNP-PL) from 2018 to 2023 that included number of patients reaching adulthood, treatment of PH, World Health Organization Functional Class (WHO-FC), control right heart catheterization (cRHC), survival, listing for lung transplant in the year before and after the transfer from pediatric to adult PH center.
Results: Among 167 pediatric patients with PH, twenty three PAH patients (12 females) reached adulthood and were transferred to adult PAH centers. Most (n=16) patients were diagnosed with PAH associated with congenital heart disease, followed by idiopathic PAH (n=6) and PAH associated with portal hypertension (n=1). Data after the transfer were available for 18 patients, with 5 lost to follow-up. Two patients died, one before and one after the transfer. Before transfer, 10 patients were on monotherapy, 8 on dual, and 5 on triple combination therapy. After the transfer, 2 patients were on monotherapy, 8 on dual and 4 on triple combination therapy. None of the patients had a cRHC before the transfer, 5 patients had cRHC after the transfer. Before the transfer, 4 patients were in WHO-FC class I, 12 patients in class II, 6 patients in class III, and 1 patient in class IV. After the transfer 1 patient was in class I, 11 patients in class II, 3 in class III and 1 patient in class IV. Two patients were listed for heart transplant before transfer, one additional patient was listed after the transfer.
Conclusions: Approximately 21% of patients were lost to follow-up during the transition, emphasizing the need for improved transition protocols. The transition process positively influenced treatment modalities, with increased utilization of combination therapy and cRHC post-transfer. Disease severity assessment and lung transplant listing remained stable.

Keywords: pulmonary hypertension, database, transfer to adult

PP-821
Dilated cardiomyopathy in children: 15 years retrospective study in tertiary center in Tunisa
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Background and Aim: Dilated cardiomyopathy (DCM) is a progressive disease characterized by ventricular enlargement and contractile dysfunction. It represents the largest group of pediatric cardiomyopathy type. This study aims to report the clinical presentation, aetiology, and outcome of DCM in children from a tertiary center in TUNISIA and to assess the prognostic factors.
Method: This is a Retrospective review of all patients with DCM, aged <15 years, presenting to Hedi CHEKER hospital between 01/01/2008 and 01/10/2023. Subjects were sourced via echocardiography database and clinical record
Results: Sixty-eight patients were identified. Median age at diagnosis was 3 years old. The age of the population vary from a 1moths to 15 years. Sex ratio was 1.06 linical features was mainly represented by heart failure (85%), fever (27%) and arrhythmias (15%). Half of patients required inotropic support during their hospitalization. Echographic findings include left ventricular enlargement and dysfunction for all patients. We note biventricular dysfunction with associated right ventricular dysfunction in 13%. Mitral valve regurgitation was noted in 85% and was severe in 3 patients. Pulmonary hypertension was noticed in 26%. The commonest aetiology was Idiopathic DCM at 50% followed by myocarditis in 25% (figure).Overall outcome was death (in 35 patients/68) rehospitalization (67%) and normalization of the ventricular function in 22 patients. No patient receive heart transplantation. Multivariable analysis identified older age at diagnosis (P<0.001), larger left ventricular end-diastolic M-mode dimension z-score (P<0.001), as independent predictors of death. Myocarditis indicate better prognosis outcome with a recovery in 9 patients /17.
Conclusions: DCM is the most common pediatric cardiomyopathy. The prognosis is still poor, especially in countries where heart transplantation has not yet been developed early diagnosis, especially of secondary forms, may improve prognosis.

Keywords: dilated, cardiomyomathy, heart failure, prognosis

PP-823
Non-invasive quantification of pulmonary vascular characteristics in children with pulmonary arterial hypertension using CTA-THORAX
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Background and Aim: Pediatric pulmonary arterial hypertension (PAH) is a progressive disease leading to right heart failure and eventually death and is characterized by pulmonary vascular remodeling. Currently right heart catheterization is the golden standard for diagnosing PAH and assessing disease progression. Following recent studies in adults with pulmonary hypertension, we performed a feasibility study using CTA-Thorax aimed to non-invasively quantify pulmonary vascular characteristics in children with PAH and compare these to pediatric controls. We further explored correlations between these characteristics and conventional invasive hemodynamic measurements.
Method: In this study, we retrospectively collected 30 CTAs, from 15 patients with idiopathic PAH and 15 age and sex matched controls. The pulmonary vessels were then segmented using a scale space vessel segmentation algorithm, and then skeletonized using the TEASAR method. Using a personalized definition of small vs large pulmonary vessels calibrated to the patient's size, we calculated the respective vessel volumes for each subject, geometric parameters of the vessels and compared patients with controls. Finally, we correlated these measurements with conventional, invasive hemodynamic variables.
Results: Children with pediatric PAH had significantly lower small vessel volumes (p<0.05) and significantly higher large vessel volumes (p<0.05) than controls (See figure). Also, patients had significantly higher vessel tapering rates (p<0.05) than controls. Lastly, explorative analyses in the patient group showed associations between small vessel volume and pulmonary vascular resistance index, as well as between small and large vessel volumes and mean pulmonary arterial pressure, although statistical significance of these was not achieved in this feasibility study.
Conclusions: In this feasibility study we successfully non-invasively quantified the pulmonary vasculature using CTA thorax in children with PAH and found significant differences when comparing vascular characteristics with those in control subjects, including small and large vessel volumes and tapering rate. These volumetric measurements tend to correlate with invasive hemodynamics. Further studies are required to investigate the potential clinical use of these the non-invasive quantification of the pulmonary

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vasculature to assess disease progression, prognosis or even treatment effect.

Keywords: pediatric pulmonary arterial hypertension, chest computed tomography, pulmonary vasculature, pulmonary vascular resistance, mean pulmonary arterial pressure, scale space segmentation

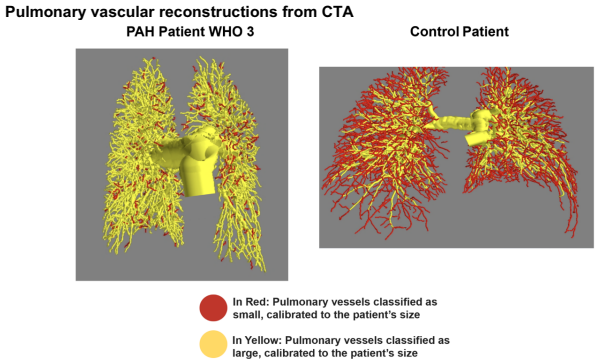

Figure Legend: *In the above figure, vascular reconstructions as measured from CTA are represented from a pediatric idiopathic PAH patient (Left Image) and a pediatric control patient (Right Image). After calibration for the patient's size, vessels represented in red were classified as small pulmonary vessels and vessels represented in yellow were classified as large pulmonary vessels.*

PP-824

Computational modeling for pulmonary arterial hypertension patients

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Background and Aim: Mathematical models based on biophysical principles formalize the relationship between the properties of the cardiovascular system and its function. Therefore, by solving the inverse problem important parameters related to pulmonary arterial hypertension (PAH) can be extracted from clinical data. The right ventricular (RV) function is an important determinant of outcome in PAH. The aim of this research is to create a personalized model for different patients to better characterize RV adaptation and ventricular vascular coupling.

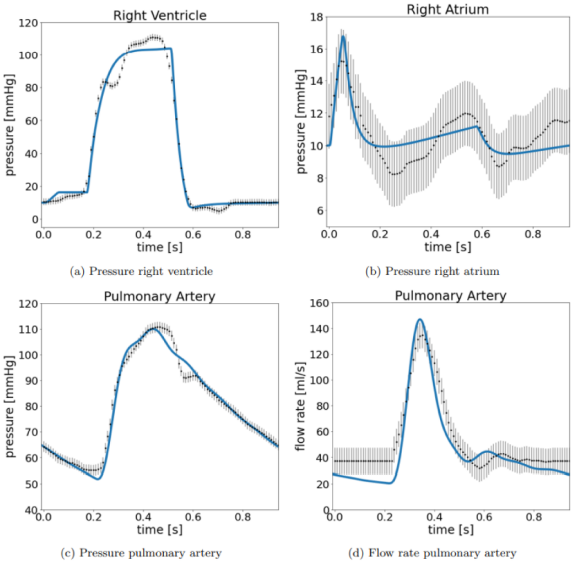
Method: The proposed 0D lumped parameter model within this research is a closed-loop system – written in terms of differential equations – consisting of the right heart, pulmonary circulation, left heart, and systemic circulation. The equations are solved for pressures and flow rates computationally using a tailor-made numerical method. Our approach places significant emphasis on the correction of heterogeneous data sources when estimating parameters.

To create a personalized model the pressure data obtained by cardiac catheterisation and MRI flow rate data are used. The sum of the squared error (SSE) is minimized by using an appropriate fitting algorithm. Personalized models are created for 9 patients diagnosed with varying types of PAH (Table 1). Important parameters related to PAH which are estimated with the model are resistances and compliance in the pulmonary artery, stiffness and contractility of the right ventricle.

Computational modeling for pulmonary arterial hypertension patients, table and figures

	Gender	Age (years)	WHO-FC	mPAP (mmHg)	PVR (WU m^2)	Diagnosis
Patient 1	Female	18	II	72	33.8	PAH-CHD
Patient 2	Female	16	III	72	26.0	IPAH
Patient 3	Female	15	III	35	7.2	PAH-CHD
Patient 4	Female	12	II	68	38.0	PAH-CHD
Patient 5	Female	13	II	28	5.4	IPAH
Patient 6	Female	9	III	72	31.8	IPAH
Patient 7	Female	8	III	79	21.2	IPAH
Patient 8	Female	12	III	82	25.8	IPAH
Patient 9	Female	14	II	30	4.0	PAH-CHD

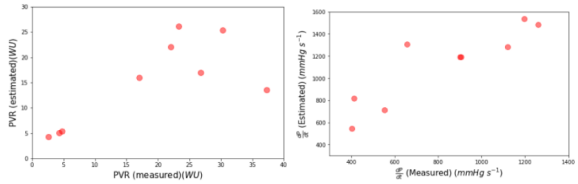
Table 1: Gender, age when MRI is taken, World Health Organization functional class (WHO-FC), mean pulmonary arterial pressure (mPAP), pulmonary vascular resistance (PVR), and the diagnosis determined within the clinic for 9 patients for which a personalized model is created. The patients are diagnosed with idiopathic pulmonary arterial hypertension (IPAH) or pulmonary arterial hypertension in combination with a congenital heart disease (PAH-CHD)



(a) Pressure right ventricle (b) Pressure right atrium

(c) Pressure pulmonary artery (d) Flow rate pulmonary artery

Figure 1: The modeled pressure in the right ventricle (a), right atrium (b), pulmonary artery (c) and modeled flow rate in the pulmonary artery (d) for one patient. The black dots with error bars are the measured pressure obtained by cardiac catheterisation or the measured flow rate during MRI.



(a) PVR estimated by the model vs the PVR measured within the clinic (b) dP/dt estimated by the model vs dP/dt determined within the clinic

Figure 2: Validation of the pulmonary vascular resistance (PVR) (a) and the dP/dt (b). The estimated dP/dt is calculated by using the contractility (force per unit area generated by the heart) estimated by the model.

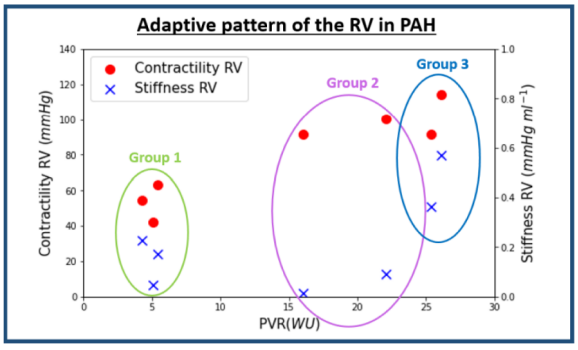


Figure 3: Adaptive pattern of the right ventricle in PAH with increasing pulmonary vascular resistance. The red dots represent the contractility of the right ventricle, and the blue crosses represent the stiffness of the right ventricle. On the horizontal axis, the modeled pulmonary vascular resistance is represented. In this plot, three groups of PAH patients can be identified. In group 2 and 3 there is an increased contractility of the right ventricle, in group 3 there is an increased stiffness of the right ventricle

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Results: In figure 1 the modeled pressure and flow rate are represented.
By comparing the model output with the data available within the clinic the model is validated (figure 2).
In figure 3 the adaptive pattern of the right ventricle in PAH is represented based on the estimated parameters for 7 patients. 3 groups can be identified depending on the resistance in the pulmonary capillaries, and RV contractility and stiffness. Furthermore, a higher resistance and lower compliance are found for the patients classified in WHO-FC III compared to WHO-FC II.
Conclusions: With the computational model, it is possible to get information about the RV adaptation (RV stiffness and contractility) within a minute which is not available with conventional measurements. These RV characteristics can be important for the risk stratification for pediatric PAH patients.

Keywords: pulmonary arterial hypertension, computational modeling, ventricular vascular coupling

PP-825

Lymphomonocytic inflammatory infiltrate with eosinophilic granulocytes in the interstitium in a surviving heart transplant recipient: A case report

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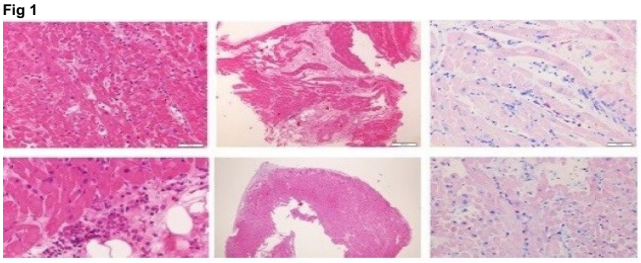
Background and Aim: Findings of eosinophilic and lymphomonocytic inflammatory infiltrates in endomyocardial biopsies (EMBs) may help in myocardial disease diagnosis identification. Eosinophilic myocarditis (EM), a rare condition, is fatal if left untreated and has rarely been described in heart transplant recipients. An extensive work up is necessary to achieve an early etiological diagnosis; however, the underlying cause remains unexplained in nearly one-third of the patients. The cornerstone of treatment is corticosteroids, comprehensive therapy and heart failure management (including advanced mechanical support for fulminant myocarditis).
Method: We have described the case of a 17-year-old heart transplant recipient who presented with a cardiogenic shock. He was admitted to our intensive care unit and treated with inotropic drugs, such as milrinone, adrenaline, vasopressin, and levosimendan; the doses of these drugs were in accordance with our internal protocol. The patient underwent cardiac catheterization, coronarography, and right ventricular endomyocardial biopsy.
Results: Endomyocardial biopsy revealed inflammatory lymphomonocytic and eosinophil granulocyte infiltrates; thus, steroid therapy was initiated, with complete recovery achieved after 15 days.
Conclusions: Performing an early differential diagnosis among eosinophilic infiltration, acute cellular rejection (ACR), and possible chemotherapeutic damage is emerging as an important challenge. To our knowledge, this is the first reported case of a lymphomonocytic inflammatory infiltration with numerous eosinophilic granulocytes in the interstitium in a surviving heart transplant recipient.

Keywords: lymphomonocytic inflammatory infiltrate, heart transplantation, endomyocardial biopsy, artero-venous extracorporeal membrane oxygenation

Fig 1



Endomyocardial biopsy showing lymphomonocytic inflammatory infiltrate

PP-826

Paediatric pulmonary arterial hypertension: an unusual course of a severe case

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Background and Aim: Pulmonary arterial hypertension (PAH) is a progressive disease that carries high morbidity and mortality. In children, the aetiology and management of PAH can be challenging.
Method: Clinical case.
Results: A 13 year old girl was referred to the Paediatric Cardiology Clinic due to two episodes of exercise related syncope. The transthoracic echocardiogram revealed severe dilation of the right-sided chambers, right ventricle hypertrophy and an estimated suprasystemic pulmonary pressures. Cardiac catheterization confirmed PAH (mean pulmonary artery pressure (PAP)of 45 mmHg; pulmonary capillary wedge pressure7 mmHg; transpulmonary pressure gradient 38 mmHg, pulmonary vascular resistance 19 WU/m2, indexed cardiac output (iCO)2.3L/min/m2) and a negative vasoreactivity test. A high resolution CT (HRCT) chest was inconclusive for pulmonary veno-occlusive disease (PVOD). The remaining workup was normal. The patient was started on sildenafil and diuretics and initially responded favourably. Bosentan was added, but was discontinued due to clinical deterioration and suspected PVOD. Two months later, she was started on epoprostenol, with a favourable response. Five months later, the condition deteriorated again and it improved after increasing the dose of epoprostenol, as well as, adding macitentan to the therapeutic regimen. Within a period of two years, the condition further declined, one episode being complicated by hemoptysis, requiring further increments in the epoprostenol dose. A repeat cardiac catherization, whilst on three vasodilatory drugs, showed no improvement of the PAP, but an iCO of 3,2 L/min/m2. A repeat HRCT chest remained inconclusive for PVOD. Due to progressive clinical worsening, she was listed for lung transplantation. At the age of 17 she underwent a double lung transplant.
Conclusions: The authors highlight the case of an adolescent girl with severe PAH of uncertain etiology, whose clinical course had been characterized by intermittent episodes of right ventricular failure with relativity clinical stability in between Despite management with triple vasodilatory drugs, no clear improvement was

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evident and she underwent a successful lung transplant. PVOD is a rare condition with a poor prognosis; lung transplant is recommended as the only curative therapy. The role of PAH targeted therapy in PVOD remains controversial.

Keywords: paediatric cardiology, arterial pulmonary hypertension, lung transplant

PP-828

Right heart failure in a child secondary to chronic airway obstruction

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Background and Aim: OBJECTIVE: to raise awareness of the importance of chronic airway obstruction in children.
BACKGROUND: Chronic airway obstruction is often ignored by pediatricians, downplaying its importance as a cause of pulmonary arterial hypertension in children.
Method: Here we describe the clinical case of a 5-year-old boy who, since he was 3 years old, has had tonsil infections, pharyngitis, repeatedly. At 5 years old, frequent bronchoobstructive symptoms with cyanosis, saturation of 87%, were managed. with salbutamol, fluticasone, ipatropium bromide with slight improvement. An echocardiogram was performed at one, two and three years of age with a report that ruled out structural heart disease. The latter was already observed with mild pulmonary hypertension of 35 mmHg. At the age of 4, he attended on several occasions with cough, fever, cyanosis, tonsillitis, respiratory difficulty, bronchoobstructive symptoms without improvement. He was hospitalized in intensive care on 2 occasions with a diagnosis of severe pneumonia. An echocardiogram was performed showing dilation of the right cavities, right ventricular dysfunction with Severe pulmonary arterial hypertension was discharged with home oxygen at night due to saturation of 85%. He attended the outpatient clinic with functional class III/IV, 65% cyanosis, with fever. The evaluation showed evidence of adenoid hypertrophy, grade IV tonsils, the airway was closed, an echocharidogram was performed with dilation of the right cavities with a z score of 3.2, and severe pulmonary hypertension. Tonsillectomy and adenoidectomy were performed, showing immediate improvement with saturation greater than 90%, with functional class I/IV. Echocardiogram one month after surgery with right chambers with z score 2.3, with mild tricuspid regurgitation.
Results: This case was a success, since with a correct diagnosis an excellent result was obtained.
Conclusions: We want to share this case since chronic airway obstruction generated right heart failure, as well as pulmonary arterial hypertension, this being reversible; however, patients with Down syndrome have a predisposition to suffer from pulmonary arterial hypertension, which could be the obstruction. Chronic airway deficiency is a determining factor in these patients, which is why it is a point of discussion to recommend early surgery.

Keywords: pulmonary hypertension, heart failure in

PP-829

High-dose angiotensin converting enzyme inhibitors and β-blocker in infantile dilated cardiomyopathy

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Background and Aim: Dilated cardiomyopathy (DCM) is characterised by ventricular dilatation and systolic dysfunction, and is the leading indication for heart transplantation in children and adults. Recently, the efficacy of high-dose angiotensin converting enzyme (ACE) inhibitors and β-blockers for heart failure has been reported; however, the optimal dosage in children, particularly infants, is unknown. Herein, we report a case series of 4 patients with infantile DCM wherein administering high-doses of ACE inhibitors and β-blockers showing effective outcomes.
Method: A retrospective review was conducted on patients with infantile DCM treated in our hospital from 2014 to 2023.
Results: There were 4 cases of infantile DCM, with onset ages ranging from 1 to 10 months. One case was Barth syndrome, and three cases were idiopathic DCM. Treatment commenced with diuretics and phosphodiesterase (PDE) III inhibitors, with the addition of catecholamines based on the severity of heart failure. Subsequently, ACE inhibitors and β-blockers were introduced with gradual increased dosage. At our hospital, the maximum dosage of cilazapril, enalapril, and carvedilol in children were 0.1 mg/kg/day, 1.0 mg/kg/day, and 1.0 mg/kg/day, respectively. High-dose ACE inhibitors and β-blockers were administered to 4 patients with infantile DCM, resulting in improved cardiac systolic function and ventricular dilatation in 3 cases. In the remaining patient, olprinone was successfully discontinued, and the patient was discharged from the hospital; however, systolic function and ventricular dilatation did not improve, and the patient died from hypovolemic shock triggered by enteritis.
Conclusions: High-dose ACE inhibitors and β-blockers may be an effective treatment option for patients with infantile DCM. When administering ACE inhibitors and β-blockers, especially at high doses, careful attention should be paid to coexisting infections and renal function.

Keywords: Dilated cardiomyopathy, Angiotensin converting enzyme inhibitor, β-blocker, infant

PP-830

The evaluation of cardiac arrythmias in the children with heart transplanation

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Background and Aim: Heart transplantation is the best option for long-term life expectancy in children with heart failure associated with cardiomyopathies and congenital heart disease and those with pathologies that cannot be corrected despite medical and surgical treatments. One of the complication in heart transplanatation patients is cardiac arrythmia. In this study, we aimed to evaluate cardiac arrythmias in our heart transplanation patients.
Method: The study included 27 children (age 0-18 years) who underwent orthotopic heart transplantation for advanced heart failure in our hospital January 2007 and January 2023. Follow-up ECHO and ECG after heart transplantation were performed at 1 week, 2 weeks, and monthly thereafter. We restrospectively evaluated the patient's files
Results: Arrhythmias were detected in 9 (33%) of the 27 patients during follow-up. Three patients developed complete atrioventricular (AV) block after transplantation. Permanent pacemakers were implanted in 3 patients. One patient developed complete AV block during acute rejection and received a temporary pacemaker. At 72 hours of follow-up, the AV block regressed and sinus rhythm resumed. Chronic sinus tachycardia was detected in 2 patients. Beta-blocker therapy was initiated in these patients because their tachycardia persisted despite eliminating all possible

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causes of sinus tachycardia. Atrial and ventricular extrasystole were each detected in 1 patient. The patient with ventricular extrasystole was found to have grade 2 rejection. A complete right bundle branch block pattern was observed in 1 patient. Except for the patient with rejection and complete AV block, none of the patients who developed arrhythmia had low ventricular ejection fraction. *Conclusions:* With this study, we observed once again that heart transplantation is not a definitive cure and that close monitoring for potential complications is essential. One of the important expected complication is cardiac arrythmias in the heart transplantation patients.

Keywords: Heart transplanation, arryhmia, children

Sports Cardiology, Physical Activity and Prevention

PP-831
Association between glycated hemoglobin and left ventricular remodeling in children with type 1 diabetes

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Background and Aim: Type 1 diabetes (T1DM) is a major risk factor for cardiovascular disease. Conflicting information exists regarding the association between the degree of glycemic control and left ventricular (LV) remodeling in children with T1DM. This study aimed to describe the frequency of the pattern of LV remodeling in children with T1DM, and its relation to glycosylated hemoglobin (HbA1c).
Method: This case-control study was conducted on 45 children with T1DM and 45 healthy, from the Department of Pediatrics, Mother and Child Institute. All participants were subjected to the full clinical examination, and investigations including biochemical dosage – HbA1c, echocardiography – LV functional and structural parameters.
Results: The results of the evaluation of the pattern of LV remodeling were distributed as follows: 6.7% (n=3) – concentric LV hypertrophy, 8.9% (n=4) – concentric LV remodeling, and 4.4% (n=2) – eccentric LV hypertrophy, 80.0% (n=36) – participants showed a normal LV geometry pattern. The correlational study between the HbA1c and the LV remodeling parameters revealed a positive correlation coefficient with interventricular septal thickness at end-diastole (mm) (r=0.5*, p=0.036), posterior wall thickness at end-diastole (mm) (r=0.5*, p=0.032), LV Mass (g) (r=0.5*, p=0.038), and LV Mass Index (r=0.5*, p=0.038).
Conclusions: Glycosylated hemoglobin appears to be one of the predictors of left ventricular remodeling in children with type 1 diabetes. The results of the study show that the increased value of the glycosylated hemoglobin is associated with a consensual and proportional increase in the values of the left ventricular remodeling parameters. Periodic echocardiographic examination is recommended for early detection of the modifications of left ventricular parameters, and initiation of the treatment if necessary.

Keywords: children, diabetes, left ventricular remodeling.

PP-832
Association between epicardial adipose tissue thickness and left ventricular parameters in children with metabolic syndrome

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Background and Aim: Epicardial adipose tissue (EAT) has a role in the development of several cardiovascular diseases through complex mechanisms - gene expression profile, pro-inflammatory and profibrotic proteome, neuromodulation, and glucose and lipid metabolism, including in the presence of metabolic syndrome (MetS). The aim of this study was to investigate the possible correlation between EAT thickness and left ventricular (LV) parameters in children with MetS.
Method: An observational analytical cohort study was conducted on 118 children, with the following criteria: the age of 10 - 18 years; and waist circumference (WC) $\geq$ 90th percentile. The diagnosis of MetS was established according to the International Diabetes Federation (IDF) criteria. EAT (by echocardiography) and other clinical-paraclinical data were analyzed.
Results: The mean value of the EAT was beyond the accepted cut-off points both, in the study group (MetS+) and control group (MetS-) (5.39 ± 0.22 mm vs. 4.97 ± 0.17 mm; $p > 0.05$); higher in males vs females (5.27 ± 0.2 mm vs. 4.97 ± 0.19 mm; $p > 0.05$); and, within the clusters, it was found to be higher in subjects with four and five positive criteria, respectively (WC - HDLc - HBP = 5.47 ± 0.44 mm, vs. WC - TG - HBP = 5.6 ± 0.84 mm, vs. WC - TG - HDLc = 4.65 ± 0.38 mm and WC - TG - HDLc - HBP = 5.77 ± 0.52 mm; F = 0.82; $p > 0.05$). A positive correlation between EAT and, interventricular septum ($r = + 0.3^*$; $p > 0.05$), left ventricular posterior wall ($r = + 0.34^*$; $p > 0.05$) and left ventricular mass ($r = + 0.3^*$; $p < 0.05$) was found. No correlation coefficient was not noted with hemodynamic, lipid metabolism, carbohydrate, and vascular remodeling parameters.
Conclusions: The increase in epicardial adipose tissue thickness probably is associated with a consensual and proportional increase of the left ventricular parameters in children with metabolic syndrome. Further studies might elucidate whether epicardial adipose tissue deserves to be included among the diagnostic factors of metabolic syndrome.

Keywords: children, epicardial adipose tissue, metabolic syndrome.

PP-833
Heart failure in long-term survivors of childhood cancer - a population-based systematic review and meta-analysis

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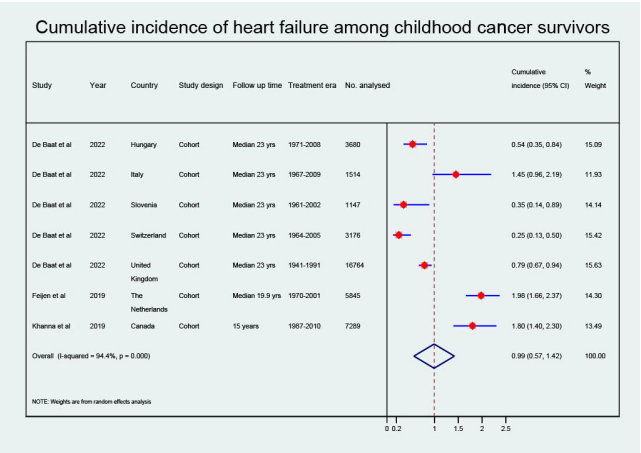
Background and Aim: The survival rate for childhood cancer has significantly improved and now exceeds 80%, resulting in a growing population of childhood cancer survivors (CCS). Among CCS, heart failure (HF) is a particularly serious non-malignant complication. We conducted a systematic review to consolidate data from population-based studies that report on the occurrence of HF in CCS who have survived for five years or more following their cancer diagnosis.
Method: We searched ten leading databases focusing on this topic from their inception up to March 14th, 2023. The process involved screening articles, extracting data, and assessing quality, all performed independently by two reviewers. A random-effects meta-analysis was conducted to summarize the findings.
Results: Out of 3,883 records screened, eight met the eligibility criteria, covering 45,659 CCS diagnosed between 1970 and 2010. These studies reported a range of cumulative HF incidences, from 0.25% to 1.98%. A meta-analysis of three of these studies yielded a cumulative incidence of 0.99% (95% CI 0.57-1.42). However, there was substantial heterogeneity among these three studies, with an I-squared statistic of 94.4% ($p < 0.001$). The overall quality assessment indicated that four studies were strong, three were moderate, and one was weak
Conclusions: The existing population-based studies on HF in CCS are limited in number and exhibit significant heterogeneity. Nevertheless, these studies confirm an elevated risk of HF in CCS. To facilitate the establishment of surveillance programs and future treatment guidelines, there is a urgent need for population-based studies that compare HF incidence in CCS to that of the general population.

Keywords: Childhood Cancer Survivors, Heart failure, Systematic review, Meta-Analysis

Cumulative Incidence



The cumulative incidence of heart failure in the different childhood cancer survivor populations shows significant heterogeneity

PP-834

PedsQL, exercise capacity, and endothelial function in pediatric fontan patients

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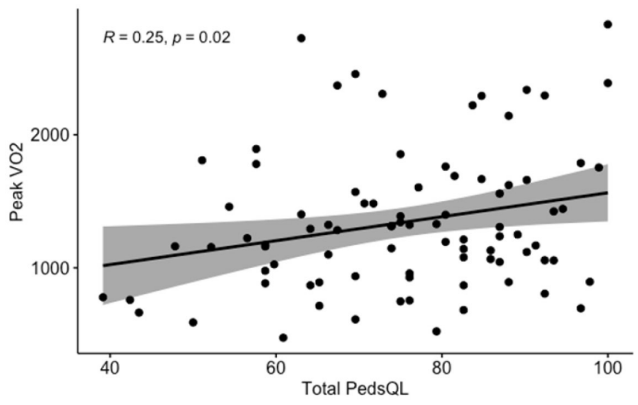
Background and Aim: Patients with Fontan circulation can experience a range of physical, motor, and/or cognitive challenges. Here, we present baseline data of pediatric Fontan patients prior to entering a controlled randomized exercise intervention (REENERGIZE FONTAN) and how their health measures relate to their Pediatric Quality of Life Inventory (PedsQL) score, a multidimensional tool assessing health-related quality of life in children and adolescents. Our hypothesis is that patients with worse exercise capacity and endothelial function will have lower PedsQL scores compared to patients with better exercise capacity and endothelial function.
Method: Fontan patients 8-19 years of age cleared to exercise are included in this randomized controlled trial that utilizes live-video conferencing to deliver a supervised exercise intervention. Patients with NYHA Class IV, recent acute illness, active protein losing enteropathy, pacemaker, and/or cognitive delay are excluded. At baseline, patients undergo a series of tests including a cardiopulmonary exercise test (peak VO2), an endothelial pulse amplitude test (reactive hyperemia index, RHI), and the 23-item PedsQL Inventory. The survey quantifies physical, emotional, social, and school functioning into 1 total and 2 summary scores pertaining to physical and psychosocial health.
Results: 86 Fontan patients (median age 13.1 years; IQR, 10.6, 15.6) completed the baseline visit. Median time from Fontan operation was 9.2 years [IQR, 7.0, 12.0]. Peak VO2 was 1343± 526 ml/min and indexed peak VO2 was 27.9±6.71 mL/kg/min. RHI and lnRHI were 1.42±0.51 and 0.31±0.32, respectively. Physical and psychosocial PedsQL summary scores were 75.7 ±17.2 and 75.3±15.6, respectively, and total scale score was 75.4±14.7. There was a mild positive correlation between peak VO2 and PedsQL total scale score (r(84)= 0.25, p=.02), and between lnRHI and PedsQL physical summary score (r(76)= 0.250, p=.03).
Conclusions: In this cohort, worse exercise capacity and endothelial function were associated with lower PedsQL scores in pediatric Fontan patients. Our RE-ENERGIZE FONTAN exercise intervention will provide further information on the impact of aerobic and strength training on the quality of life in this patient population.

Keywords: Fontan, Quality of life, Exercise capacity, PedsQL

Peak VO2 vs Total PedsQL



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PP-835

Differences in ergospirometry between fallots with severe and non-severe pulmonary regurgitation

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Background and Aim: Pulmonary regurgitation (PR) is one of the most common complications in patients undergoing for Tetralogy of Fallot (TOF) surgery. Chronic PR is known to lead to right ventricular (RV) dysfunction, symptoms such as fatigue and exercise intolerance and therefore a worse long-term prognosis.
Our aim is to compare cardiopulmonary stress test (CPET) variables: peak oxygen consumption, OUES, VE/VC02 slope, maximal workload, 02 pulse, maximal heart rate and maximal oxygen consumption at anaerobic threshold between TOF patients corrected with transpulmonary valve patch with severe PR and those with preserved pulmonary valve annulus with mild-moderate PR.
Method: Retrospective observational study was performed in a Spanish population of 41 patients with TOF aged 7-17(14,5) years, all of whom had undergone a CPET on the cycloergometer and magnetic resonance imaging (MIR) during their follow-up. They were divided into two groups according to whether they had severe PR or not. Seven CPET variables and PR fraction and RV end-diastolic volume were collected. Severe PR fraction >40% MIR was considered severe.
Results: A total of 41 patients were recruited, 20 in the preserved annulus group without severe PR and 21 in the severe PR group, without any degree of stenosis in the pulmonary bed, (these groups were comparable in age and sex). CPET variables VO2/kg, VE/VCO2 slope, OUES, maximal workload,02 pulse, maximal heart rate and maximal oxygen consumption at anaerobic threshold were compared without finding statistically significant differences in any of them. The detailed results are shown in the table (see tables below).

Image 1

	Severe Pulmonary regurgitation	Mild-moderate pulmonary regurgitation	p value
Number of patients	21	20	
Mean age (years)	14,41	15,67	
Percentage of female	23,8%	40%	
Mean regurgitation fraction (MRI)	47,02%	16,35%	P<0,001
Mean right ventricular end-diastolic volume standardised by body surface area (MRI)	144,51 ml/m2	105,02 ml/m2	P<0,001
Mean VO2 peak consumption per kilo	30,70 ml/kg/min	31,03ml/kg/min	P>0,05
VE/VC02 slope	30,49	27,63	P>0,05
OUES	1746,67	1780,00	P>0,05
Mean maximum workload(Watt)	118,00	129,65	P>0,05
Mean 02 pulse	9,98	10,20	P>0,05
Mean maximum heart rate	171,52	176,50	P>0,05
Mean V02 consumption at anaerobic threshold (VT1)	17,28 ml/kg/min	18,75ml/kg/min	P>0,05

Conclusions: We found no statistically significant differences in oxygen consumption between the two groups, which may be due to the fact that in the paediatric age group, despite the clear RV dilatation associated with PR, it is rare to observe an overt deterioration in RV function. On the other hand, the lack of a statistically significant difference in VE/VC02 overlap may be due to the fact that this variable is more affected in situations of stenosis in the pulmonary tree than in situations of pulmonary insufficiency.
Important evolutionary follow-up with multiple diagnostic tests to monitor both potential decline in functional capacity and RV function throughout life.

Keywords: Fallot, Pulmonary regurgitation and Cardiopulmonary stress test

PP-836

Oxygen consumption in congenital heart disease

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Background and Aim: Cardiorespiratory fitness (CRF) is usually reduced in people with CHD, and often deteriorates more rapidly than in healthy people. Cardiopulmonary stress test (CPET) is used to measure functional capacity. CRF is known to be prognostic for future mortality and morbidity.
Objectives: Our goal is to compare the functional capacity of different congenital heart defects in Spanish children.
Method: This is a retrospective observational study in a Spanish paediatric population with congenital heart disease between 8-14 years of age. We analyzed 423 patients with 551 test from the national paediatric registry of CPET on the cycloergometro in Spain was used from 2017 to the present. We selected maximum tests and compared percentage of maximum peak oxygen consumption achieved (V02 peak %) in each pathological group. Due to small sample sizes in some groups and the presence of extreme values, we use the median as a measure of the centralisation of values.
Results: In our study, we found that the congenital heart defects with the best peak oxygen consumption were pretricuspid shunts, followed by corrected alkapas and non-interventional ebsteins with completely normal predicted consumption compared to the healthy population. On the other hand, the group with the worst peak oxygen consumption was the Glenns, followed by Fontan and Truncus patients whose median predicted consumption was far away from the expected consumption.
In the table below we add the rest of the heart diseases studied with the median peak and predicted consumption.
Conclusions: We provide an overview of how each congenital heart disease affects oxygen consumption. CPET is a very useful tool for monitoring these patients and allows us to follow their functional capacity in great detail. We recognise the limitations of the study (anatomical heterogeneity, small size sample). For these reasons, the database needs to be expanded in order to obtain more consistent information and to be able to determine the predicted V02 for each congenital heart disease based on age and sex.

Keywords: Cardiopulmonary stress test, Congenital heart disease and Oxygen peak consumption

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Image 1

Congenital heart disease	N of patients	Mean age	Median V02 peak /kg.	Median V02 Predicted
Alcapa	N=3 (3 test)	15,3	33,10 ml/kg/min	85,60%
Ebstein	N=11 (14 test)	12,4	27,31 ml/kg/min	84,00%
AP + CIV	N=7 (10 test)	14,1	25,29 ml/kg/min	69,00%
APSI	N=7 (11 test)	14,0	28,32ml/kg/min	74,61%
Glenn	N=7 (13 test)	15,1	16,44 ml/kg/min	40,21%
Canal AV	N=6 (12 test)	12,7	31,96 ml/kg/min	80,55%
Pretricuspid shunts (CIA, CIA OP y DVPAP)	N= 22 (27 test)	12,2	34,19 ml/kg/min	87,45%
DVAPT	N=5 (5 test)	12,4	30,30 ml/kg/min	68,72%
Fallots	N=103 (132 test)	13,7	28,49 ml/kg/min	75,49%
Postricuspid shunts(CIVs + Ductus)	N=37 (45 test)	12,3	34,15 ml/kg/min	80,46%
Aortic coarctation	N=28 (32 test)	13,1	33,84 ml/kg/min	80,56%
DTGA	N=65 (95 test)	13,1	33,29 ml/kg/min	76,98%
LTGA (without the double switch)	N=8 (12 test)	13,9	28,41ml/kg/min	73,36%
Aortic stenosis	N= 46 (63 test)	12,5	30,03 ml/kg/min	73,61%
Ross	N=6 (8 test)	11,6	31,63 ml/kg/min	71,24%
Aortic regurgitation	N=10 (12 test)	13,2	34,6ml/kg/min	74,67%
Truncus	N=7 (8 test)	13,0	29,9 ml/kg/min	63,06%
Non-valvuloplasty pulmonary stenosis	N=19 (20 test)	11,9	29,33ml/kg/min	79,69%
Fontan	N=26 (32 test)	14,4	26,62ml/kg/min	59,79%

PP-837

Exercise capacity in children with moderate to severe pulmonary regurgitation after repair of tetralogy of fallot

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Background and Aim: Tetralogy of Fallot is the most common form of cyanotic congenital heart disease and accounts for 10% of all forms of congenital heart disease. Complete repair of this condition can lead to progressive pulmonary regurgitation which has been thought to contribute significantly to exercise intolerance in children. Objective assessment of exercise capacity with a maximal cardiopulmonary exercise testing can provide valuable prognostic information.
Method: Children were tested on a modified treadmill protocol keeping the gradient constant (10%) and ramped increases in speed. Breath to breath analysis was performed with a Medraphics metabolic cart connected to the treadmill.
Results: Thirty five children (21 females, 14 males, 9 – 17 years) with moderate to severe pulmonary regurgitation were tested 12.68 (7.83 – 16.83) years after full repair of their Tetralogy of Fallot. The mean height was 160.44 cm (range 127.6 – 182), mean weight was 54.27kg (range 24.4 – 108) and mean body surface area was 1.55 m2 (range 0.94 – 2.29). The mean exercise time was 8.62 minutes (range 5 – 11) with maximum heart rate achieved mean was 88.45% of predicted (range 71 – 99). None of them had any arrhythmias during the test or during recovery. On the Borg scale of patient reported symptoms at end exercise mean rate of perceived exertion was 6 (range 3 – 10) and mean shortness of breath was 6 (range 3 – 10). Test Parameter Patients (n=35) Predicted P Value VO2 AT 23.28±4.21 26.73±2.44 <0.001 VO2max 35.7±7.19 44.59±4.11 <0.001 VE/VO2 33.65±6.15 37.26±4.01 0.008 VE/VCO2 31.02±5.16 30.88±3.25 0.9
Conclusions: Children with moderate to severe pulmonary regurgitation after repair of Tetralogy of Fallot show significant reduction in submaximal and maximal exercise capacity despite demonstrating a normal ventilatory response to exercise.

Keywords: Cardiopulmonary exercise test, Tetralogy of Fallot, Pulmonary regurgitation, Oxygen uptake efficiency slope

Cardiovascular Morphology

PP-838

A rare case of double outlet right atrium with atrio-ventricular septal defect

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Background and Aim: We describe a newborn baby with a very unusual congenital cardiac malformation and our initial clinical management.
NG was born on 2/9/2023 at 38 weeks of gestation by SVD with Apgars 9 and 9. His birth weight 2760 grams. On newborn check he was found to have preductal saturations of 92 – 94% and post-ductal saturations of 94 – 96%. On examination he had no dysmorphic features, no respiratory distress, his femoral pulses were palpable and he had a soft systolic murmur over his precordium. There was no family history and his parents are non consanguineous.
Method: His echocardiogram showed normal situs, pulsatile abdominal aorta, apex was in the midline, pulmonary veins were draining into a left sided atrium and systemic veins and coronary sinus draining into a right sided atrium. He had a common atrio-ventricular valve with a small ventricular septal defect, good sized left and smaller right ventricular chambers, normally arranged great arteries and unobstructed aortic arch. His atrial septum was deviated leftward extending from the superior vena cava to the left atrioventricular groove excluding the left atrial mass from the ventricular mass. There was a small restrictive atrial septal defect with flow from left to right, dilated pulmonary veins. (Images 1 and 2)
Results: Balloon atrial septostomy was performed on 19/9/2023 with a 13.5 mm Z-MED balloon catheter with creation of an unrestrictive atrial septal defect. (Image 3)
Conclusions: Double outlet right atrium is a very rare congenital cardiac malformation. This happens due to deficient rotation of the left ventricular mass from right to left very early in embryogenesis. The baby's clinical condition would have been aggravated if Dinoprostone was given as the standard treatment of cyanosis if echocardiography had not been performed. Good understanding of the anatomy and timely management is needed for this condition.

Keywords: Double outlet right atrium, cor triatriatum dextra, common atrioventricular valve, absent left atrioventricular connection

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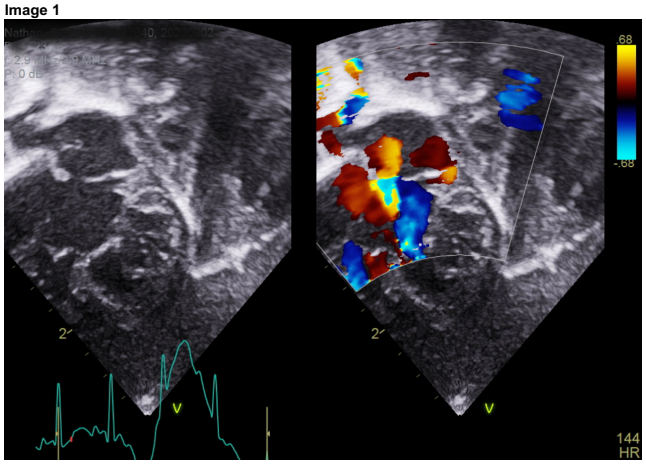
Image 1



4 Chamber view showing common atrioventricular valve, malaligned atrial septum walling off the left atrial cavity with a small restrictive atrial septal defect, double outlet from the right atrium

General Cardiology

PP-839

Cardiovascular response to graded exercise challenge in children with moderate to severe pulmonary regurgitation after repair of tetralogy of fallot

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Background and Aim: Activities of daily living require an appropriate cardiovascular response to exercise. Children require a level of cardiovascular fitness to enable them to participate in all aspects of school life. Children with repaired congenital heart disease need to enjoy the same activities as their peers. Tetralogy of Fallot (TOF) is the most common form of cyanotic congenital heart disease (CHD) and complete repair of this condition can lead to progressive pulmonary regurgitation which has been thought to contribute significantly to exercise intolerance in children. Objective assessment of cardiovascular response with a graded cardiopulmonary exercise testing can provide valuable information to enable proper management
Method: Children were tested on a modified treadmill protocol keeping the gradient constant (10%) and ramped increases in speed. Breath to breath analysis was performed with a Medraphics metabolic cart connected to the treadmill.
Results: Thirty five children (21 females, 14 males, 9 – 17 years) with moderate to severe pulmonary regurgitation were tested 12.68 (7.83 – 16.83) years after full repair of their Tetralogy of Fallot. Twenty four age matched normal children (17 males, 7 females) were tested as control group. None of them had any arrhythmias during the test or during recovery. Cardiac output was calculated from oxygen consumption with a standard formula (reference 2). Statistical comparison was made with the unpaired t-test and significance was accepted for $p \leq 0.01$. Table 1 (attached)
Conclusions: Children with moderate to severe pulmonary regurgitation after repair of Tetralogy of Fallot show appropriate increase in their cardiovascular response to exercise compared to normal children. However they demonstrated a significant reduction in time for which they can exercise. This suggests that providing exercise training may be an important intervention to improve their quality of childhood experience.

Keywords: Tetralogy of Fallot, Cardiac Output, Stroke Volume, Chronotropic Index, Exercise

Table 1

Table 1. Comparison of cardiovascular response to graded exercise challenge in children with repaired Tetralogy of Fallot to normal controls

Test Parameter	Patients (n=35)	Controls (n=24)	P Value
Height mean (range)	160.44 cm (127.6 – 182)	156.12 (127.2 – 180.6)	NS
Weight mean (range)	54.27kg (24.4 – 108)	45.75 (24.35 – 64)	NS
BSA mean (range)	1.55 m^2 (0.94 – 2.29)	1.42 (0.94 – 1.79)	NS
Exercise time mean (range)	8.62±1.45 (5 – 11)	10.66±1.76 (8 – 14),	>0.001
RPE	6.25±1.96 (3 - 10) (34)	6.45±1.26 (5 – 10) (22)	NS
SOB	6.1±2 (3 – 10) (30)	6.26±1.16 (4 – 8) (15)	NS
Mean Heart Rate at rest (range)	88.28±13.36 (66 – 120)	85.04±16.7 (49 – 114) (24)	NS
Mean Heart Rate atVO2 max (range)	182.05±12.74 (150 – 203)	186.33±15.71 (153 – 203) (24)	NS
Chronotropic index (bpm/L)	64.37±15.45 (36.97 – 115.08)	58.81±19.63 (50.94 – 66.67) (24)	NS
Stroke volume mls (rest)	44.3±17.11 (12.2 – 96.2)	44.97±23.64 (35.5 – 54.44)	0.8
Stroke volume mls (AT)	81.76±23.34 (39.69 – 132.24)(34)	80.61±25.73 (70.3 – 90.92)	0.8
Stroke volume mls (VO2max)	75.72±22.57 (37.3 – 137.29) (34)	73.25±20.34 (65.1 – 81.4)	0.6
Stroke Volume at AT/ Rest	2.02±0.54 (1.34 – 4.18) (34)	2.28±1.75 (1.58 – 2.98)	0.4
Stroke Volume Max/ Rest	1.9±0.69 (1.22 – 4.86) (34)	2.11 ± 1.6 (1.47 – 2.75)	0.5
Cardiac output Litres (rest)	3.88±1.22 (1.33 – 6.5)	3.85±1.61 (3.21 – 4.49)	0.9
Cardiac output Litres (AT)	11.77±3.54 (5.2 – 22.35)	11.21±3 (10.01 – 12.41)	0.5
Cardiac output Litres (max)	14.01±4.14 (6.65 – 25.4)	14.05±3.85 (12.51 – 15.59)	0.9
Cardiac Output At AT/ Rest	3.15±0.78 (2.09 – 5.75)	3.55±2.54 (2.54 – 4.56)	0.3
Cardiac Output Maximum/ Rest	3.77±0.96 (2.29 – 6.69)	4.52±3.51 (3.12 – 5.92)	0.2

Table is an integral part of results of research

Sports Cardiology, Physical Activity and Prevention

PP-840

Handgrip-strength and international physical activity questionnaire – surrogate markers for cardiovascular fitness

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Background and Aim: Usually, Cardiovascular fitness is tested with Cardiopulmonary Exercise Testing (CPET). This validated method is time consuming and cost intensive. In daily practice and for identification of high cardiovascular (CV) risk groups on a large scale, e.g. at school or university, cost efficient and easy to use tests are needed. Handgrip-strength (HGS) and the self-reported physical activity using the International Physical Activity Questionnaire (IPAQ) could serve to identify the CV fitness. Both are known predictors of CV health and mortality. This study examines the association of HGS and IPAC to the objective CPET measurement of VO2peak in a healthy student cohort at the Technical University of Munich, Germany. The project is part of TUM4Health, a university-based health promotion program (Ethical Board number 379/19 S-SR).
Method: From 2017 to 2023, a sub cohort of 389 Students participating in the TUM4Health program, which offers a medical check-up and motor performance test, this includes the evaluation

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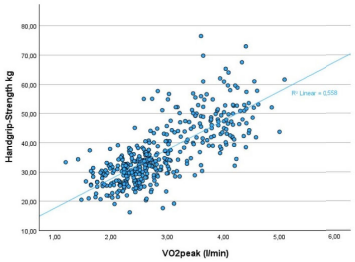
of anthropometric data, the IPAQ, a CPET on a treadmill and a digital HGS Test. The students were aged 25 +/- 4 years, and 63% were female; students with health issues were excluded from this study.
Results: HGS (r =.747, p <.001, n = 389), and IPAQ (r =.309, p <.001, n = 303) showed a positive association with VO2peak. Weight, size, and sex of the participants were strongly associated VO2peak and HGS; therefore, a linear regression model including these covariates was applied: the positive association with VO2peak was still significant for HGS (T=8.912, p=0.023) and IPAQ (T=2.430, p=0.016). In the linear regression model the R^2 value of HGS to VO2peak was 56% and R^2=10% in the case of the IPAQ.
Conclusions: Both HGS and IPAQ were significantly associated with VO2peak, but HGS seems to be the more predictive surrogate marker for CV fitness. Thus, HGS is a cost-efficient way to estimate the cardiorespiratory fitness when CPET measurements are not available or too expensive.
For future applications HGS could be used as predictor variable in the emerging field of machine learning (AI) for maximal oxygen uptake calculations. A high associated surrogate marker like HGS could optimize the performance.

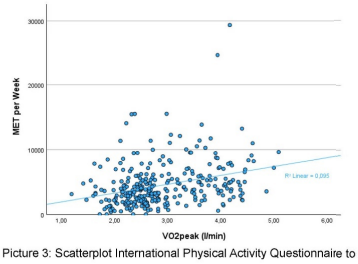
Keywords: Cardiovascular, fitness, IPAQ, handgripstrength, prevention, CPET

Graphs

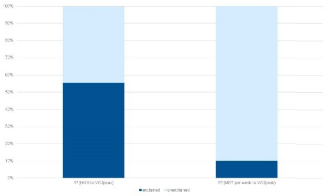
Chair of Preventive Pediatrics
TUM School of Medicine and Health
Technical University of Munich



Picture 2: Scatterplot Handgrip-Strength to VO2peak



Picture 3: Scatterplot International Physical Activity Questionnaire to VO2peak



Picture 4: R² of Handgrip-Strength and MET per week to VO2peak

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Technical University of Munich

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Sport & Exercise Science

Supervisor
Prof. Dr. Renate Oberhoffer-Fritz

PP-841

A single-center 10-year retrospective review of maximal exercise capacity in children with different types of congenital heart disease

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Background and Aim: In the last decades, children with congenital heart disease (CHD) demonstrate lower mortality and morbidity due to improved surgical, interventional and medical treatment options. Physical functioning evaluated through cardiopulmonary exercise testing (CPET) is an important factor of health and quality of life. This study evaluates maximal exercise performance in different disease groups with CHD in a large cohort of patients.
Method: Data from 836 CPET performed in children between 6-16 years with different types of CHD over a period of ten years were reviewed and compared with a control group of 294 healthy children. Maximal heart rate (HRmax) in bpm and maximal oxygen consumption (VO2max) and maximal load (Loadmax) expressed as % of the predicted value were evaluated. Statistical analysis, using Student T-test and Mann-Whitney U test as appropriate, was used to compare the different subgroups with the control group.
Results: Children with CHD had lower exercise performance compared to the healthy control group (VO2max=98.4±14.5%; Loadmax=92.8±17.1%; HRmax=189.7.9±10.0bpm). Patients with Pulmonary Atresia with intact septum (VO2max=66.6 ±18.0%; Loadmax=59.8±16.8%; HRmax=164.9±24.7bpm), Univentricular heart (VO2max=70.9±18.1%; Loadmax=60.1 ±12.7%; HRmax=163.8±21.2bpm), Ebstein malformation (VO2max=76.5±20.2%; Loadmax=73.7±15.5%; HRmax= 186.4±10.2bpm), Double Outlet Right Ventricle (VO2max= 76.7±16.8%; Loadmax=73.1±13.4%; HRmax=178.4±14.8bpm) and Tetralogy of Fallot (VO2max=76.5±20.2%; Loadmax=73.7 ±15.5%; HRmax=186.4±10.2bpm) have the worst exercise performance of all patient groups. Children with Atrial Septal Defect (VO2max=91.3±22.7%; Loadmax=81.8±18.5%; HRmax=186.5 ±10.9bpm), Ventricular Septal Defect (VO2max=90.4±17.5%; Loadmax=82.5±15.9%; HRmax=176.0±23.6bpm), Atrio-ventricular Septal Defect (VO2max=86.9±19.9%; Loadmax=81.6 ±21.1%; HRmax=172±25.1bpm), Transposition of the Great Arteries (VO2max=91.5±20.2%; Loadmax=80.0±16.6%; HRmax=175.8±18.9bpm) or Coarctation (VO2max=90.8 ±18.4%; Loadmax=80.6±18.6%; HRmax=181.7±18.2bpm) had better CPET results, although the results were still lower compared to healthy controls.
Conclusions: As children with CHD have lower exercise performance it is important to evaluate their exercise capacity during the follow-up at young age without exception. Children with Pulmonary Atresia with intact septum, Univentricular Heart, Ebstein malformation, Double Outlet Right Ventricle and Tetralogy of Fallot perform the worst and need specific attention in the evaluation of their physical fitness. When appropriate, a rehabilitation program needs to be proposed, especially in these patient groups with the lowest exercise performance.

Keywords: maximal exercise performance, physical fitness, congenital heart disease

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PP-842

Kijani app to promote physical activity in children and adolescents – preliminary results of a mixed method evaluation

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Background and Aim: Digital approaches have the potential to make activity promotion for children and adolescents attractive and age-appropriate. KIJANI is a mobile application aiming to promote safe outdoor physical activity (PA) in children and adolescents via gamification and augmented reality. The aim of this work is the conception and development of KIJANI as well as its evaluation through a mixed-method approach.
Method: The KIJANI app is based on the concept that virtual coins can be earned through PA e.g. by daily steps. With these coins, in turn, blocks can be bought in the app, which can be used to create virtual buildings and integrate them into the player's real-world environment via augmented reality. KIJANI can be played at predefined outdoor play locations that were comprehensively identified as safe, child-friendly, and attractive for PA by the target group in a partner project. To evaluate the user experience and obtain feedback from the target group, a mixed-methods approach was used. After playing KIJANI in groups of three for 25 minutes, participants took part in one-on-one semi-structured interviews and completed the user experience questionnaire (UEQ).
[ranging from -3 to +3; >0.8 indicates positive evaluation; 1-2 indicates excellent evaluation]. All interviews were audio recorded, transcribed verbatim and evaluated based on Mayring's content analysis.
Results: So far, 17 adolescents (12.5 ± 1.8 years, 5 girls) participated in the study. Overall, feedback on KIJANI was good and participants liked the game concept especially the opportunity of playing KIJANI in a group. Various categories concerning improvements of KIJANI were identified, including more variety of blocks, problems with accurate GPS location, as well as precise handling of the blocks. The average UEQ scale was (mean ± SD): attractiveness (1.96 ± 0.86), perspicuity (2.06 ± 0.68), efficiency (0.94 ± 1.05), dependability, (1.34 ± 0.86), stimulation (1.44 ± 1.37), and novelty (1.27 ± 1.56).
Conclusions: Results from this evaluation will be implemented in the KIJANI app before the evaluation will proceed in the form of a two-arm randomized controlled trial in which the effectiveness of KIJANI will be assessed via objectively measured PA.

Keywords: activity promotion, digital health, gamification

PP-843

Exercise-based cardiac rehabilitation for children and adolescents with long QT syndrome: rythmo'fit program

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Background and Aim: Children with long QT syndrome (LQTS) have impaired physical fitness and quality of life. Despite the potential benefits of exercise-based cardiac rehabilitation in pediatric cardiology, such interventions have not been evaluated in pediatric LQTS. This study aimed to assess the safety, feasibility, and benefits of a tailored exercise-based cardiac rehabilitation program in children with LQTS.
Method: Eight participants, aged between 6 to 18 years old, with positive LQTS-related genotype mutations and impaired cardiorespiratory fitness, were prospectively enrolled in a 12-week center-based cardiac rehabilitation program. This program included supervised group sessions of exercise training (aerobic, resistance training, and outdoor activities for 60-75 minutes per session) and patient education (30 minutes with a specialist nurse). The primary outcomes were safety (cardiac and non-cardiac events) and feasibility (recruitment and retention rates, adherence, and acceptability). We also investigated the impact of the program on cardiorespiratory and muscle fitness, health-related quality of life (HRQoL), physical activity, and clinical outcomes. Qualitative feedback from parents, children, and professionals involved was reported during an interview held at the end of the program.
Results: Safety was excellent as no cardiac event occurred during the 12-week intervention. The feasibility was deemed satisfactory with retention and adherence rates of 88%, while the recruitment rate remained low at 42%. Overall, participants, parents, and professionals reported being highly satisfied with the intervention program, despite experiencing some transportation- and school-related problems. The intervention led to a significant increase in ventilatory anaerobic threshold (21.7±5.2 vs. 28.7±5.1 mL/Kg/min, P=0.01, effect size= 0.89), grip strength (17.98±5.33 Kg vs. 19.98±4.67 Kg, P=0.02, effect size= 0.90), lower limb explosive strength (142±36.5 cm vs. 148±24 cm, P=0.02, effect size= 0.90) and proxy-reported physical HRQoL (65.6±9.75 vs. 84.4±20.35, P=0.03, effect size= 0.87).
Conclusions: The 12-week supervised and individualized center-based cardiac rehabilitation RYTHMO-FIT program was safe and feasible for children with LQTS. Preliminary results from this pilot study suggested the effectiveness of the intervention, in terms of physical capacity, muscle fitness and HRQoL. Further randomized controlled trials remain necessary to increase the level of evidence.

Keywords: Long QT syndrome, Exercise, Cardiac rehabilitation, Children, Adolescents, Pilot study

PP-844

The effect of exercise training in patients with single ventricle physiology: A systematic review & meta-analysis

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Background and Aim: Patients with single-ventricle physiology are usually treated with three palliative operations, eventually creating

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Fontan circulation. Over the past few decades, these procedures have improved significantly. Nevertheless, Fontan patients' exercise capacity and overall quality of life remain limited. Exercise training programs appear promising for addressing these challenges in postoperative management. However, the practical implementation of these programs and adherence of Fontan patients to exercise guidelines still need to be improved. Accordingly, this systematic review and meta-analysis aimed to elucidate the safety, efficacy, and optimal modalities of exercise interventions in Fontan patients.
Method: On 24 August 2023 a systematic search of PubMed, Scopus, Web of Science, and Cochrane Library was performed. Rayyan QCRI was used to screen the studies for eligibility based on title, abstract, and full text, following pre-set exclusion criteria. The quality of the included studies was assessed using the Cochrane Collaboration's RoB Tool 2 and Strengthening the Reporting of Observational Studies in Epidemiology checklist. A meta-analysis was conducted using a continuous random-effects model, focusing on mean and standardised mean differences to evaluate the efficacy of different cardiac rehabilitation interventions and settings.
Results: A total of 26 studies (7 RCTs, 19 cohort studies) describing 22 individual cohorts were included. Aerobic exercise training (AET) was investigated in 18 studies, lower-extremity focused exercise (resistance training) in 11 studies, and inspiratory muscle training (IMT) in 6 studies. The overall pooled effect of cardiac rehabilitation interventions demonstrated a significant improvement in VO2, indicating an enhanced exercise capacity. Either Home-based and supervised rehabilitation programs have shown a significant positive effect.
Conclusions: Substantial variation in exercise training programs was observed. Although the types of training differed considerably, exercise training seems promising for addressing challenges in postoperative management. Exercise training might enhance exercise capacity, cardiac biomarkers, lung function, and lower limb muscle strength, ultimately contributing to improved quality of life. Further research is needed to refine exercise protocols, assess long-term effects, fully elucidate the underlying mechanisms, and test exercise programs in Fontan patients with severe clinical conditions.

Keywords: Fontan, cardiac rehabilitation

PP-845
A practical approach to evaluate cardiovascular function of children and adolescence VIA stress exercise echocardiography
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Background and Aim: Identifying young and asymptomatic athletes with early-stage cardiomyopathy remains a major challenge in sports cardiology. While resting echocardiography and cardiopulmonary exercise-test (CPET) are easily available standard procedures, diagnostic yield in early disease stages is poor. In adults with cardiac disease, exercise-stress echocardiography (ESE) is an established tool for identifying cardiovascular dysfunction masked at rest. Therefore, a combination of CPET and simultaneous ESE (CPET-SE) has the potential to differentiate between training-associated remodeling and cardiomyopathy when assessing adolescent athletes for early-stage cardiac disease. However, CPET-SE has not been established in pediatric cohorts so far.
Method: Fifty-five healthy adolescent male athletes performed CPET-SE in a semi-supine position on a recumbent cycle-ergometer using a step protocol, and echocardiography was evaluated for imaging quality. Workload increments of 25 or 50W were used, adapted to body size and physical fitness of the subjects. For each step, assessment of cardiac function included 2-dimensional strain analysis via speckle-tracking, peak systolic mitral-annular velocity, early diastolic velocity of mitral-annular motion, and early diastolic velocity of mitral inflow.
In 42 of the subjects, standard CPET (S-CPET) data were obtained at their next annual visit using our standard ramp protocol during upright cycle ergometry.
Results: Cardiac function analysis was feasible in 70% of subjects at moderate effort (60% of max workload) and 50% at submaximal effort (80% of max workload). Imaging quality was lowest in apical segments, especially in 2- and 3-chamber views. In CPET-SE, heart rate, respiratory exchange rate (RER) and aerobic capacity (peakVO2) at maximal effort did not differ significantly from results of S-CPET, although maximum workload in CPET-SE was significantly lower than in S-CPET. Examination time for this CPET-SE protocol was significantly higher than S-CPET (29 vs.18 minutes, $p<0.001$).
Conclusions: Cardiac strain imaging in children and adolescence under exercise-stress during CPET was feasible at moderate workloads using our approach, but insufficient at high workload and examination time was very long. For future research, we propose a modified approach based on a ramp exercise protocol and focusing on improved image quality rather than maximal exertion.

Keywords: cardiac function, exercise tolerance, stress echocardiography, cardiopulmonary exercise test, return to sport

PP-846
Stress tolerance and relaxation ability of children with congenital heart diseases and healthy counterparts during a winter camp - a pilot study
Nicola Stöcker, Florian Katzl, Renate Oberhoffer Fritz and Christina Sitzberger
Department Health and Sport Sciences, TUM School of Medicine and Health (MH)

Background and Aim: Children and adolescents are frequently exposed to stressors from a young age. An inadequate recovery from these stressors can lead to distress and an increased risk of chronic stress. This is especially pertinent for children and adolescents living with chronic diseases, as they are at a heightened risk of experiencing stress. That can result in prolonged physiological activation, compromising the body's ability for self-regulation and impeding the return to a state of relaxation during rest periods, ultimately disrupting inner homeostasis and potentially contributing to further health issues. Heart rate variability (HRV), which reflects the autonomic nervous system's balance, offers insight into one's recovery capacity.
Method: This Pilot study was conducted as part of the KidsTUMove wintercamp 2023 in South-Tirol, Italy, involving vigorous physical activity. The aim was to examine HRV in both chronically ill children and adolescents and their healthy counterparts. HRV was assessed using the Low Frequency to High Frequency ratio (LF/HF), which indicates the sympathovagal balance. Three Measurements were conducted during the week by using the Firstbeat©-System. Sample sizes for the first and second measurements were both n=13, while n=5 were included for the third measurement. HRV was recorded over a five-minute resting period.

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Results: A total of 13 children (12.26±4.6 years) were included, 7 with chronic diseases including congenital heart disease. No statistically significant differences in HRV within healthy and chronical ill children in all three measures (HzDay1 1.18±0.60/1.06 ±0.37, p=0.73; HzDay2 1.29±0.32/1.54±1.07, p=0.94; HzDay3 1.30±0 /2.31±1.0, p=0.4) were revealed. No age- (p=0.94/p=0.46/p=0.25) or gender-specific (p=0.45/p=0.84/ p=0.4) disparities in LF/HF were identified.
Conclusions: Although differences in HRV between chronically ill and healthy children and adolescents are known to exist, this study could not detect these differences due to small sample sizes. Comparative studies often incorporate additional HRV parameters, such as RMSSD, SDANN, or SDNN, to provide a more comprehensive assessment of HRV. Additionally, it's essential to consider that participants in the study experienced increased physical and social stress during the camp week, which could have influenced the results. Future studies should prioritize improved study quality and a larger number of participants to obtain more meaningful and reliable results.

Keywords: chronic diseases, Heart rate variability (HRV), recovery capacity, children with congenital heart diseases

PP-847

Is vigorous physical activity a good predictor of exercise capacity and quality of life among adolescents with congenital heart disease?

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Background and Aim: Moderate-to-vigorous PA (MVPA) has positive associations with many health biomarkers; however little is known about the interactions between vigorous physical activity (VPA), sedentary time (ST), exercise capacity (peak VO_2) and health-related quality of life (HRQoL) in adolescents with CHD. Furthermore, there is a growing need to understand the role of VPA compared to MPA in health outcomes. Therefore, the aim of the study was to assess the associations between device-measured MVPA, VPA, ST with peak VO_2 and HRQoL among adolescents with CHD.
Method: Twenty-eight adolescents with CHD (mean age 14.4 ±1.7y, 57% male) were recruited. PA (MVPA and VPA) and ST were measured using accelerometer for 7-consecutive days. Data were analysed if wear time was ≥16 hours/day and ≥4 days using age-specific cut-offs. Peak VO2 via an incremental cycle ramp test and HRQoL using a self-reported Teens (12-18 years) Paediatric Quality of Life Inventory Questionnaire were assessed. Linear and multiple regression examined the association between all variables.
Results: Mean MVPA, VPA and ST were 57.0 ±28.2, 4.0 ±3.3 and 763.0 ±105.4 min·d-1, respectively. Mean peak VO2 was 31.6 mL·kg-1·min-1 and HRQoL overall score was 72/100. MVPA and VPA were positively associated with peak VO2 with, β=0.2, P=0.001, and β = 1.8, P<0.001, accounting for 33% and 44% of variation, respectively. MVPA and VPA demonstrated a positive association with overall HRQoL (β=0.2, P=0.04, and β=1.43, P=0.04, respectively) accounting for 15.7% and 15.8% of variability, respectively. Only VPA was associated with predicting the HRQoL sub-components, physical health score by 25.9% (β = 2.4, P = 0.007). ST was negatively associated with peak VO2 (β=-0.02, P=0.24) and HRQoL (β=-0.02, P=0.43). Multiple regression between VPA and HRQoL revealed that only VPA was a significant predictor of peak VO2 (β=1.57, P<0.01), accounting for 49% of the variance, (P<0.001). Between HRQoL and peak VO2, only peak VO2 significantly predicts VPA (β=0.23, P<0.01), accounting for 46% of the variability. Meanwhile, VPA and peak VO2 did not significantly predict HRQoL (R2=0.22, P=0.052).
Conclusions: Both VPA and MVPA independently predict exercise capacity and HRQoL, with VPA showing a stronger association. Incorporating VPA into personalised interventions may enhance exercise capacity and HRQoL significantly.

Keywords: VPA, MVPA, congenital heart disease, health-related quality of life, exercise capacity

PP-848

24 hour ambulatory blood pressure monitoring in young adults conceived through assisted reproductive technologies

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Background and Aim: Since 1978, approximately 8 million individuals have been conceived through assisted reproductive technologies (ART). The literature suggests that ART offspring might be at increased cardiovascular risk. However, data on vascular function of young ART adults is scarce and ambiguous. This study aimed to assess vascular function of young ART adults through 24h ambulatory blood pressure monitoring (ABPM).
Method: Young ART adults as well as age- and sex-matched spontaneously conceived peers were recruited by our center. 24h ABPM was conducted in all study participants. Blood pressure readings were taken every 15 minutes during the day and every 30 minutes during the night. 24h systolic/diastolic blood pressure (SBP/DBP) were assessed. Moreover, participants were asked to note sleep- and wake times to measure nocturnal blood pressure

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decrease. Subjects were included in the final analysis if ≥50 measurements were performed within 24h and ≥10 measurements were recorded during nighttime. The unpaired t-test was used for normally distributed data and the Mann-Whitney-U-test for non-normally distributed data.
Results: In total, 15 ART subjects (IVF, n=9; ICSI, n=6) and 20 spontaneously conceived controls were included in the final analysis. Both groups did not differ significantly in age (21.11±2.28 years vs. 21.69±1.95 years, p=0.427) and sex. 24h SBP (118.62±9.74 mmHg vs. 116.74±6.47 mmHg, p=0.525), 24h DBP (70.99 ±7.62 mmHg vs. 71.21±7.20 mmHg, p=0.931) as well as nocturnal blood pressure decrease did not display significant differences between ART subjects and peers.
Conclusions: Unlike previous publications, this study does not indicate a significantly higher blood pressure measured by 24h ABPM in young ART adults compared to spontaneously conceived peers. In the future, multi-centric studies are required to validate these preliminary results.

Keywords: Assisted Reproductive Technology, blood pressure

PP-849

Cardiopulmonary exercise test performance in children with various counts of idiopatic ventricular extrasystoles

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Background and Aim: Idiopathic ventricular extrasystoles (VEs) are defined as VEs in patients without structural heart disease and are considered benign in children. However VEs induced cardiomyopathy is described in literature. Cardiopulmonary exercise test (CPET) reports a large number of physiologic measurements during exercise on maximum effort. Aim of the study: to compare the results of CPET performance in children with different numbers of ventricular extrasystoles.
Method: A prospective study performed between 1st January 2022 and 30th September 2023 Vilnius University Hospital Santaros Klinikos. The inclusion criteria: ≥5% PVB in 24-hour electrocardiography and/or group, and/or multiform PVB. The exclusion criteria: hemodynamically significant congenital heart diseases, confirmed diagnoses of cardiomyopathies and accessory pathways. Patients underwent 24-hour electrocardiography (24ECG) and cardiopulmonary exercise testing (CPET) on treadmill (BTL Cardiopoint CPET) using modified ramp protocol. Peak oxygen uptake per kilogram (VO2/kg), anaerobic threshold (AT), respiratory exchange rate (RER), VE/VCO2 were measured during the CPET. The exercise test was terminated if patients get exhausted, develop symptoms or have abnormal blood pressure results during exercise. According to the amount of PVCs in 24ECG, we divided patients into two groups ≤10% and >10% PVCs per 24 hours.
Statistical analysis performed with R software. Nominal variables tested for normal distribution with Shapiro-Francia test. Nominal values were presented as mean and standard deviation or minimum, maximum, median respectively. Student's t-test used to compare means between normally distributed nominal variables. Chi-square used to compare categorical variables. The p value ≤0,05 was considered statistically significant.
Results: Sixty-two patients (53,2% male) were 12.3±2.5 years old with median activity time 217,5 hours per week (maximum 1725 hours per week) and VEs 10.8±4,7% per 24-hour ECG. CPET duration was 5,5-11 (median 8,2) minutes. Patients reached VO2 peak 41,5±6.1 ml/kg/min; AT 759-1634 (median 922) ml/min, VE/VCO2 30,9-100 (median 40,85), RER 1,8±0.5. Peak VO2/kg was lower in <10 VEs group., We found no significant difference in other CPET factors and ≤10% and >10% VEs per 24 hours.
Conclusions: Peak VO2/kg was lower in the >10% VEs group. We found no statistically significant difference in patient's activity time, AT, RER, VE/VCO2, CPET duration between the groups.

Keywords: ventricular extrasystoles, children, cardiopulmonary exercise test

PP-851

The super mario way: Assessing the suitability of trampoline jumping for cardiopulmonary exercise testing and developing an appropriate testing protoc

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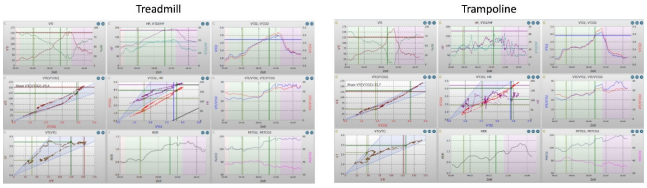
Background and Aim: Cardiopulmonary exercise testing (CPET) of children can be difficult, since they often lack the coordination for treadmill testing and are too small for cycle ergometry. Previous alternative testing methods using mobile equipment outdoors have proven to be successful. However, motivation and safety issues necessitate new and alternative approaches. The objective of this study was to assess the suitability of trampoline jumping as a modality for CPET compared to traditional treadmill tests as well as developing an appropriate testing protocol. We used adults for safety reasons.
Method: 30 young healthy adults (17 male, 13 female) with a mean age of 23.9 years were recruited to perform a treadmill test and a trampoline test with a minimum of one week apart. The running test started at 4 km/h, increasing the speed by 2 km/h every two minutes until exhaustion. The jumping test was performed using a portable CPET device. The testing protocol with five 2-minute levels started with slow walking in place and ended with maximum intensity jumping. Samples for capillary blood gas analysis were taken after each test to determine blood lactate levels. In both tests, portable ECG-devices were used to measure the heart rate (HR).
Results: The peak oxygen uptake (V.O2peak) on the trampoline did not differ significantly from the treadmill (43.4 vs. 43.9 ml/kg/min). The maximum HR was lower while jumping than running (182 vs 190 bpm). The maximum RER on the trampoline was significantly lower than on the treadmill (1.04 vs. 1.12). Lactate levels after jumping did not differ significantly from running (10.2 vs. 11.1 mmol/l). An example for a trampoline test is presented in figure 1.
Conclusions: The trampoline seems to be an adequate alternative for the treadmill as both tests produce comparable results. Due to the lower maximum RER measured during the trampoline tests, it can be assumed that even higher performance values are possible

Figure 1



Example of a participants' matching 9-field-graphics

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on the trampoline. We are planning the implementation of the test protocol in children and patients with congenital heart disease.

Keywords: mobile exercise testing, physical activity, treadmill, sports

PP-852

Trends of submaximal slope parameters in cardiopulmonary exercise testing provide underlying mechanisms of sex differences in obese adolescents

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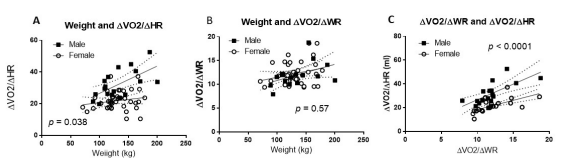
Background and Aim: Because obese individuals generally show decreased exercise performance due to early physical exhaustion caused by excessive non-muscle body mass or fat, peak CPET values and weight-index values are not physiologically relevant to represent their cardiopulmonary reserve. Sex differences of CPET in obese adolescents are poorly understood.
Method: Anthropometric measurements and CPET parameters (bicycle ergometer) were analyzed retrospectively for obese adolescents seen at weight management program. Peak heart rate (HR), peak oxygen consumption (VO2), peak work rate (WR), peak minute ventilation (VE), ventilatory anaerobic threshold (VAT), and submaximal sloop parameters including δVO2/δHR (stroke volume or SV), δHR/δWR (HR-dependency), and δVO2/δWR (oxygen cost) were obtained. Data are shown as mean ± standard deviation.
Results: Nineteen males (17.0 ± 1.9 years; weight 134 ± 32 kg; BMI 49.1 ± 10.0 kg/m2) and 40 females (16.9 ± 1.9 years; weight 129 ± 23 kg; BMI 47.4 ± 8.8 kg/m2) were enrolled. δVO2/δHR and δHR/δWR are surrogates of SV and HR-dependency, respectively. pVO2 and δVO2/δHR were significantly higher in male than in female (3.18 ± 0.39 vs. 2.50 ± 0.46 L/min, $p < 0.0001$ and 32.0 ± 8.9 vs. 21.5 ± 5.4, $p < 0.0001$, respectively) but lower δHR/δWR (0.37 ± 0.08 vs. 0.55 ± 0.09, $p < 0.0001$), suggesting males had higher peak exercise performance, higher SV, and lower HR-dependency than females. VAT and δVO2/δWR were comparable between males and females (2.39 ± 0.72 vs. 2.11 ± 0.63 L/min and 12.1 ± 2.2 vs. 11.9 ± 2.4, respectively). The correlation between weight and δVO2/δHR demonstrated a positive linear relationship with a steeper slope in males, suggesting higher SV with the same weight in males (Figure 1A) whereas correlations between weight and δVO2/δWR almost overlapped each other (1B). With the same oxygen cost (δVO2/δWR), males showed distinctively higher SV (δVO2/δHR) than females (1C), suggesting increase of oxygen cost was more efficiently resulting in increase of SV increase in males.
Conclusions: Interpretation of CPET in obese individuals are challenging because of excessive non-muscle mass and early exercise termination with exhaustion. Submaximal CPET parameters are useful in understanding exercise physiology with obese individuals.

Keywords: Cardiopulmonary reserve, exercise performance, obesity, Cardiopulmonary exercise testing, sex differences, submaximal exercise

Two dimensional CPET analysis

Figure 1



By creating a graph by two CPET parameters, the sex differences of exercise performance in obese adolescents were delineated.

PP-853

Limited stroke volume reserve detected by cardiopulmonary exercise testing represents subclinical cardiotoxicity in childhood cancer survivors

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Background and Aim: Childhood cancer survivors (CCS) show decreased exercise performance despite normal ventricular function at rest. Multiple underlying mechanisms are considered. Decreased stroke volume reserve (SVR) may be one major determinant responsible for poor exercise performance in CCS.
Method: Cardiopulmonary exercise testing (CPET) was performed with bicycle ergometer in asymptomatic off-treatment CCS with normal left ventricular function by echocardiogram; LVSF ≥ 28% or LVEF ≥ 55%. Patients were divided into two groups by the estimated SVR represented by submaximal slope of [VO2/kg]/heart rate (HR) (δ [VO2/kg]/δHR) in males and females. An average δ [VO2/HR]/δHR values of 0.30 and 0.25 were set as thresholds between "Good" and "Poor" SVR in male and female CCS, respectively. Data are shown as mean ± standard deviation.
Results: Demographic and CPET data are presented in Table 1 (38 and 41 CCS males and females, respectively) in comparison with 67 male and 70 female controls. Older ages and larger body size were characteristic in "Poor" CCS groups in both sexes. There was no significant difference in years after the diagnosis in CCS. In the "Poor" groups, peak VO2 (pVO2)/kg, peak work rate (pWR)/kg, peak oxygen pulse/kg (pOP)/kg, and ventilatory anaerobic threshold (VAT)/kg were significantly lower whereas HR-dependency (δHR/δ [WR/kg]) was significantly higher in both sexes, consistent with the limited SVR in "Poor" groups. Peak respiratory exchange ratio (RER) was significantly higher in "Poor" than in "Good" group only in males. Submaximal slope of VO2/WR (δVO2/δWR) was significantly lower in "Poor" groups in both sexes, suggesting limited VO2 increase in response to exercise. There was no difference in ventilatory efficiency (δVE/δCO2) between "Good" and "Poor" groups in either sex. "Good" CCS groups showed comparable peak and submaximal CPET values with control groups.
Conclusions: Limited SVR with increased HR-dependency is one major feature of poor exercise performance in CCS in both sexes. Those with low SVR tended to be older and exhibit worse exercise performance, suggesting progression of exercise intolerance during adolescence. There are some sex differences in exercise performance of CCS, suggesting complex mechanisms of cardiovascular toxicity by cancer treatment.

Keywords: cardiovascular toxicity, cancer treatment, oxygen consumption, exercise performance, stroke volume, anthracycline

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Cardiopulmonary Exercise Testing in CCS and Controls

	Male CCS (N = 38)		Male Control	Female CCS (N = 41)		Female Control
Δ[VO2/kg]/ΔHR	"Poor": ≤ 0.30	"Good": > 0.30		"Poor":≤ 0.25	"Good": > 0.25	
N	18	20	67	24	17	70
Age (years)	**15.8** ± 3.1†	13.6 ± 2.3	14.6 ± 1.9	**15.6 ± 2.7**†	13.4 ± 2.6	14.5 ± 1.9
Height (cm)	169 ± 14	160 ± 13	167 ± 12	**162 ± 8**†	151 ± 12§	161 ± 8
Weight (kg)	**65.6 ± 16.7**†	53.2 ± 13.6	60.4 ± 15.1	**62.3 ± 10.3**†§	45.4 ± 12.4§	55.9 ± 10.4
Total Anthracycline Dosage (mg/m^2)	191 ± 99	202 ± 98	N/A	228 ± 109	209 ± 125	N/A
Age at Diagnosis	9.3 ± 5.5	7.0 ± 4.4	N/A	**7.0 ± 4.5**†	3.9 ± 3.1	N/A
Years after Diagnosis	8.62 ± 4.90	9.49 ± 3.99	N/A	6.57 ± 4.76	6.51 ± 3.26	N/A
CPET						
pHR (beat/min)	192 ± 10	185 ± 11	187 ± 12	188 ± 9	185 ± 8	185 ± 11
pRER	**1.29 ± 0.15**†§	1.19 ± 0.09	1.18 ± 0.09	1.24 ± 0.11	1.22 ± 0.13	1.18 ± 0.07
pVO2/kg (ml/kg/min)	**31.3 ± 6.3**†§	40.5 ± 7.9	44.0 ± 7.6	**25.3 ± 5.8**†	36.2 ± 7.6	33.8 ± 6.2
pOP/kg (ml/kg)	**0.17 ± 0.07**†§	0.22 ± 0.04	0.24 ± 0.04	**0.14 ± 0.03**†	**0.22 ± 0.04**§	0.18 ± 0.03
pWR/kg (Watts/kg)	**2.10 ± 0.46**†§	**2.58 ± 0.60**§	3.08 ± 0.62	**1.73 ± 0.44**†§	2.06 ± 0.40	2.40 ± 0.43
VAT/kg (ml/kg/min)	**18.7 ± 4.5**†§	26.3 ± 5.8	28.4 ± 7.0	**17.3 ± 4.4**†§	25.1 ± 5.4	23.0 ± 5,9
Δ[VO2/kg]/ΔHR (ml/kg)	**0.23 ± 0.05**†§	0.37 ± 0.05	0.35 ± 0.07	**0.18 ± 0.07**†§	**0.35 ± 0.07**§	0.29 ± 0.07
ΔHR/Δ[WR/kg] (kg/60l)	**40.6 ± 11.4**†§	30.8 ± 7.6	30.6 ± 6.2	**47.6 ± 12.5**†§	35.1 ± 8.4	36.9 ± 7.5
ΔVO2/ΔWR (l/60l)	**9.7 ± 3.1**†	11.3 ± 2.1	10.9 ± 1.5	**8.6 ± 2.0**†§	**13.4 ± 4.9**§	10.6 ± 1.9
ΔVE/ΔVCO2	24.4 ± 4.1	23.3 ± 1.9	24.6 ± 3.0	25.3 ± 3.0	24.2 ± 3.1	25.4 ± 2.9

† p < 0.05 compared with "Good" group, § p < 0.05 compared with Control group

CPET results of CCS with poor and good stroke volume reserve ("Poor" and "Good", respectively) in males and females.

PP-854

Decreased submaximal slope of vo2/work rate in cardiopulmonary exercise testing characterizes physical deconditioning in healthy nonobese adolescents

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Background and Aim: Physical deconditioning (PD) due to lack of exercise is prevalent among contemporary adolescents, which can be a substantial risk for long-term adverse cardiovascular events. However, physiological definition of PD is poorly understood. Peak and submaximal parameters in cardiopulmonary exercise testing (CPET) were studied to characterize the underlying physiology of PD.
Method: Anthropometric measurements and CPET parameters (bicycle ergometer) were analyzed retrospectively in nonobese healthy children age ranging from 11 to 19 years old. Resting and peak heart rate (HR), % systolic blood pressure (SBP) response to peak exercise, peak oxygen consumption (VO2), peak work rate (WR), peak oxygen pulse (OP), peak minute ventilation (VE), and ventilatory anaerobic threshold (VAT) were measured. Submaximal sloop parameters including δ [VO2/kg]/ δHR (stroke volume or SV), δHR/δ [WR/kg] (HR-dependency), δVO2/ δWR (oxygen uptake by work rate), and δVE/ δVCO2 (ventilatory efficiency) were obtained. The cohort was divided into 3 groups by %predicted max VO2 (%PmaxVO2); < 80%, 80 to 100%, and > 100%, males and females separately. Data are shown as mean ± standard deviation.
Results: Data are presented in Table 1 (71 males and 62 females). Height was comparable in all age groups. Although normal, BMI was significantly higher in %PmaxVO2 ≤ 100% groups than > 100% group in both sexes. Peak HR and %SBP increase were comparable in all groups. Peak pVO2/kg and peak WR/kg were lowest in < 80% group and highest in > 100% in both sexes, consistent with the definition of grouping. Peak OP, VAT/kg, and δ [VO2/kg]/ δHR in < 80% group were significantly lower than those in ≥ 80% groups whereas δHR/δ [WR/kg] (heart rate-dependency) was significantly higher in < 80% group than ≥ 80% groups in both sexes. δVO2/δWR was significantly lower in < 80% group than > 100% groups, suggesting limited VO2 increase in response to exercise in PD. Significantly high δVCO2/δVE, lower ventilatory efficiency, was only noted in < 80% males.
Conclusions: The physiological features of PD were characterized as a combination of lower stroke volume estimate (lower peak OP and lower δ [VO2/kg]/δHR), higher HR-dependency (higher δHR/δ [WR/kg]), lower δVO2/δWR (decreased VO2 generation per work), and higher BMI.

Keywords: Physical deconditioning, exercise intolerance, cardiopulmonary exercise testing, submaximal exercise, cardiovascular risk, stoke volume

Cardiopulmonary exercise testing

Table 1 Cardiopulmonary Exercise Testing (CPET)

	Male			Female		
%Predicted max VO2	< 80%	80-100%	> 100%	< 80%	80-100%	> 100%
Age (years)	14.7 ± 2.5	14.9 ± 1.7	15.0 ± 2.0	15.2 ± 1.3	15.1 ± 1.4	14.8 ± 1.2
Number	11	24	36	12	30	20
Weight (kg)	66.4 ± 17.1	66.9 ± 13.5	58.1 ± 14.2	**62.0 ± 8.8** †	56.1 ± 8.6	53.6 ± 8.4
Height (cm)	168 ± 16	171 ± 10	169 ± 12	164 ± 6	162 ± 6	162 ± 7
BMI (kg/m^2)	**23.3 ± 3.3** †	**22.7 ± 1.7** †	20.1 ± 2.7	**22.9 ± 2.9** †	**22.2 ± 2.9** †	20.3 ± 2.0
Rest HR (bpm)	75 ± 20	74 ± 16	70 ± 13	77 ± 11	**85 ± 15** †	75 ± 15
Peak HR (bpm)	181 ± 11	185 ± 13	188 ± 11	183 ± 11	186 ± 8	186 ± 12
%SBP increase	149 ± 14	152 ± 17	152 ± 21	149 ± 17	138 ± 23	146 ± 20
Peak VO2/kg (ml/kg/min)	**30.9 ± 3.7** †§	**40.5 ± 6.3** †	49.5 ± 4.5	**25.9 ± 1.7** †§	**31.9 ± 2.6** †	39.8 ± 4.3
Preak WR/kg (watts/kg)	**2.26 ± 0.33** †§	**2.89 ± 0.47** †	3.45 ± 0.43	**2.02 ± 0.23** †§	**2.27 ± 0.24** †	2.81 ± 0.35
peak OP (ml)	**11.2 ± 2.6** †§	15.0 ± 3.5	15.4 ± 3.9	**8.9 ± 1.4** †	**10.0 ± 1.5** †	11..6 ± 2.2
Peak VE (L/min)	85 ± 28	97 ± 29	107 ± 33	65.0 ± 18.8	68.6 ± 12.6	76.7 ± 18.5
Peak RER	1.26 ± 0.12	1.18 ± 0.09	1.19 ± 0.09	1.22 ± 0.10	1.20 ± 0.06	1.16 ± 0.06
VAT/kg (ml/kg/min)	**21.2 ± 3.2** †	**26.1 ± 6.1** †	30.5 ± 6.1	**17.9 ± 3.3** †	**21.3 ± 4.4** †	27.3 ± 5.5
Δ[VO2/kg]/ΔHR (ml/kg)	**0.30 ± 0.08** †	**0.33 ± 0.06** †	0.38 ± 0.06	**0.23 ± 0.08** †	0.28 ± 0.06	0.32 ± 0.07
ΔHR/Δ[WR/kg] (kg/60J)	**36.0 ± 13.2** †	31.2 ± 6.2	28.4 ± 3.6	**41.8 ± 9.2** †	36.9 ± 8.6	34.5 ± 6.3
ΔVO2/ΔWR	**8.4 ± 1.7** †§	**10.3 ± 1.4** †	11.3 ± 1.1	**9.1 ± 1.3** †	**10.1 ± 2.0** †	11.2 ± 1.4
ΔVCO2/ΔVE	**28.7 ± 3.8** †§	24.4 ± 2.8	24.4 ± 2.9	24.7 ± 1.8	25.1 ± 2.7	26.1 ± 3.0

† p < 0.05 compared with > 100% group, § p < 0.05 compared with 80-100% group.
SBP: sytolic blood pressure, WR: work rate, OP: oxygen pulse, VE: minute ventilation, RER: respiratory exchange ratio

Nonobese adolescents were divided into 3 groups based upon %predicted max VO2.

PP-855

A comparison of hybrid vs in-person cardiac fitness program outcomes for patients with congenital heart disease

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Background and Aim: Cardiac fitness and rehabilitation programs have numerous benefits for children and adults living with congenital heart disease (CHD). Recently, delivery methods have expanded from all in-person training to hybrid training (virtual plus in-person). The aim of this study was to compare in-person and hybrid programs by evaluating for differences in fitness outcome metrics and progression, safety, satisfaction, and program adherence.
Method: Cardiac fitness program outcome metrics were retrospectively analyzed from 01/2017 through 09/2023. Based on enrollment factors, patients with CHD either participated in all in-person or hybrid training. Effectiveness was assessed by physiological (aerobic, strength, flexibility metrics), psychologic (mindset survey), and highest fitness progression level achieved (entry, intermediate, advanced). Data on safety (adverse events), satisfaction, and program adherence (number of sessions) were collected.
Results: 105 participants engaged in the cardiac fitness program, 51 were in-person (median age 18.8 years, range 8.7-52.4, 45% female) and 54 were hybrid (median age 20.4 years, range 9.2-55.3, 67% female). Patient characteristics (other than female preponderance in hybrid group, p=0.023) were similar between groups, including CHD classification, hemodynamic level, and distance traveled. Pre- to post-program aerobic, strength, flexibility and mindset fitness metrics improved for both groups. Table 1 shows the comparison between groups, with no significant physiological or psychological differences other than a favorable increase in plank time and decrease in parent anxiety for the hybrid group (p=<0.001, p=0.002, respectively). Patient satisfaction and adherence were similar. Figure 1 shows twice as many participants progressed from entry to advanced level (33% vs. 16%) and fewer

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Figure 1 and Table 1

Figure 1: Exercise Progression Level for the Hybrid vs In-Person Groups

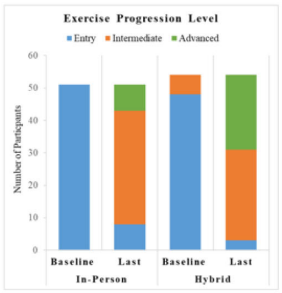

Table 1: Comparison of Patient and Program Outcomes in Hybrid vs In-Person Cardiac Fitness Programs

	N (in-person, hybrid)	In-Person Only	Hybrid	P Value
Aerobic		*Change Pre-Post*	*Change Pre-Post*	
Percent predicted peak VO_2	33, 26	6.7 ± 9.2	4.0 ± 8.2	0.24
Percent predicted VAT at VO_2	33, 24	2.0 ± 6.2	4.5 ± 9.5	0.26
METs (peak)	33, 26	0.7 ± 1.0	0.3 ± 0.8	0.11
Strength		*Change Pre-Post*	*Change Pre-Post*	
Push-ups (total number)	36, 53	7 [4, 10]	8 [2, 11]	0.68
Squats (total number)	9, 35	12 [6, 20]	16 [10, 30]	0.36
Plank (seconds)	41, 53	13 [2, 27]	30 [18, 44]	<0.001
Flexibility		*Change Pre-Post*	*Change Pre-Post*	
Sit and reach (cm)	46, 50	3.8 ± 5.5	4.0 ± 4.4	0.83
Mindset		*Change Pre-Post*	*Change Pre-Post*	
Meaning and Purpose				
Participant	14, 20	3 ± 6	3 ± 6	0.97
Parent	14, 15	4 ± 9	6 ± 10	0.52
Anxiety				
Participant	15, 16	-4 ± 9	-2 ± 8	0.48
Parent	13, 13	0 ± 8	-7 ± 8	0.020
Satisfaction				
Program Very satisfied Satisfied Neutral Unsatisfied Very unsatisfied	22, 44	 20 (91%) 1 (5%) 1 (5%) 0 (0%) 0 (0%)	 35 (80%) 6 (14%) 2 (5%) 0 (0%) 1 (2%)	0.82
Adherence				
Total number of sessions	51, 54	17 [12, 22]	19 [16, 22]	0.28

Mean ± standard deviation or median [25th, 75th percentiles]

Comparisons are made using Fisher's exact test and the Wilcoxon rank sum test

Figure 1: Exercise Progression Level for the Hybrid vs In-Person Groups Table 1: Comparison

stayed at entry level (7% vs. 16%) in the hybrid group, but this did not reach significance (p=0.08). No adverse events occurred in either group.

Conclusions: Both hybrid and in-person delivery methods were safe and effective, showing similar fitness improvements. Virtual training not only did not reduce improvements, but may even show some gains over the all in-person program. Interestingly, there was similar geographic distance, adherence, and satisfaction with both delivery methods. These key findings support the inclusion of virtual training in fitness programs for patients with CHD.

Keywords: Cardiac Fitness, Congenital Heart Disease, Virtual Training, Hybrid Fitness Program, Cardiac Rehabilitation

PP-856

Relationship between maximal and submaximal exercise variables in patients with fontan circulation

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Background and Aim: Most Fontan patients live with major limitations in cardiorespiratory fitness (CRF). Peak oxygen uptake (VO_2peak) is a strong prognostic variable in a variety of cardiopulmonary diseases. However, its assessment requires maximum effort during the cardiopulmonary exercise test (CPET) which is often not achieved by Fontan patients. Consequently, there is a need of reliable submaximal variables. From research in other cardiopulmonary diseases and in older Fontan patient populations, we know that both ventilatory threshold (VT) and oxygen uptake efficiency slope (OUES) are valuable variables. These two, and other submaximal variables, have previously shown to have independent predictive value. Their value in assessing CRF in the younger Fontan population is still to be demonstrated. To increase the prognostic yield from tests not reaching maximal effort, we aimed to evaluate the association between submaximal and maximal variables derived from CPET in adolescent Fontan patients.

Method: This cross-sectional study included a national cohort of 44 adolescents (median age = 16.6 years, 18 females) with Fontan circulation during pre-transition clinical work-up. A CPET was performed on a treadmill using a stepwise protocol until exhaustion. Gas exchange and ventilatory variables were continuously determined by breath-by-breath sampling, averaged over 30-s intervals. The association of VO_2peak, VO_2@VT and OUES in both mL-min^{-1} and mL-kg^{-1}-min^{-1} were assessed by Pearson correlation coefficient. A weak correlation was defined as r<0.3, moderate as r=0.3-0.7, and high as r>0.7. *Results:* The VO_2peak in both mL-kg-min^{-1} and mL-min^{-1} were highly correlated with VO_2@VT (figure 1: A/D) and the OUES (figure 1: B/E). There was also a high correlation between VO_2@VT and OUES in both mL-kg-min^{-1} and mL-min^{-1} (figure 1: C/F).

Conclusions: Submaximal variable measurements obtained through CPET in adolescent Fontan patients show a significant correlation with CRF. Submaximal reference values for children and adolescents are needed for assessing CRF in individuals who fail to achieve maximal effort during exercise testing.

Keywords: Cardiopulmonary exercise testing, Fontan circulation, cardiorespiratory fitness, ventilatory threshold, oxygen uptake efficiency slope

Figure 1

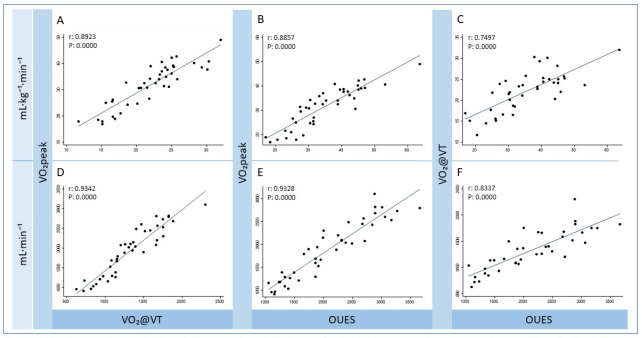

Figure 1: Correlation of VO_{2peak} with VO_2@VT in mL·kg·min^{-1} (A) and mL·min^{-1} (D), VO_{2peak} with OUES in mL·kg·min^{-1} (B) and mL·min^{-1} (E), and VO_2@VT with OUES in mL·kg·min^{-1} (C) and mL·min^{-1} (F).

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