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Cardiology in the Young

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Basic science, Genetics

O-001
Defining genotype-phenotype correlation in paediatric patients with hypertrophic cardiomyopathy

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Background and Aim: Hypertrophic Cardiomyopathy (HCM) is the most common inherited heart disorder. In most cases, it is due to mutations in cardiac sarcomere protein genes. In < 10% children HCM may be associated with inborn errors of metabolism, neuromuscular diseases, and malformation syndromes.

Recent studies support that certain genetic variants may be related to a more severe evolution or a modulating effect. Paediatric onset HCM involve relevant impact, and risk of major events assessment remains a challenge.

The aim of this study is to describe the genotype and phenotype of a cohort of young patients with HCM.

Methods: Data of 0-25 years-old patients diagnosed with HCM during paediatric age (0-18 yo) and currently followed-up in a national reference centre for Family Heart Disease were collected. Those with genetic variants of interest (pathogenic, likely pathogenic or of uncertain significance but with suspected pathogenicity) were included.

Clinical data were prospectively collected from diagnosis or follow-up beginning. Genetic studies were performed by gene panels and Sanger or Next Generation Sequencing. Variants pathogenicity was classified by the American College of Medical Genetics criteria.

Results: 63 patients were included. 41 had variants in sarcomeric genes (Table 1.A). Most prevalent variants were found in MYBPC3 gene (n = 20), followed by MYH7 (n = 10). 14 were associated with RASopathies. Major events (sudden cardiac death or appropriate implantable cardioverter defibrillator - ICD - therapies) or need for intervention (myomectomy, ICD implantation or transplantation) were present in 100% patients with variants in TPM1 and 70% patients with variants in MYH7, comparing with 35% in MYBPC3 or 21% of RASopathy-associated genes (p = 0.01).

Some cases with double mutation in distinct sarcomeric protein genes with early and aggressive phenotype are described (Table 1.B). We have further identified equal variants – in MYBPC3, TPM1, ACTC1 and BRAF – with similar medical evolution in different patients from diverse families (Table 1.C).

Conclusion: Genetic study in paediatric HCM provides not only useful information for genetic counselling, but also it may add a prognostic value, involving a significant impact on evaluation and early management of children and teenagers with HCM. Genotype-phenotype records are essential to evaluate genetic assessment application for predictive purposes.

Keywords: Genotype-Phenotype, Hypertrophic Cardiomyopathy, Paediatrics, Assessment, Prediction.

O-002
Shaping the heart tube by modulation of cardiomyocyte differentiation: identification of greb1l as a genetic determinant of criss-cross heart in MICE

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Background and Aim: Criss-cross heart is a rare congenital heart defect (1/125 000) defined by abnormal twisting of the atrioventricular valves and often associated with mispositioned supero-inferior ventricles. However, the origin of this disease has remained unknown. We have uncovered a mouse model of criss-cross heart with supero-inferior ventricles: Greb1l mutants. Exploiting this model, we aim to understand the embryological origin of criss-cross heart and identify the molecular mechanisms involved in this malformation.
Methods: We developed new 3D quantitative procedures of cardiac anatomy to analyze comparatively mouse mutants and patients. We used a combination of mouse genetics, 3D spatially-resolved gene expression and transcriptomic analyses to tackle molecular mechanisms.
Results: Using two mouse mutant lines, we show that Greb1l is required for cardiac chamber alignment at E10.5, after the process of heart looping. Our quantitative measures support a model where growth arrest of the outflow tract mechanically twists the heart tube to generate a criss-cross heart with supero-inferior ventricles.

Reconstitution of the spatio-temporal dynamics of Greb1l expression indicates that it is mainly expressed in heart progenitors before myocardial differentiation. Greb1l, which encodes an unknown protein, had been previously identified as a target of retinoic acid signaling. By mapping the expression profile of genes dependent on retinoic acid signaling and quantifying the phenotype of retinoic acid deficient mutants, we show that Greb1l inactivation phenocopies partial retinoic acid deficiency. The unchanged pattern of the RARE-lacZ reporter transgene in Greb1l mutants suggests that GREB1L is a novel effector of retinoic acid signaling. Transcriptomics of Greb1l mutants coupled with the analysis of published single cell datasets show that GREB1L represses cardiac differentiation and promotes ribosome biogenesis.
Conclusion: Our work deciphers for the first time an embryological origin of criss-cross heart in mice. In addition, we show that modulation of cardiomyocyte differentiation after cardiac looping is important to shape the heart tube. The molecular mechanisms that we identify downstream of GREB1L are relevant to other human congenital defects, which are associated with GREB1L genetic variants.

Keywords: Heart morphogenesis, Criss-cross heart, supero-inferior ventricles, Greb1l, mouse genetics

O-003
Pathophysiological role of dexmedetomidine for monocrotaline-induced pulmonary arterial hypertension in rats

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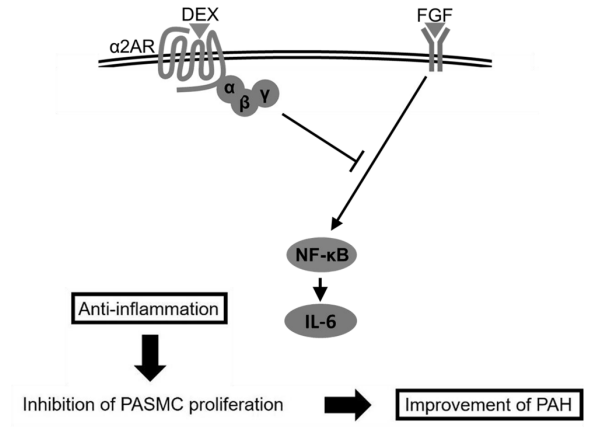
Background and Aim: The abnormal proliferation of pulmonary arterial smooth muscle cells (PASMCs) is one of pathological characteristics of pulmonary arterial hypertension (PAH). Inflammation is thought to play an important role for PASMCs proliferation. Dexmedetomidine (DEX) is a selective α_2-adrenergic receptor agonist that is used as a sedative in clinical settings. It has been reported that DEX decreased pulmonary arterial pressure, and suppressed inflammation. Thus, our aim in this study is to demonstrate that DEX improves Monocrotaline (MCT)-induced PAH in rats through its anti-inflammatory effect.
Methods: In vivo, 6-week-old male Sprague-Dawley rats were injected 60 mg/kg of MCT subcutaneously. At day 14 after MCT injection, continuous infusions of DEX (2 μg/kg/hour) by osmotic pumps were started in one group (MCT + DEX group), and not in another group (MCT group). We performed cardiac catheterization to measure right ventricular systolic pressure (RVSP) at day 23, and conducted histological analyses with paraffin-embedded rat lung tissues. *In vitro*, we examined human PASMCs with fibroblast growth factor 2 (FGF2) stimulation.
Results: Both RVSP and survival rate markedly improved in MCT + DEX group compared with those in MCT group (RVSP; 34mmHg ± 11mmHg vs. 70mmHg ± 25mmHg, survival rate 42% vs. 0% at day 29 ($P < 0.01$)). In histological analyses, medial hypertrophy of pulmonary arterioles and phosphorylated-p65 positive PASMCs reduced in MCT + DEX group. Western blotting with Rat lung homogenates showed that phosphorylated-p65 protein expression was diminished in MCT + DEX group. DEX also inhibited human PASMCs proliferation stimulated with FGF2 dose dependently. Furthermore, interleukin-6 mRNA expression was suppressed by DEX in human PASMCs stimulated with FGF2.
Conclusion: These results indicate that DEX improves MCT-induced PAH in rats by suppressing PASMCs proliferation through the anti-inflammatory effect, which may result from the inhibition of nuclear factor-kappaB activation induced by FGF2. DEX can be a new therapeutic tool for PAH, and the result that DEX had no toxicity for human PASMCs is desirable as a therapeutic agent.

Keywords: Pulmonary arterial hypertension (PAH), Dexmedetomidine, Pulmonary arterial smooth muscle cells (PASMCs), Nuclear factor-kappaB (NF-kB), Interleukin-6, Fibroblast growth factor 2 (FGF2)

Speculated signaling pathways



DEX improved PAH by suppressing PASMCs proliferation through anti-inflammatory effect which probably result from inhibition of NF-κB activation.

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O-004
Identifying new genetic causes of pediatric cardiomyopathy by exome sequencing and zebrafish modeling

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Background and Aim: Genetic pediatric cardiomyopathies define a heterogeneous group of heart muscle diseases associated with cardiac dysfunction. In children, cardiomyopathy is a rare condition with high morbidity and mortality. Whole exome sequencing (WES) in genetic testing has significantly improved our ability to detect cardiomyopathy-causing genetic variants. However, in a substantial proportion of pediatric cases the cause remains unknown. Also, for many identified causative variants, the underlying mechanisms by which they impair cardiac function and ultimately result in cardiomyopathy remain poorly understood. In this research project, we aim to identify new pediatric cardiomyopathy genes and explore their underlying pathophysiologic mechanisms using the zebrafish as a model system.
Methods: Exome sequencing was used to discover potentially disease-causing variants in children with otherwise unexplained cardiomyopathy. The patient-specific variants were introduced in the corresponding zebrafish orthologs by CRISPR-Cas9. Using *in vivo* imaging cardiac development and function was assessed in established mutant zebrafish lines.
Results: We identified bi-allelic variants in the *FLII* gene (flightless-1 homolog), which encodes an actin-binding member of the gelsolin family and is highly expressed in the sarcomere of striated muscle. *Fli1* mutant zebrafish displayed morphological and functional abnormalities of the heart, including ventricular dilatation and reduced contractility, similar to those found in human disease.
Conclusion: Our data confirm the role of *FLII* in pediatric cardiomyopathy. Further functional analysis of the zebrafish mutants will yield a better understanding of the underlying pathophysiological processes. The discovery of new pediatric cardiomyopathy genes and their underlying pathogenic mechanisms will provide important insights in the disease progression and should ultimately lead to new targets for therapeutic intervention.

Keywords: pediatric cardiomyopathy, exome, zebrafish

O-005
Genetic testing outcomes in a cohort of 21,159 children with heart disease

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Background and Aim: Pediatric heart conditions affect 1 in 77 US children. Guidelines recommend cardiogenetic testing in children and relatives (for some conditions at 10-years-old and over). To our knowledge, a large clinical cohort undergoing genetic testing across the spectrum of pediatric cardiogenetic conditions has not been described. This study investigated outcomes in pediatric probands who had clinician-ordered cardiogenetic testing in one laboratory.
Methods: Probands less than 18-years-old had next generation sequencing and deletion/duplication analysis for cardiogenetic conditions including up to 224 established genes. Using a point-based refinement of ACMG guidelines, a molecular diagnosis (positive result) was defined as a single pathogenic/likely pathogenic (P/LP) variant in an autosomal dominant or X-linked gene or two P/LP variants in a recessive gene. Outcomes were compared to a cardiogenetic testing cohort of 61,368 adults by t-tests with multiple comparison correction.
Results: A total of 21,159 probands were tested (median = 10.03 years, SD = 5.75). Positive results were reported in 16.6%, most frequently for aortopathy (5.4%), cardiomyopathy (3.8%), and arrhythmia (3.4%). Positive syndromic gene results were identified in 41.4% (commonly Marfan syndrome, 19.6%; hereditary hemorrhagic telangiectasia, 3.7%; Loeys-Dietz syndrome, 3.1%). The frequency of positive results in infants (less than 1-year-old) was 16.0%; 51.6% in a syndromic gene. Cascade testing was pursued in 3.9% of families (mean 3.48 relatives/family; 59.1% aged 10 years and under; 14.6% infants). Compared to adults, positive results were more frequent in children (16.6% vs 15.3%), including syndromic genes (41.4% vs 29.7%). Families with probands aged 10 and under had cascade testing completed more often (4.6%; p = .0069), but families with adult probands were larger (mean 4.88 relatives/family; p = .0014).
Conclusion: This study showed that 1 in 6 children had a positive result, highlighting many cardiogenetic management opportunities. In families with probands 10 years-old and under, cascade testing was pursued more often than in families with adult probands. Given higher healthcare utilization among children with heart disease, these findings provide a rationale for family-based care, to identify patients of all ages who can benefit from genetics-guided interventions.

Keywords: genetic testing, heart disease, pediatrics

O-006
Platelet adhesion, fibrin deposition and neo-endothelialization in bovine jugular vein conduits and homografts after implantation in sheep

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Background and Aim: Infective endocarditis remains an associated complication after RVOT valve replacement despite excellent hemodynamic valve function. Knowledge on the in-vivo adaptation of the graft tissues is rare. As platelet and fibrin deposition can favour bacterial attachment, the aim was to evaluate this aspect and the degree of in-vivo re-endothelialization of RVOT conduits implanted in sheep. In addition, the potential effect of anti-aggregant therapy was evaluated.
Methods: Four cryopreserved ovine homografts (CH) and 11 bovine jugular vein conduits (BJV) were implanted in the RVOT of sheep. Hemodynamic function was monitored by echocardiography and MRI. The effect of anti-platelet therapy

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was evaluated for the BJV group. After 4 months, 3 conduits per group were analysed by immunostaining and scanning electron microscopy to evaluate the endothelial cell coverage, platelet adhesion and fibrin deposition. Sheep blood was tested in additional ex-vivo experiments to investigate platelet and S. aureus adhesion to homograft and BJV tissue.
Results: Re-endothelialization was seen in the region of the surgical sutures in CH and BJV, propagating towards the valve leaflets with varying extent. Fibrin covered 43.5% of the surface in the pulmonary wall region of CHs and 30.6% of BJVs. Valve leaflets were fibrin-covered to 20.7% in CHs and 18% in BJVs. Platelet adhesion, detected using an anti-CD42b antibody, was largely concomitant with positive fibrin staining, although generally detected to a lower extent. Treatment with Ticagrelor reduced platelet adhesion of BJV at the surface of leaflets from 13.5 to 8.1% and at the pulmonary side from 11.2 to 5.6%. Also fibrin coverage was reduced at the surface of the leaflets (18 vs. 10.2%) and on the surface of the pulmonary wall (30.6 vs. 6.5%). Platelet and S. aureus adhesion to CH and BVJ tissue was not different when testing blood from ticagrelor treated compared to untreated sheep in the flow chamber in-vitro.
Conclusion: Histophathological findings demonstrate similar platelet adhesion and fibrin deposition characteristics in CH and BJV. However, anti-aggregant treatment reduced but not abolished platelet adhesion and fibrin coverage on BJV leaflet and pulmonary wall region. A potential pathophysiological role in preventing bacterial infection awaits further investigation.

Keywords: infective endocarditis, platelets, re-endothelialisation, RVOT conduits

Imaging/Functional assessment

O-007

What's in a shape? predicting neonatal coarctation of the aorta using 3D statistical shape models of normal and abnormal fetal arch morphology

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Background and Aim: Antenatal diagnosis of coarctation of the aorta (CoA) is challenging with high false positive (FP) rates posing a burden on both parents and healthcare. Motion-corrected three-dimensional (3D) fetal cardiac magnetic resonance imaging (CMR) permits novel in-vivo study of shape and size variability between outcome groups. Statistical shape modelling can be applied to study these variations in shape across different populations by using principal component analysis (PCA). We applied this technique in the largest known 3D *fetal* CMR cohort to study in-utero arch variations between true and FP CoA and controls.
Methods: Fetal CMR examination was offered to women carrying fetuses with suspected CoA and healthy controls between July 2015 and April 2021. Two-dimensional single-shot fast spin echo sequences were acquired and processed using motion-corrected slice-volume registration, resulting in high resolution 3D volumes. Semi-automatic segmentation with manual refinement was used to produce 3D meshes. Centrelines were automatically extracted from aortic and ductal arch and descending aorta. A statistical shape model capturing relevant shape variations was built using PCA. A linear discriminant analysis (LDA) with the first 10 PCA modes was used to generate three optimal axis of anatomical variation to distinguish between controls, FP and neonatal CoA.
Results: In total 165 fetuses (median gestational age 31 weeks, IQR 30-33) were included for statistical shape analysis, of which 112 suspected CoA patients. Outcome was available for 106/112, neonatal CoA was confirmed in 43/106 and one fetus in the FP group developed late CoA. The first 10 anatomical PCA modes captured 86% of the shape variability in the population. Each discriminant axis based on outcome groups (43 CoA, 63 FP, 53 controls) showed distinct morphological differences [Figure 1]. LDA classification performance was excellent across all groups (AUC 0.99 controls - CoA; AUC 0.95 controls - FP; AUC 0.90 FP - CoA).
Conclusion: Fetal arch shape variability exists not only between true CoA and healthy controls, but between FP CoA and controls. This provides unique insights into fetal arch geometry with potential to improve our understanding and antenatal diagnosis of suspected neonatal CoA.

Keywords: Congenital heart disease, Coarctation, Magnetic Resonance Imaging, Statistical shape modelling

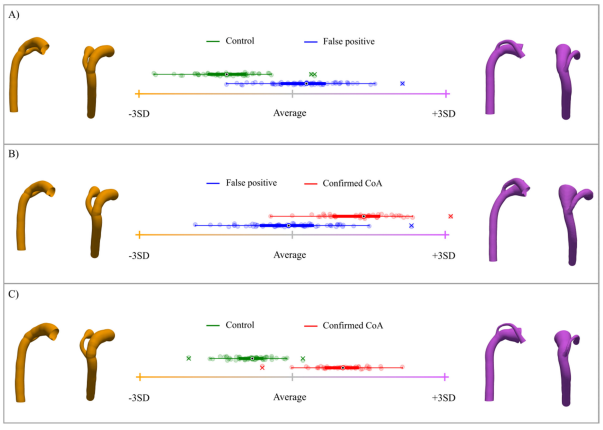

Figure 1 Axis of anatomical variation that best discriminate between confirmed CoA, false positive cases and healthy controls. Boxplots represent controls (green), FP (blue), CoA (red), and Z-score distributions across each axis. Orange cross and 3D shape show -3SD from the mean shape (grey cross), purple cross and shapes +3SD. Axis A) represents axis between controls versus FP, B) FP and confirmed CoA, C) controls and confirmed CoA.

O-008

Impaired cardiac performance during exercise in preterm born young adults with and without bronchopulmonary dysplasia: an exercise mri study

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Background and Aim: Advances in neonatal care have increased survival rates of extreme preterm infants with and without bronchopulmonary dysplasia (BPD) into adulthood. Long-term, functional cardiopulmonary consequences of prematurity with and without BPD are largely unknown. The aim of this study is to examine cardiorespiratory structure and function during (sub)maximal exercise in young adult survivors of extreme prematurity with or without BPD, to reveal dynamic abnormalities that are not apparent on conventional tests at rest.
Methods: 20 premature born with BPD (aged 22 ± 2 y; gestational age [GA] 27(26; 28) wks; >28 days of postnatal oxygen dependency), 20 premature born without BPD (aged 23 ± 2 y; GA 28(27; 29) wks; <28 days of postnatal oxygen dependency) and 20 at term born young adults (aged 22 ± 3 y; GA 39 (38; 40) wks) were subjected to a cardiopulmonary exercise test, baseline cardiovascular magnetic resonance (CMR) and CMR during exercise at 40% and 60% of their maximal oxygen consumption (VO_2max) using a push-pull ergometer.
Results: BPD subjects displayed lower exercise tolerance compared to the at term born subjects (92 ± 18 vs. 106 ± 17% of predicted VO_2max, $p = 0.03$), but not compared to preterm subjects (102 ± 14%, $p = 0.14$). Figure 1 shows cardiac performance at rest and during exercise at 40% and 60% of maximum oxygen consumption in all three groups. At rest, BPD subjects had significantly smaller biventricular end-diastolic volumes and decreased stroke volume compared to at term born subjects (left ventricular end-diastolic volume: 80 ± 11 vs. 91 ± 14 ml/m², $p = 0.01$; stroke volume 47 ± 7 vs. 53 ± 9 ml/m², $p = 0.04$), but not compared to preterm born subjects without BPD (81 ± 13 ml/m², $p = 0.95$ and 49 ± 8 ml/m², $p = 0.69$). Cardiac response to exercise revealed an impaired increase in stroke volume and cardiac index in both preterm groups compared to at term born subjects, but more extensive in BPD subjects. This was attributed to a more profound decrease in left ventricular end-diastolic volume during exercise in BPD subjects.
Conclusion: Preterm born young adults with BPD presented lower exercise capacity and more impaired cardiovascular response to exercise than preterm born subjects without BPD and at term born controls. Reduction of left ventricular end-diastolic volume during exercise could indicate pulmonary vascular disease and/or left ventricular diastolic dysfunction.

Keywords: Exercise, prematurity, bronchopulmonary dysplasia, CMR

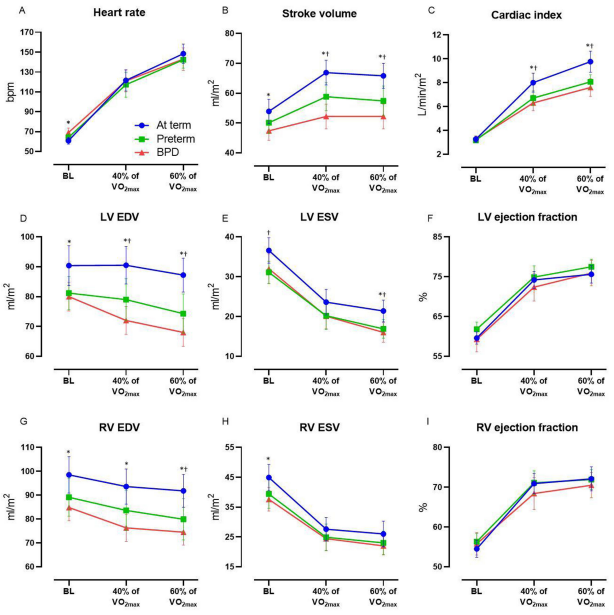

Figure 1: Cardiac performance at rest (BL) and during exercise at 40% and 60% of maximal oxygen consumption (VO_2max) in preterm born young adults with and without bronchopulmonary dysplasia (BPD). LV – left ventricular, EDV – end-diastolic volume, ESV – end-systolic volume, RV – right ventricular. Mean±confidence interval are shown. *p<0.05 for BPD vs. At term, †p<0.05 for preterm vs. at term.

O-009

Fibrotic myocardial remodeling in children and adolescents after cardiac transplantation – a CMR native T1 mapping study

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Background and Aim: Adverse fibrotic myocardial remodeling has been reported in patients after heart transplantation (HT). The aim of our study was to quantify diffuse myocardial fibrosis using CMR in a cohort of children and adolescents after HT, to identify potential underlying factors that are related to increased fibrosis and to assess its hemodynamic and prognostic relevance.
Methods: 105 HT recipients (age 17.9 ± 7.1 years) without any clinical signs of acute rejection were assessed by CMR 11.8 ± 6.8 years after HT. Left ventricular (LV) volumes and mass were derived from short-axis cine images and LV strain/strain rate was assessed using tissue tracking technique. A T1 mapping sequence (MOLLI) was used to quantify native T1 times within the LV myocardium at a midventricular short axis slice. The collagen volume fraction (CVF) was obtained from histological analysis of endomyocardial biopsy samples.
Results: Septal native LV T1 times showed a weak but significant association with CVF ($r = 0.36$, $p = 0.01$). T1 times were not related to LV size, mass and ejection fraction but correlated

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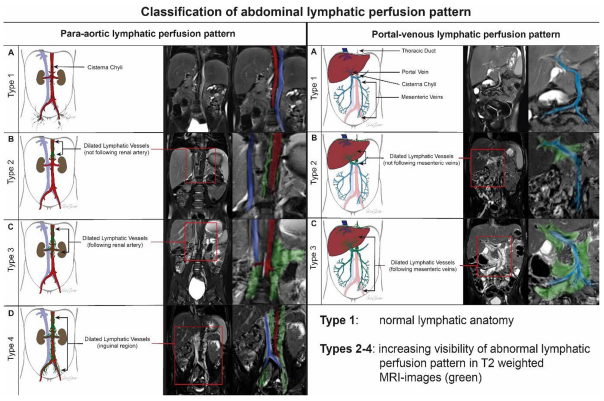
inversely with LV mass-to-volume ratio (r = 0.26, p = 0.001) and systolic (r = 0.32, p = 0.001) and diastolic longitudinal (r = 0.25, p = 0.01) strain/strain rate. Neither a history of rejection, organ ischemic time, BSA ratio donor-acceptor and time since HT were significantly related to T1 times. During a mean follow-up of 5.3 ± 1.6 years, ten patients developed adverse events (death n = 1, re-HT n = 2, listing for re-HT n = 1, coronary interventions n = 6). Univariate Cox-Regression analysis revealed septal native T1 times (HR 1.01, 95% CI 1.00-1.02, p = 0.003) and LV longitudinal strain (HR 1.73, 95% CI 1.25-2.40, p = 0.001) as the only significant predictors for adverse events.
Conclusion: Fibrotic myocardial remodeling after HT affects LV systolic and diastolic longitudinal function, is associated with an increased mass-to-volume ratio and emerged as a univariate predictor for adverse outcome. Future studies should aim to identify underlying factors and mechanisms that promote the development of fibrotic changes post HT.

Keywords: Cardiac Magnetic Resonance, Heart Transplantation, Fibrotic myocardial remodelling



Schematic presentation of our classification of abdominal lymphatic perfusion pattern with T2-weighted MRI samples.

O-010

Classification of abdominal lymphatic perfusion pattern after fontan surgery and association to hypoproteinemia

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Background and Aim: Complications after Fontan surgery have been associated with arise and classification of abnormal thoracic lymphatic perfusion pattern. This study compiles a classification of abnormal abdominal lymphatic perfusion pattern and investigates the impact on serum protein readings.
Methods: Retrospective analysis of 71 patients who underwent magnetic resonance imaging with T2-weighted lymphatic imaging and serum protein measurement 6 month after Fontan surgery. Abdominal lymphatic imaging was classified according to anatomical lymphatic drainage into two categories: (1) para-aortic (type 1-4); (2) portal-venous (type 1-3). Thoracic lymphatic imaging was classified (type 1-4) as described earlier.
Results: Serum protein was 65 ± 8 g/l (< standard value in 37%). Para-aortic lymphatic perfusion pattern classified as type 1 in 4, type 2 in 13, type 3 in 37 and type 4 in16 out of 71 patients. Portal-venous lymphatic perfusion pattern classified as type 1 20, type 2 in 10 and type 3 in 41 patients. Thoracic lymphatic perfusion pattern classified as type 1 in 8, type 2 in 11, type 3 in 39 and type 4 in 13 patients. Higher-grade para-aortic (p = 0.0062), portal-venous (p = 0.022) and thoracic (p = 0.011) lymphatic abnormalities were correlated with lower total serum protein. Higher ratings of para-aortic lymphatic abnormalities were significantly associated with higher ratings of portal-venous abnormalities (p<0.0001). Ratings of para-aortic and portal-venous classifications were correlated with thoracic classification (p<0.001).
Conclusion: Abnormal abdominal lymphatic perfusion pattern can be classified according to anatomical structures with increasing severity. Abdominal and thoracic lymphatic perfusion pattern are associated with serum protein values.

Keywords: Fontan surgery, cardiac MRI, lymphatic imaging, classification of abdominal lymphatic perfusion pattern

O-011

Altered white matter microstructure is associated with executive dysfunction in congenital heart disease across childhood and adulthood

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Background and Aim: Congenital heart disease (CHD) patients are at risk for brain alterations and neurodevelopmental impairments. However, it is unclear whether the extent of alterations decrease with age, especially during adolescence when the white matter microstructure (WMM) undergoes intense maturation processes. Thus, this study investigates differences in WMM between patients with CHD and healthy controls in a pooled sample of subjects from childhood to young adulthood. Secondly, the association between WMM and executive functioning is examined.
Methods: In total, 79 patients with different types of CHD (23 with univentricular CHD) and 137 healthy controls underwent diffusion tensor imaging and standardized neuropsychological tests measuring executive functions. Subjects were between 9 and 32 years of age and originated from three study cohorts with matching inclusion criteria, neuropsychological tests and MRI protocols. A set of major white matter tracts were extracted and mean fractional anisotropy (FA) was calculated for each tract. Multiple linear regression models were conducted including FA as outcome and group and age as predictors. An interaction between age and group was added to investigate differences in white matter alterations across age. Relative variable importance was calculated to estimate the independent contribution of the following factor to the variability in executive functions: Mean FA of the white matter tracts, group, CHD complexity, socioeconomic status, and sex.
Results: Mean FA of all tracts was lower in patients compared to controls (all p<0.01). There was no significant interaction indicating that the extent of WMM alteration in CHD did not change

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across ages (all p>0.1). Executive functions were lower in patients compared to controls – especially in those with more complex CHD. For the total sample, executive functions correlated with FA in several tracts and with socioeconomic status (Figure 1).
Conclusion: WMM is altered in CHD patients from childhood to young adulthood. There is no evidence for improvement after extensive brain maturation in adolescence. Altered WMM is associated with executive dysfunction in CHD patients across age. CHD is a chronic disease with cerebral and neurocognitive impairments persisting into adulthood and, thus, this population requires continuous attention when growing older.

Keywords: Congenital heart disease, cerebral white matter microstructure, diffusion tensor imaging, neurodevelopment, executive function

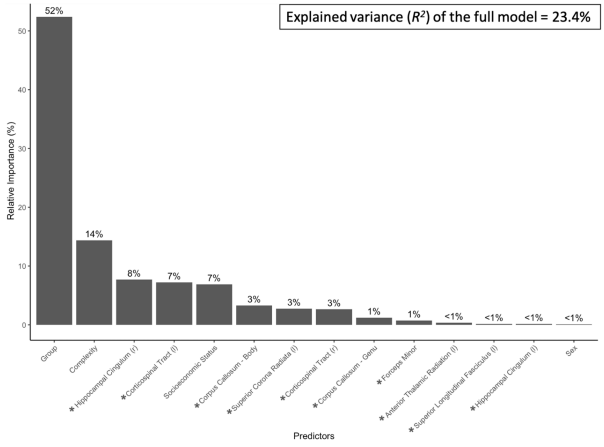

Figure 1 Relative importance of factors predicting executive functions.
Note. (l) = left hemisphere, (r) = right hemisphere.
★ White matter tracts significantly associated with executive functions

O-012

Effects of enzyme replacement therapy on cardiac function and structure in classic infantile pompe disease – up to 22 years of follow-up

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Background and Aim: Patients with classic infantile Pompe disease are born with a hypertrophic cardiomyopathy. Treatment with enzyme replacement therapy (ERT) significantly improves motor function and normalizes cardiac hypertrophy and function. We aimed to assess long-term effects of ERT on cardiac structure and function.
Methods: Twenty-seven Pompe patients treated with ERT were included. Cardiac function was assessed at regular time intervals (before and after start of ERT) using conventional echocardiography and myocardial deformation analysis. Separate linear mixed effect models were used to asses temporal changes within the first year and the long-term follow-up period thereafter. Echocardiograms of 103 healthy children served as controls.
Results: A total of 192 echocardiograms were analyzed. Median follow-up was 9.9 years (IQR: 7.5 - 16.3). Mean LVMI before start of ERT was 292.3 g/m2 (95% CI: 202.8 - 381.8, Z-score +7.6) and after 1 year of ERT 87.3 g/m2 (CI: 67.5 - 107.1, LVMI, Z-score +0.8, p<0.001 for change). Mean shortening fraction was within normal limits before start of ERT, up to 22 years of follow-up. Cardiac function measured by RV/LV longitudinal, and circumferential strain were decreased before start of ERT, but normalized (< -16%) at 1 year after start of ERT. During long-term follow up, only LV circumferential strain gradually worsened in Pompe patients (+0,24%/year) compared to controls. LV longitudinal strain was worse in Pompe patients, but did not change significantly over time compared to controls. Despite these statistical differences in Pompe patients compared to controls, RV/LV longitudinal, and circumferential strain remained within normal limits (< -16%) up to 22 years of follow-up.
Conclusion: Cardiac function assessed by conventional and innovative echocardiographic techniques normalizes after start of ERT and seems to remain stable over a median follow-up period of 9.9 years.

Keywords: Classic infantile Pompe disease, hypertrophic cardiomyopathy, enzyme replacement therapy, myocardial deformation analysis, long-term follow-up.

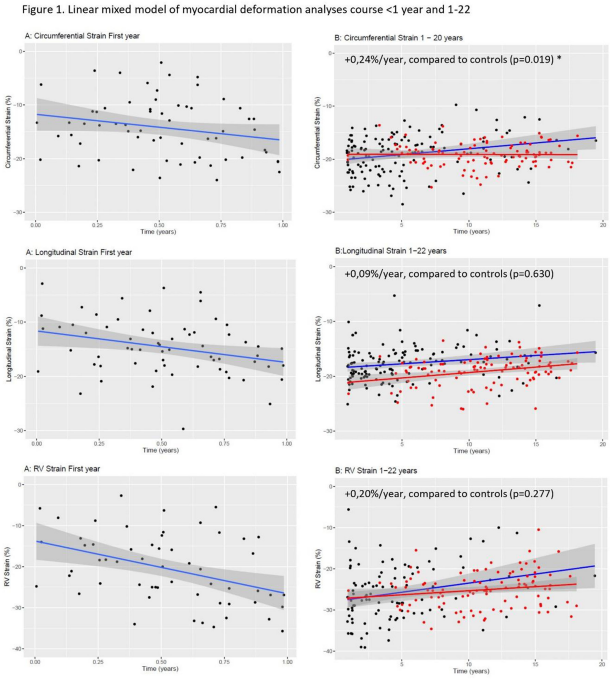

Figure 1. Myocardial deformation analyses course

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Interventional Cardiology

O-013
Piggyback mounting for stent and valve deployment during percutaneous pulmonary valve implantation

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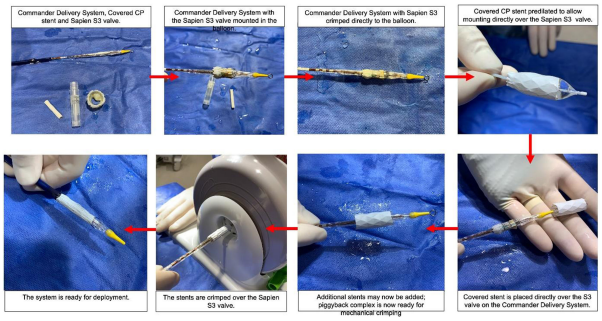
Background and Aim: Heterogeneity and complexity of the Right Ventricular Outflow Tract may complicate stent deployment when preparing a landing zone for transcatheter pulmonary valve implantation. We report our experience in simultaneously implanting multiple stents and valves mounted on a single balloon before and during transcatheter pulmonary valve placement.
Methods: Retrospective analysis of patients from Children's Hospital of Colorado, USA; and Oslo University Hospital, Norway, undergoing transcatheter pulmonary valve replacement that had at least 2 stents mounted on a single balloon, deployed in the RVOT.
Results: Over a 42-month period, a total of 50 subjects from the two centers met inclusion criteria for the study. Subjects were predominantly male (58%), and the median age was 17 years (4-78 years). In 6 subjects (12%) there was need for pre-stenting with use of the double or triple stent Piggyback technique. Forty subjects (80%) had a Melody ™ TPV implanted. In 45 cases (90%) one or more stents were mounted over the pulmonary valve using its delivery system, either the Ensemble for the Melody™ TPV or the Edwards Commander for the SAPIEN 3 THV. 37 subjects (74%) had one stent mounted and 8 subjects (16%) had two stents mounted over the pulmonary valve for simultaneous deployment. In the majority of cases, low profile crimping was achieved by using a modified commercially available valve crimper. No complications related to this technique were reported.
Conclusion: The Piggyback technique aims to simplify and facilitate adequate conduit preparation and valve insertion by minimizing manipulation across the outflow tract and decreasing the risk of stent distortion, misalignment, and embolization.

Keywords: Percutaneous Pulmonary Valve; Pre-stenting; Conduit preparation; Congenital heart disease.

PiggyBack Process Schematic



Piggyback crimping process. An example of our process shows us crimpiing a covered CP stent over the top of a Sapien S3 valve; in this case we were temporizing a patient with profound sepsis secondary to endocarditis in a surgical conduit using a Sapien valve for hemodynamic improvement and a covered stent to try to temporarily exclude the infected conduit valve from the blood pool.

O-014
Echocardiographically guided interventional PDA closure on the NICU in prematures – initial experience on 22 patients

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Background and Aim: Although potentially superior to surgery and drug therapy, percutaneous interventional closure of the patent ductus arteriosus in premature infants is classically dependent on radiation and, hence, transportation of the patients. We present the results of a novel treatment for catheter interventional closure of PDA in premature infants with ultrasound guidance only in an incubator at the local neonatology clinic.
Methods: We developed a method for echocardiographically guided transvenous percutaneous PDA closure in prematures. Clinical and interventional details are provided.
Results: Twenty-two premature infants were treated percutaneously for PDA with echocardiographic guidance only. The median weight of the patients was 1220g, range 750-2800g. In one patient the femoral artery was inadvertently punctured und used for the procedure, in all other patients the intervention was performed through a 4F sheath in the femoral vein. Piccolo devices were used in all patients, the size was based on the echocardiographic measurement of the size of the PDA. Procedural time from puncture to sheath removal was in median 57min (range 25-195 minutes) with a clear tendency towards shorter times in the latter procedures. The ductus was closed successfully in all prematures. In one patient a residual shunt persisted for 3 weeks before spontaneous closure occurred. Two possible postductal thromboembolic events occurred. There were no other complications.
Conclusion: The presented method enabled safe and effective percutaneous closure of the PDA in prematures solely under ultrasound guidance at the bedside. Further establishment of the technique should provide an optimal treatment method for PDA in extremely low birth weight infants.

Keywords: premature infant, echocardiography, pda closure

O-015
Embolization of vascular abnormalities in children with congenital heart diseases using medtronic micro vascular plugs

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Background and Aim: Micro Vascular Plug (MVP) is one of the most recent addition to the armamentarium for peripheral embolization. Data on its use in congenital cardiology is scarce. We describe and evaluate our experience with MVP for embolization of vascular abnormalities in children with congenital heart diseases (CHDs).
Methods: Medical records of children with CHDs who had embolizations with MVPs between April 2015 and September 2020 were reviewed. Immediate, and follow-up data were assessed.
Results: 153 patients underwent 172 procedures during which 240 embolizations using 259 MVPs were attempted. Median age and weight were 34.9 months (IQR, 4-75 months) and 12.5 kg (IQR, 4.8-19.4 Kg), respectively. Targets were abnormal systemic arteries (n = 163), patent arterial ducts (n = 26), venous (n = 45), and coronary-cameral fistulas (n = 6). Median vessel diameter was 3.3mm (IQR, 2.5-4 mm), ranging from 1.4 to 9 mm. Veins were larger than arteries (2.3 mm> 2.1 mm, p<0.01).

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MVPs were implanted in narrower diameters compared to the manufacturer's recommendations, particularly for the MVP-5Q and MVP-7Q. Compared to veins, solitary implanted MVPs in arteries were additionally oversized at 12.4%, 5.1%, and 7.9% for MVP-3Q, 5Q, and 7Q, respectively. Additional occlusion material (16.7%) and 2 MVPs (5.8%) were needed at the same site for complete closure. Implantation and procedure success rates were 99.2% and 96.7%, respectively. Five complications were treated percutaneously (n = 4), and surgically (n = 1).
Conclusion: The safety and efficacy of vascular embolization using MVPs were demonstrated in the largest cohort of children with CHDs and a variety of clinical settings. Immediate and stable closure is obtained with a single device in most cases. Based on our experience, we present a detailed device selection protocol according to vessel type, site, and diameter to achieve intended outcomes.

Keywords: congenital heart defect; device closure; microvascular plug; vessel embolizations; children

	Single MVP-3Q	Oversizing*	Single MVP-5Q	Oversizing*	Single MVP-7Q	Oversizing*
Unconstrained device diameter (mm)	5.3		6.5		9.2	
Device nominal diameter (mm)	3		5		7	
IFU target vessel diameter (mm)	1.5 - 3		3 - 5		5 - 7	
Overall embolized vessel diameter (mm), median (IQR)	2.1 (1.8, 2.4)	42.8%	3.2 (2.6, 3.7)	56.2%	4.2 (3.9, 4.7)	66.7%
Arterial vessel diameter (mm), median (IQR)	2.1 (1.8, 2.3)	42.8%	3.1 (2.6, 3.6)	61.3%	4.1 (3.7, 4.4)	70.7%
Venous vessel diameter (mm), median (IQR)	2.3 (1.9, 2.5)	30.4%	3.2 (2.8, 3.5)	56.2%	4.3 (4.1, 4.6)	62.8%
Institutional recommended target vessel diameter (mm)						
Arterial vessels	1.8 - 2.5		2.5 - 3.5		3.5 - 4.5	
Venous vessels	2 - 2.8		2.8 - 4		4 - 5	

IFU: Instructions for use
*Compared to device nominal diameter

Fig. 1. Oversizing percentage of implanted MVP according to device size and vessel type

O-016

Transcatheter occlusion of PDA in extremely premature infants weighting less than 1200 g: a single-center experience with the micro vascular plug

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Background and Aim: Patent ductus arteriosus (PDA) in premature infants continues to be a significant problem contributing to both morbidity and mortality. Surgical ligation and medical therapy both have their drawbacks. The aim of our study is to describe a single center experience with an endovascular occlusion device Medtronic Micro Vascular Plug (MVP) for transcatheter PDA closure in extremely preterm infants.
Methods: Between March 2018 and November 2021, 44 percutaneous PDA closures were performed in our center on infants ≤1.2kg. 45% of these infants were less than 1kg and 9% were less than 800g. Procedural details, complications, and short-term outcomes were recorded.
Results: 35 premature infants ≤1.2kg underwent transcatheter PDA closure using the MVP device and 9 utilizing the Piccolo™ device. The gestational age and birth weight were 24.0 ± 0.9 weeks (range 22-26) and 651 ± 100 g (range 380-800). The mean weight and age at the time of intervention were 1.0 ± 0.13 kg (range 700-1200) and 39 ± 10.2 days (range 20-67). Mean PDA diameter was 2.8 ± 0.7 mm. 75% of the ducts were described as tubular in nature with 8 type F ducts by the Santhanandam modification of the Krichenko classification. All MVP devices were deployed via a 4F Glidecatheter in prograde fashion without arterial access. Fluoroscopy and echocardiography were utilized for guiding device placement. Mean fluoroscopy time and radiation dose were 10.8 ± 10.5 min and 37.0 ± 158 mGy, respectively. Heparin was not given due to concerns for intracranial hemorrhage. Complete PDA closure was achieved in all infants. One MVP device had to be removed and replaced due to aortic obstruction (AO) and one Piccolo device had to be removed and replaced with an MVP device due AO. There were no major complications related to the procedure or noted during short-term follow-up (mean 85.4 ± 120 days).
Conclusion: This study demonstrates that transcatheter PDA closure with the MVP device can be successfully performed in extremely preterm infants weighting less than 1.2 kg. using the MVP with a high success rate and a low incidence of complications. Transcatheter PDA closure continues to be an excellent alternative to surgery.

Keywords: Extreme prematurity, patent ductus arterioles, transcatheter closure, micro vascular plug

O-017

Assessment of computer-simulated stent implantation and virtual reality as tools to plan interventional treatment of aortic coarctation

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Background and Aim: A number of studies on using both three-dimensional printing and virtual models in analysing aortic coarctation has been published, yet none of them assesses virtual modelling as a planning tool in a blind retrospective analysis. The aim of our study was to assess the usefulness of virtual modelling and virtual reality in planning interventional treatment of aortic coarctation.
Methods: The study involved 20 CT scans performed prior to the interventional treatment of aortic coarctation, which were then processed by two medical students in Materialise Mimics in order to produce a virtual three-dimensional model of the narrowed aorta. The models were subsequently exported to Materialise 3-Matic in which, based on the diameter and the length of the stenotic segment, a group of possible stents was simulated. The complete models were then assessed in Mimics Viewer using virtual reality headset in order to choose an optimal stent, which was later compared with the actually used stent.
Results: A group of stents ranging from 1 to 6 was simulated in each virtual model. A stent with identical diameter was proposed in 11 cases and in the remaining the difference did not exceed 2 mm. The length comparison was expressed as a ratio of the simulated to actual stent length. In 6 cases the length ratio ranged between 95-105%, in 5 cases between 80-94.9% and in the remaining 9 cases the difference was more than 20%. The location of the proximal end of the stent was described by its relation to the left subclavian artery and it was concordant between the simulation and reality in 14 cases. A complete comparison can be seen in Table 1.
Conclusion: The method of computer modelling provided a satisfactory success rate of predicting the possible stents to use during procedure. The differences in chosen stents may have been caused by lack of experience in Interventional Cardiology of the main researchers, the lack of availability of certain stents in the Cardiovascular Interventions Laboratory at the time of the procedure, the lack of information about the diameter of the vascular access and differences in dimensions measured on the model, CT and angiography.

Keywords: coarctation of the aorta, stenting, 3D-modelling

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Group	Concordant SA	Discordant SA
Concordant stent	2	1
Stent with identical diameter and length difference up to 20%	2	0
Stent with diameter difference up to 2 mm and length difference up to 20%	5	1
Proposed stent did not meet the previous criteria	5	4
Identical diameter, length difference over 20%	4	2
Diameter difference up to 2mm and length difference over 20%	1	2

Table. 1 – Distribution of stents in the groups. Concordant SA – relation to the left subclavian artery was concordant, Discordant SA – the relation to the subclavian artery was discordant – the modelled stent was located either superior or inferior to the actual stent

O-018

First experiences with myval transcatheter heart valve system in conduit dysfunction and native rvot with severe pulmonary regurgitation

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Background and Aim: The rate of morbidity and mortality related to pulmonary regurgitation and pulmonary stenosis are big concerns after the surgery for CHD. Percutaneous pulmonary valve implantation has been established as a less invasive technique compared to surgery with promising results according to long-term follow-up of the patients. There are only two approved valve options for percutaneous pulmonary valve implantation until now, which are Melody and Sapien. Both valves have limitations and do not cover entire patient population. Therefore, the cardiologists need more options to improve outcomes with fewer complications in a such promising area. Herein, we present a case series applying for pulmonary position in conduits and native right ventricular outflow tract of a new transcatheter valve system Myval® which is designed for transcatheter aortic valve implantation procedures. This is the first patient series in which the use of Myvalv in dysfunctional RVOT is described, after surgical repair of CHD.

Methods: We present our experiences with twelve cases between 8 and 34 years of age using Myval for the pulmonary position between June 2020 and November, 2021. The demographic, clinical characteristics, and pre-procedural MRI findings,procedural details of the patients were demonstrated.

Results: In all patients, the stent implantation was preceded the valve implantation and the valve implantation was performed successfully. In two patients with native right ventricular outflow tract, the stent and valve were implanted in the same session.The valve size was 23 mm for the patients with the conduit, 29 mm for the two cases, and 26 mm for the four cases with native right ventricular outflow tract. There is no valvular leakage immediately after the procedure by TTE and angiography. The patients were examined by TTE, ECG, and chest X-ray before the discharge and in the first 3 and 6 months. No dysrhythmia was detected. The valve functions are also competent after the sixth month.

Conclusion: In conclusion, this study demonstrates our first experiences with Myval® in the pulmonic position for both conduit and native right ventricular outflow tract. Regarding this experience, Myval® is a clinically feasible, safe, and effective system for use in the pulmonic position.

Keywords: Pulmonary valve implantation RVOT Conduit dysfunction Pulmonary regurgitation

Adult Congenital Heart Disease

O-019

Pulmonary hypertension in adults with congenital heart disease: updated and extended results from the international compera-chd registry

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Background and Aim: Pulmonary arterial hypertension (PAH) is a common residuum or complication in patients with congenital

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heart disease (CHD). Because clinical-trial data on patients with PAH associated with CHD (PAH-CHD) remain limited, registry data on the long-term course and management of these conditions are essential. The aim of this analysis was to provide updated information from the COMPERA-CHD registry on current management strategies for adults with PAH-CHD, based on real-world data.
Methods: The prospective international pulmonary hypertension registry COMPERA has since 2007 enrolled more than 10,000 patients. COMPERA-CHD is a subregistry for patients with PAH-CHD enrolled in 62 specialized centers in 12 countries.
Results: 769 patients with PAH-CHD were included into COMPERA-CHD from January 2007 through September 2020. At the last followup in 09/2020, patients had either post-tricuspid shunts (n = 359), pre-tricuspid shunts (n = 249), complex CHD (n = 132), congenital left heart or aortic valve or aortic disease (n = 9), or miscellaneous other types of CHD (n = 20). The mean age was 45.3 ± 16.8 years; 512 (66%) were female. The mean 6-minute walking distance was 369 ± 121 m, and 28.2%, 56.0%, and 3.8% were in WHO functional class I/II, III or IV, respectively (12.0% unknown). Compared with the previously published COMPERA-CHD data, after 21 months of followup, the number of included PAH-CHD patients increased by 91 (13.4%). Within this group the number of Eisenmenger patients rose by 39 (16.3%), the number of "Non-Eisenmenger PAH" patients by 45 (26.9%). Currently, among the 674 patients from the PAH-CHD group with at least one followup, 450 (66.8%) received endothelin receptor antagonists, 416 (61.7%) PDE-5 inhibitors, 85 (12.6%) prostacyclin analogues, and 36 (5.3%) the sGC stimulator riociguat. While at first inclusion in the COMPERA-CHD registry, treatment was predominantly monotherapy (69.3%), this has shifted to favoring combination therapy in the current group (53%). For the first time, the nature, frequency, and treatment of significant comorbidities requiring supportive care and medication are described.
Conclusion: While individual therapy was dependent on the underlying CHD, there was an overall trend towards more aggressive treatment strategies and combination therapies. In the future, particular attention must be directed to the "Non-Eisenmenger PAH" group and to patients with complex CHD.

Keywords: Congenital heart disease, Eisenmenger syndrome, Pulmonary hypertension, Registry, Targeted treatment

O-020

Neuropsychological and structural brain abnormalities in adults with D-transposition of the great arteries: the boston circulatory arrest study

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Background and Aim: Neurocognitive impairments are frequent in congenital heart disease. The Boston Circulatory Arrest Study (BCAS) is a randomized trial comparing the developmental and neurological outcomes of newborns born with d-transposition of the great arteries (d-TGA) who underwent the arterial switch operation (ASO) with predominantly either deep hypothermic circulatory arrest (DHCA) or continuous low-flow bypass (LFBP) as the method of vital organ support. We report the neuropsychological and structural brain MRI findings of this cohort at adulthood.
Methods: Adults with d-TGA enrolled at birth in the BCAS study underwent a neuropsychological evaluation including IQ, executive function, social cognition, and memory functioning. A structural brain MRI was conducted on a Siemens Skyra 3T with a 64-channel head coil. Focal and multifocal brain abnormalities were recorded including brain mineralization/iron deposits and abnormal T2 hyperintensities (≥3mm) in subcortical grey, periventricular, and deep white matter. d-TGA outcomes were compared to population norms or to a control group.
Results: Eighty-seven adults (28.9 ± 1.3 y, mean age ± SD) participated. Their mean scores were not significantly lower compared to population norms except for working memory (9.2 ± 2.6, p = 0.01), visual social cognition (9.1 ± 2.9, p = 0.01), and reading comprehension (97.2 ± 10.8, p = 0.02). Adults who had been randomly assigned to predominant DHCA vs. LFBP during the neonatal ASO had lower scores on working memory (8.7 ± 2.6 vs. 9.8 ± 2.6, p = 0.04), cognitive flexibility (9.2 ± 2.9 vs. 10.8 ± 2.4, p = 0.01), and social cognition (25.9 ± 5.3 vs. 28.0 ± 4.4, p = 0.05) after adjustment for childhood socioeconomic class and ventricular septal defect status. Structural brain MRI findings included data from 69 adults with d-TGA and 38 controls. Compared to controls, the d-TGA group had greater prevalence of focal and multifocal abnormalities including brain mineralization/iron deposits (41% vs. 3%) and T2 hyperintensities (28% vs. 3%). Congenital brain abnormalities were also more common in the d-TGA group (13% vs. 0%).
Conclusion: Adults with d-TGA present with average neuropsychological functioning except for vulnerabilities in working memory, social cognition, and reading abilities. Structural brain abnormalities are prevalent. Adverse sequelae of DHCA extends to adulthood in higher-order neurocognitive areas. Follow-up into later adulthood should evaluate the risk of early neurodegenerative processes.

Keywords: d-transposition of the great arteries, arterial switch operation, neuropsychological outcomes, MRI brain abnormality, longitudinal cohort

O-021

Ebstein's anomaly: functional outcomes after cone reconstruction

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Background and Aim: Ebstein's Anomaly (EA) is a rare disease of the tricuspid valve. In the past decade, the Cone repair was established as the preferred surgical procedure for EA. Numerous studies showed good mid-term outcome regarding tricuspid valve competence. However, there is a knowledge gap concerning exercise performance and right ventricular remodeling.
Methods: Between 01/2010 and 12/2019, 48 patients underwent a Cone repair at our center. We excluded one patient due to in-hospital death and two patients due to missing follow-up, resulting in a study population of 45 patients. Mean age at operation was 30.0 ± 18.3 years, 29 patients (64%) were female. Indication for operation

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was a relevant tricuspid regurgitation (TR) in combination with clinical symptoms, increasing right ventricular (RV)-volume or deterioration in exercise capacity. We analyzed postoperative changes in (1) tricuspid valve competence, (2) peak oxygen uptake (peakVO2) assessed by cardiopulmonary exercise test and (3) end-diastolic volume of the right-ventricle (EDV-RV) and antegrade net stroke volume of the right-ventricle (net-SV-RV) assessed by cardiac magnetic resonance imaging (volumetry and flow measurement).
Results: Mean follow-up was 5.0 ± 2.8 years. At the end of follow-up, 39 patients had none/mild TR, five patients had moderate TR and one patient had severe TR. PeakVO2 was 23.4 ± 8.7 ml/kg/min preoperatively, and 23.8 ± 5.8 ml/kg/min postoperatively (paired T-test p = 0.79). During follow-up PeakVO2 did not change significantly (0.14 ± 0.19ml/kg/min increase per year, p = 0.45). EDV-RV decreased significantly from 300 ± 137 ml preoperatively to 211 ± 96 ml postoperatively (paired T-Test p<0.001) and remained stable during follow-up (0.9 ± 3.8 ml increase per year, p = 0.81). Net-SV-RV increased significantly from 58 ± 21ml preoperatively to 70 ± 22ml postoperatively (paired T-test p = 0.014) and remained stable during follow up (1.6 ± 1.3ml increase per year, p = 0.22).
Conclusion: The Cone repair results in a decrease in tricuspid valve regurgitation. Objective exercise capacity did not improve, however the right ventricle decreased in size and had an improved antegrade stroke volume.

Keywords: Congenital Heart Disease, Ebsteins Anomaly, Tricuspid Valve, Heart Surgery, Cone reconstruction

O-022

The burden of late thrombo-embolic complications following fontan palliation among polish adolescent patients – multicenter study

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Background and Aim: Thromboembolic events (TE) are a significant cause of morbidity among patients post Fontan operation (FO). The purpose of the study was to describe prevalence of TE complications and characterize the clinical profile of Polish Fontan patients at risk of TE.
Methods: We performed retrospective multicenter baseline analysis of consecutive patients after FO who remained under a regular ambulatory care between 2018 and 2021 at three Adult Congenital Heart Disease(ACHD) departaments in Poland. Next we performed outpatient-based prospective observation of TE occurrence. TE complications, clinical, and imaging data was based on the medical records. Median time between FO and baseline evaluation was 22.1 ± 5.1 years.
Results: 91 post-FO patients (mean age 25 ± 6 years; 49.5% male); 53.8% on aspirin, 22.1% on VKA, 11.0% on NOAC and 13.2% not on any antithrombotic medication were analysed.
21 out of 91 patients (23.1%) had developed 24 TE complications since FO in the baseline analysis. Most frequent TE type was pulmonary embolism which was reported in 12 (13.2%) patients. Other thrombus locations included Fontan circuit in 6 (6.6%) cases, intracranial in 4 (4.4%) and peripheral vasculature in 2 patients (2.2%). Mean time since FO to first TE was 17.8 ± 5.1 years. Actuarial freedom from TE since FO at 5, 10, 15, 20, 25 and 30 years was 99.80%; 97.80%; 94.46%; 81.92%; 78.31% and 68.67% respectively.
Total follow-up duration was 2341 patient-months with a median of 31 (IQR: 28 - 33) months. Total amount of thrombo-embolic events during follow-up was 9 and occurred in 7 (7.7%) patients, including pulmonary embolism in 5 patients (5.5%); Fontan circuit thrombus in one (1.1%) and peripheral venous thrombosis in 3 cases (3.3%).
Patients complicated with TE were 50% male, in 28.6% with right type systemic ventricle, in 85.7% unfenestrated. TE during follow-up occurred in 14.3% of patients with a history of previous TEs and in 5.7% with no previous events. 3 (42.9%) TEs occurred on aspirin; 1 (14.3%) during secondary prophylaxis with VKA, 2 (28.6%) TEs occurred on NOAC therapy and 1 (14.3%) without prophylaxis.
Conclusion: In this multicenter analysis thromboembolic events were not rare in patients with Fontan circulation.

Keywords: thromboembolism, Adult Congenital Heart Disease, Fontan procedure

O-023

Long-term results in patients with repaired tetralogy of fallot: up to 38-year follow-up of 814 patients

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Background and Aim: To evaluate long-term outcome of tetralogy of Fallot (ToF) repair analyzing an unbiased country-wide surgically treated population with ToF.
Methods: Retrospective analysis of consecutive patients aged <18 years who underwent ToF repair from January 1979 through December 2016 at a single nation-wide pediatric cardiac center. ToF repair type was coded as transatrial (REP/TOF/TAT), transventricular (REP/TOF/TV) and transannular (REP/TOF/TAN) with or without monocusp (REP/TOF/MC). Death from any cause and need for surgical or catheter reintervention were considered as study endpoints.
Results: A total of 814 patients (male 56 %) was analyzed. Staged repair was performed in 17%. Early mortality (20/814, 2.46% patients) was confined to the early surgical periods. Late mortality was 3.27% (26/794 of patients). Probability of freedom from death was 95.5%, 94.3% and 93.1% at 10, 20 and 30 years after repair, respectively. Early surgical period (P = 0.013) and surgical/catheter reinterventions (P<0.001) were multivariable predictors of death. A total of 357 reinterventions was performed after initial repair in 188/814 patients (23.1%) with pulmonary artery revalvulation (142/814 patients, 17.4%) being most frequent. Probability of freedom from first reintervention was 87.7%, 73.9% and 57.1% at 10, 20 and 30 years after primary repair, respectively. Transannular repair was associated with the need for pulmonary artery revalvulation (P<0.001), transatrial repair carried higher risk of reinterventions for right ventricular outflow tract obstruction (P = 0.004).
Conclusion: In an unbiased nation-wide cohort ToF repair carried a favorable survival of >90 % at 30 years. Reinterventions were significant risk factors for mortality. Type of initial repair predicted the need for specific surgical or catheter reinterventions. (transannular repair is associated with the need for pulmonary artery

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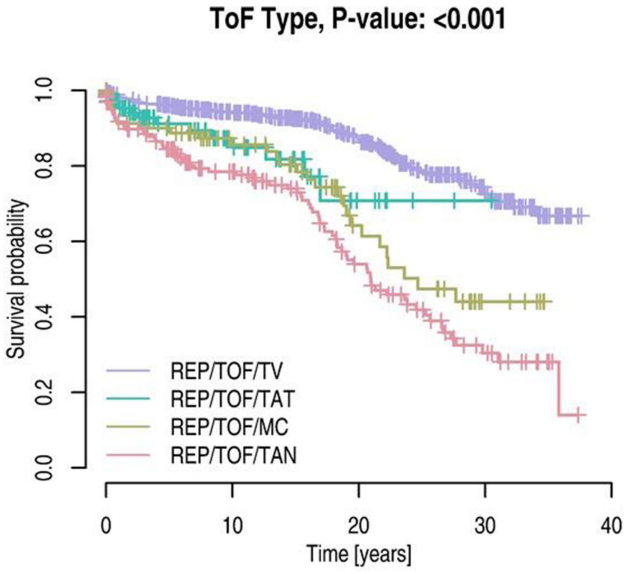
revalvulation whereas transatrial repair carries higher risk of reinterventions for right ventricular outflow tract obstruction).

Keywords: Tetralogy of Fallot, Outcome, Single-center study, Pulmonary valve replacement

Probability of freedom from any first surgical or catheter reintervention by the type of primary repair.



O-024

Medical rehabilitation in adults with congenital heart disease

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Background and Aim: The number of adults with congenital heart disease (ACHD) is steadily rising. In the long-term course, many of the affected patients require medical rehabilitation after interventional or surgical treatment of residua, sequels or complications of their congenital heart defect (CHD). However, up to now only scarce data exist about indication, performance and outcomes of cardiac rehabilitation in ACHD.
Methods: The course and outcome of rehabilitation after previous interventional or surgical treatment in ACHD was analyzed in a retrospective cohort study.
Results: 205 ACHD (54% female; mean age 34 ± 12 [16-68] years) with mild (23.9%), moderate (35.1%) or severe CHD (41,0%), of whom 32% had complex CHD, 21% right-ventricular outflow tract obstructions, 20% pre-tricuspid shunts, 18% left heart or aortic anomalies, 9% post-tricuspid shunts and 2% other congenital cardiac anomalies were included into analysis. The main indications for rehabilitation were a preceding surgical (92%) or interventional (3%) treatment of the underlying CHD immediately before rehabilitation. During rehabilitation, no severe complications occurred. The number of patients in function class I/II increased from 189 to 200 and decreased in class III/IV from 16 to 5. Cardiac medication could be reduced or stopped during rehabilitation in 194 patients, with the exception of ACE-inhibitors. There was an improvement of cardiovascular risk factors. While before the medical treatment 77% (n = 157) patients were capable of working, the number increased to 82% (168) at the end of rehabilitation. Throughout follow-up 9.3% (n = 19) of patients needed further cardiac interventions.
Conclusion: The current study provided for the first-time comprehensive data on the course of rehabilitation in a large cohort of ACHD after surgical or interventional treatment. The over-all outcome of ACHD after rehabilitation was uneventful and favorable. Further analysis is required to assess the clinical long-term outcome, the impact of rehabilitation on quality of life, disease coping and employment. The results of this study can serve as a benchmark for the development of specific rehabilitation programs in ACHD.

Keywords: Adults with Congenital Heart Disease, ACHD, Prevention, Rehabilitation

Nursing

O-025

Oral breastmilk is safe pre- and early post-operatively in babies with coarctation of the aorta

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Background and Aim: The incidence of necrotising enterocolitis (NEC) is significantly higher in infants with congenital heart disease, compared with healthy term infants, especially those with reduced cardiac output such as Coarctation of the Aorta (CoA) and hypoplastic aortic arch (HHA). The lack of well controlled, large trials in children with congenital heart defects has resulted in an absence of standardised evidence-based feeding guidelines. We therefore aimed to assess whether stable babies with suspected CoA/HHA could safely receive breastmilk pre- and post-operatively in preference to being nil by mouth.
Methods: All babies operated for CoA/HHA in 2013-2018 in a single tertiary unit were included in this audit. Feeding data was retrospectively reviewed including diagnosis of NEC, type of feeding, and length of time to full feeds. Based on these results, a feeding guideline was developed and after implementation the audit was repeated, reviewing babies operated for CoA/HHA from June 2020 to October 2021.
Results: 98 babies were included (74 pre feeding guideline and 24 post feeding guideline). 3 NEC cases (4%) were identified in the pre-guideline cohort, all of which received formula as main feed. None of the breastfed babies developed NEC. The feeding guideline developed therefore outlined the use of breast milk only (maternal or donor) if clinically appropriate to feed prior to and where possible until on full feeds after surgery before switching to formula. No babies developed NEC in the post-guideline group.
Pre-guideline 28% babies were not fed orally which was similar (21%) after guideline implementation. On average feeds started on day 2 post-operatively in both groups.

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Pre-guideline 46% babies were discharged on breastmilk, which increased to 63% post-guideline. After guideline implementation babies were on full feed on an average 2.5 days sooner with less variation (pre-guideline 8.5 ± 7 days vs post-guideline 6 ± 2 days; $p = 0.013$).

Conclusion: Feeding stable babies with CoA/HHA with breastmilk is safe in the pre- and early post-operative period. Implementation of a feeding protocol increases the number of babies discharged breastfeeding. Babies were safely on full feeds sooner and able to be discharged home sooner.

Keywords: Coarctation of the Aorta, feeding, breastmilk, necrotising enterocolitis, guideline

O-026

A mobile application to support parents of infants with severe cardiac disease

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Background and Aim: Every year approximately 125 infants are born in Norway with severe congenital heart disease (CHD). Around 10% of these children die during the two first years of life, and recent research documents that 17% of these children die from unexpected deaths after a gradual deterioration at home. Parents of infants with cardiac disease have shown higher levels of anxiety, depression and distress compared to other parents. Many parents find it difficult to recognize worsening of symptoms, and this uncertainty contributes to their burden. We report initial experiences of a mobile application, aiming to support the parents' understanding and management of their child's condition.

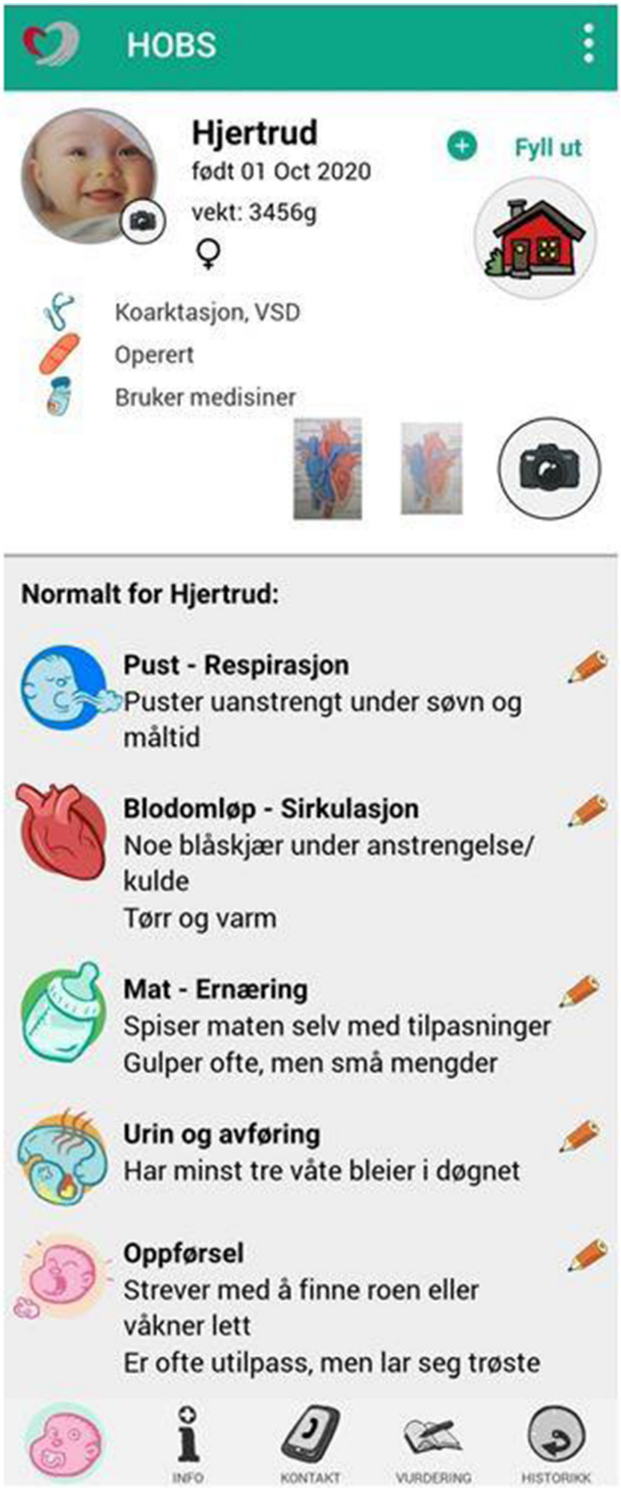
Methods: To meet the challenges of unexpected deaths and psychological distress in parents, Oslo University Hospital, has in close collaboration with parents and local health professionals, developed at mobile application called Heart OBServation app (HOBS). HOBS is a personalized decision support tool for parents that aims to increase parent's awareness of their infant's normal condition and appearance. It also helps them to assess deterioration, decide who and when to contact if necessary and what to report. We investigated the acceptance of HOBS in a pilot study of 10 families. We performed semi structured interviews at discharge and after one month and used thematic analysis to explore feasibility and acceptance of the application.

Results: Analysis revealed that HOBS was easy and intuitive to use. It increased awareness of normal appearance of the child and the personalized information was considered relevant and comprehensible. The assessment function was well received as a useful checklist and a help to understand what to look for. One mother (without proper training in use from the hospital) became stressed, and one family chose not to use the application. Some minor suggestions to revisions were proposed.

Conclusion: The mobile application HOBS was well received as a tool to provide tailored information, increase awareness of the child's normal condition and to support parents to assess worsening of symptoms. Some training is necessary to achieve these benefits. A controlled trial of HOBS as part of the service pathway is in progress.

Keywords: Congenital heart disease, mobile application, decision support tool, discharge preparation

HOBS screenshot



Example of personalized baseline of normal appearance of the child in HOBS

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Psychosocial

O-027

Infant growth trajectories post-surgery and over four months predict parental stress in parents of infants with complex congenital heart defects

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Background and Aim: Among the salient indicators associated with the mortality risk and morbidities in infants with complex congenital heart defects (CHD) are growth deficits in early life. These may be a major source of stress for parents of those infants, especially for those lacking partner's support. This study examined the associations between infant growth trajectories and parental stress following cardiac surgery, while testing the moderating role of parental partner's support.
Methods: A longitudinal study utilizing data from a previous clinical trial in which parents and infants were recruited from three pediatric cardiac centers in Northeastern America, and followed over the infant's hospital stay and over 4-months post-discharge. The sample included 136 parent-infant dyads, who had data on infant growth and parental stress measures. Data included demographics and clinical indicators, infant's daily weight measurements, and parental self-report surveys of stress and other psychosocial parameters. Analyses included latent class growth models and multivariable GEE models.
Results: We identified distinct growth trajectories- "stable around WAZ = 0" (37.5%), "maintaining WAZ>0" (8.8%), "partially-recovered" class (32.4%), and "never-recovered" class (21.3%). The first two classes were collapsed and referred to as the reference group. Multivariable GEE models revealed that parents of infants in the 'Never recovered' (OR = 4.78) and the 'Partly recovered' (OR = 2.47) groups were at a greater risk to experience higher stress levels, both at hospital discharge and after 4-months, compared to the reference group. A moderation analysis revealed that parents with low dyadic adjustment, whose infants 'Partly recovered', were at a greater risk to experience higher stress on all stress domains (OR = 2.63-5.45), both at the time of hospital discharge and after 4-month. Parents whose infants had 'Never recovered', were also at a greater risk to experience higher stress on the Parent Domain (OR = 4.91) at both time points, compared to the reference group.
Conclusion: Parents of infants with poorer growth paths, and especially those lacking partner's support, are at a greater risk to experience parental stress. Findings may be used to screen families at risk of poor illness adaptation and in the design of interventions to target parental stress in this vulnerable population.

Keywords: parental stress, infants, growth trajectories, complex cardiac defects

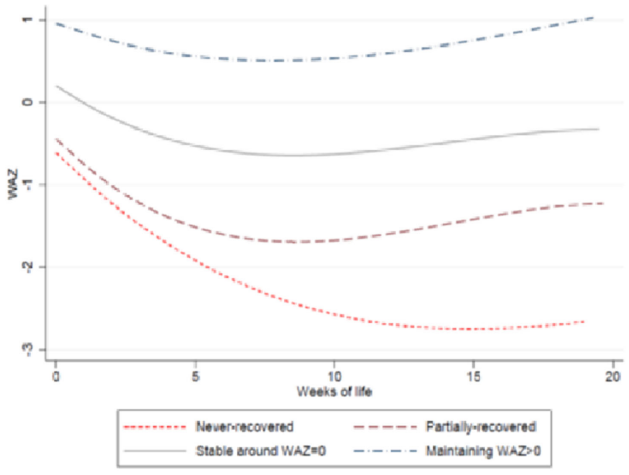

Figure 1 Infant growth trajectories identified in Latent Growth Trajectory Analysis, and used for further GEE modeling.

O-028

Longitudinal comparison of neurodevelopmental outcomes from 18m to 36m of children born preterm and children diagnosed with congenital heart disease

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Background and Aim: Children born preterm or diagnosed with congenital heart disease (CHD) have similar patterns of brain injury and dysmaturation, and are at risk for poorer neurodevelopment. Limited research has compared neurodevelopment of these groups and longitudinal outcomes remain unclear. This longitudinal study compared cognitive, language, motor, and behavior ratings of preterm children to children with CHD at 18 and 36-months of age.
Methods: 87 children born preterm (<32 weeks; n = 60) or with CHD (n = 27) were included and assessed at 18-and 36-months of age. Development was assessed using the Bayley Scales of Infant and Toddler Development-3rd Edition. Parents rated child behaviour using the Child Behavior Checklist (CBCL; 1.5-5 years). Three 2x3x2 repeated measures ANOVA with post-hoc tests compared group differences on the Bayley over time. A series of paired t-tests explored potential group and time differences in CBCL scores. Reliable change index (RCI) scores were calculated.
Results: There was a main effect of time for cognitive scores ($F(1,85) = 12.11$, $p<.001$, $\eta2 = .125$), with lower scores at 36-months for both groups. There was a time by group interaction for language ($F(1,73) = 8.868$, $p = .004$, $\eta2 = .108$) and motor scores ($F(1,75) = 8.725$, $p = .004$, $\eta2 = .104$). Language scores in the preterm group were stable but significantly increased from 18m to 36m in the CHD group. Motor scores in the preterm group significantly decreased over time but were stable in the CHD group. Using RCI, similar proportions of children demonstrated significantly low cognitive scores (19% CHD; 17% preterm). The preterm group had more children with significant declines in language (8%) and motor (17%) scores compared to the CHD group (0% and 7%; respectively). Significant group differences on behaviour ratings were found for withdrawn ($p = .019$) and internalizing problems ($p = .010$), with the preterm parent group endorsing higher symptoms, yet within normative ranges. Parents of preterm children endorsed fewer ADHD symptoms at 36m ($p = .047$), while parents of CHD children endorsed more symptoms at 36m vs. 18m ($p = .05$). Parents of preterm children endorsed lower scores on total problems over time ($p<.001$).

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Conclusion: Findings highlights the unique and shared cognitive and emotional changes in children born preterm with CHD in early development, which may inform targeted early interventions in the future.

Keywords: neurodevelopmental outcomes, behaviour, cardiology, preterm birth, congenital heart disease

Appendix

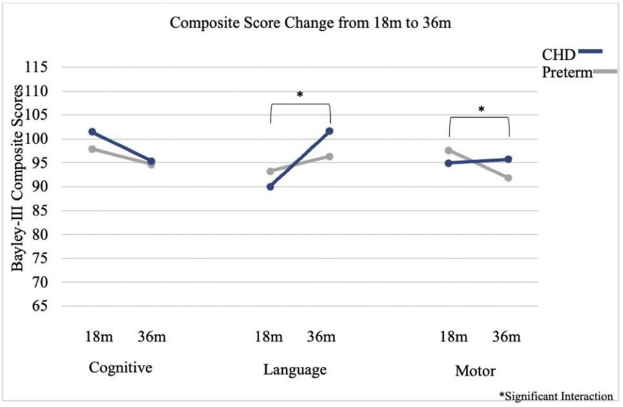

Figure 1. Group comparison of Bayley-III scores over time

This figure depicts the interactions and group comparisons of the cognitive, language and motor composite scores over time.

O-029

The prevalence of fatigue in children with congenital heart disease: is there a correlation with disease-specific factors?

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Background and Aim: The prognosis of children with congenital heart disease (CHD) has improved tremendously over the past decades through major advances in paediatric medicine; however, children who grow up with a chronic disease often face several challenges. Fatigue is a common and disabling symptom in childhood chronic disease. It appears to have a similar prevalence and presentation in several paediatric chronic disease groups, transcending diagnosis. Presumptively fatigue also presents similarly in children with CHD, however little is known about fatigue in these children. The aim of this study was therefore to investigate the prevalence of (severe) fatigue in children with CHD and the correlation with quality of life and disease-specific factors.
Methods: In this cross-sectional study, data were obtained from the PROactive cohort study (chronic diseased children). We examined the prevalence of severe fatigue, the effect of fatigue on health-related quality of life, and the effects of disease activity on fatigue severity among paediatric patients 2–18 years of age with CHD. Fatigue and health-related quality of life were assessed using the paediatric quality of life inventory multidimensional fatigue scale (PedsQL MFS) and generic core scales (PedsQL GCS), respectively. Linear regression analyses and an analysis of covariance were used to compare the fatigue scores with the scores obtained from a healthy control group. Data were adjusted for age and sex.
Results: 259 patients were included (55% participation rate, median age 8.9 (interquartile range 4.9-13.6), 57% boys). Paediatric patients with CHD reported significantly higher fatigue levels compared to healthy peers, with an 34% prevalence of fatigue and 16% of severe fatigue. Moreover, higher fatigue levels were inversely associated with health-related quality of life in all domains. Two disease-specific factors (number of procedures and exercise tolerance test results) were significantly correlated with fatigue. The disease-specific model explained 19% of the variance in fatigue.
Conclusion: Fatigue is common in children with CHD and its prevalence is corresponding to other paediatric chronic disease groups. In addition, severe fatigue negatively affected the patient's quality of life. The total explained variance of fatigue by disease-related factors is low. A biopsychosocial approach should be considered for management of fatigue in children with CHD.

Keywords: Paediatric, Congenital Heart Disease, Fatigue, Health-related Quality of Life; Disease-activity

Nursing

O-030

Motor development and physical fitness in children with congenital heart disease

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Background and Aim: Children have a basic need for physical activity. Frequently, heart disease means a limitation of the affected child's perceptual and motor experience. There are relatively few studies focusing on the motor development of children with congenital heart disease (CHD). We aim to assess the impact of CHD on the fundamental motor skills of children aged 6 to 9 years.
Methods: Children with CHD, of both sexes, aged 6 to 9 years were included. Exclusion criteria were: presence of other risk factors for motor development disorder, visual or auditory alterations, presence of extra-cardiac malformations or syndromes that could interfere with the study variables. The participants were divided into 3 groups according to the underlying disease and surgical status: corrected simple acyanotic CHD (group I); corrected cyanotic or complex acyanotic CHD (group II); uncorrected complex cyanotic CHD, i.e. after palliative surgery (group III). The assessment included the 6-Minutes' Walk-test; an interview with the children and their parents to collect clinical, demographic and life habits data; and at the end the Bruininks-Oseretsky motor proficiency test, 2nd Edition, was applied.
Results: Forty-four participants were recruited, 24 male and 20 female. Group I included 15 children, group II 12, and group III 17. In the Composite Motor Proficiency Score, Group I presented a score corresponding to the 48th percentile (34.51-61.76), Group II presented a score corresponding to the 25th percentile (10.06-40.25); finally, the participants in Group III present a score corresponding to the 5th percentile (2.54-7.32), much below the average for their age. Statistically significant differences between

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the 3 groups were documented for the different subtests assessed. A correlation was documented between Motor Proficiency and the results of the 6MWT, the impact being greater the more complex the disease.
Conclusion: This study documents that children with complex or palliated CHD present reduced motor competence, with limitations in fine and gross motor skills. These limitations, based on skills acquired since early childhood, present an additional negative effect on the physical fitness and muscle strength of individuals. These results point to the need for early intervention in the development of these patients.

Keywords: Motor Development; Physical Fitness; Children; Congenital Heart Disease

Interventional Cardiology

O-031
Transcatheter correction of sinus venosus atrial septal defect: from bench testing to clinical success

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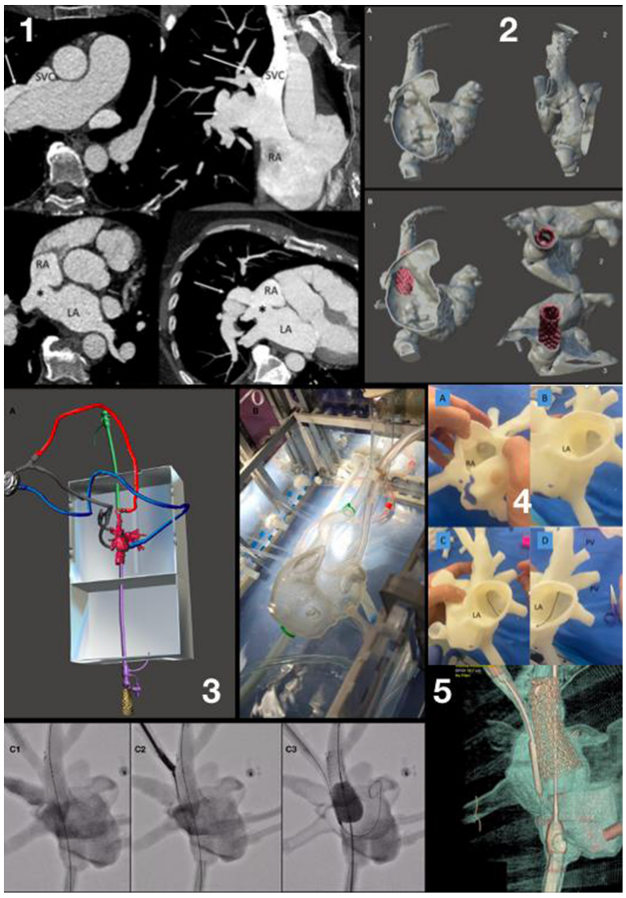
Background and Aim: Transcatheter correction of sinus venosus atrial septal defect (SVASD) has emerged as an alternative to open-heart surgery when the anatomic configuration is suitable. The three main challenges of the procedure consist in achieving complete shunt occlusion, maintaining APVR patency, and achieving stable stent implantation.For at-risk cases, we developed a step-by-step simulation program with virtual simulation, 3D printing, and hands-on simulation training (HOST) to assess procedure feasibility.
Methods: When we faced a complex anatomy, a 3D stereolithography (STL) model was electively segmented using cardiac CT (1); then, a created STL stent was merged with the SVASD STL to virtually simulate the procedure on screen (2).
If at risk, a 3D printed-model of the SVASD was created. Material was developed to produce similar echogenicity, radiotransparency, and distensibility to those of cardiovascular tissue.
To achieve bench-testing, the 3D-printed model was fixed in a container filled with radiotransparent and echogenic liquid and plugged to a pump-driven circuit to simulate the procedure in a catheterization laboratory, with close-to-reality conditions including transesophageal echocardiography (TEE), fluoroscopy, and angiography (3).
Results: In 6 cases, the virtual simulation confirmed the high risk of PV obstruction or residual shunting but did not rule out feasibility of transcatheter correction.
HOST permitted to test PV obstruction risk by inflating differents balloons in the SVC. When compliant balloon inflation produced a bulge that obstructed a PV, it gave an important warning to test the PV with a non-compliant one. However, when PV stenosis/obstruction was observed with a non-compliant balloon inflated at a similar diameter than the SVC diameter above the APVR, it made us choose to protect the concerned PV during the in vitro and then the in vivo procedure (3). In vitro stent implantation was also performed.
At the end of the bench-testing, dissection (4) and cone-beam CT (5) of the 3D model could confirm the final result of the in-vitro procedure.
In 2 years, we performed bench-testing in 6 cases before real percutaneous correction.
Findings, features and warnings were always comparable between in vitro and in vivo procedures.
Conclusion: Complex cases of SVASD percutaneous correction can be prepared and guided by hands-on simulation training.

Keywords: sinus venosus atrial septal defect - percutaneous correction - hands on simulation training - virtual simulation

Virtual simulation and bench testing of SVASD percutaneous correction



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Basic science, Genetics

O-032

Perinatal hypoxia aggravates occlusive pulmonary vasculopathy in rats later in life: association with epigenetic changes

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Background and Aim: Pulmonary arterial hypertension (PAH) is a progressive and fatal disease. Although the SUGEN5416/hypoxia rat model reproduces human PAH-like occlusive pulmonary vascular disease (PVD), this model is not fatal and PVD is not progressive or rather reversed. Since perinatal insults increase the risk of developing various adult-onset vascular diseases via epigenetic mechanisms, we tested the hypothesis that perinatal hypoxia, simulating perinatal insults, aggravates survival prognosis and occlusive PVD in SUGEN5416/hypoxia rats via such mechanisms.
Methods: Perinatal hypoxia rats were produced by exposure to hypobaric hypoxia before and after birth for 10 days. To induce occlusive PVD, 7-week-old adolescent rats were treated by SUGEN5416 injection and subsequent exposure to hypobaric hypoxia for 3 weeks. At the end of the experimental period for 8 weeks, right ventricular systolic pressure (RVSP), right ventricular hypertrophy (RV/LV+septum, RVH) and pulmonary vascular morphometric parameters (% occlusive or % plexiform lesions/total vessels; % medial wall thickness) were evaluated in 4 groups: SUGEN5416/hypoxia or control rats with/without perinatal hypoxia (n = 40). Baseline hemodynamic and morphometric analysis at 7 weeks of age was performed in 2 additional groups: perinatal hypoxia or normoxia group (N = 8). Proliferation, cytokine production and genome-wide DNA methylation profile were evaluated in cultured pulmonary artery smooth muscle cells (PASMCs) isolated from the baseline rats.
Results: In SUGEN5416/hypoxia rats, perinatal hypoxia reduced body weight gain and survival (60% vs 100%), and increased RVSP, ratio of RV/LV+septum and occlusive PVD ($p<.05$), which were associated with an increase in perivascular macrophages, expression of prepro-endothelin-1 mRNA and eNOS protein in the lung ($p<.05$) but not with impaired alveolarization or worsening right ventricular fibrosis. In PASMCs, perinatal hypoxia enhanced PDGF-BB-induced cell proliferation and phosphorylation of extracellular signal-regulated kinase as well as TNFα-induced expression of IL-6, monocyte chemotactic protein 1 and phosphorylation of p38 ($p<.05$). Such hyperproliferative and proinflammatory phenotypes were associated with altered DNA methylation status in several proliferation-related candidate genes.
Conclusion: We established the first animal model of the fatal and progressive PAH with occlusive PVD. Hyperproliferative and proinflammatory phenotypes of PASMC accompanied by altered DNA methylation status may contribute to the development of the fatally occlusive PVD.

Keywords: pulmonary hypertension, perinatal hypoxia, DNA methylation

Other

O-033

Kawasaki disease and the current SARS-COV-2 pandemic: rare lessons from a cohort of more than 1000 kawasaki patients in the netherlands

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Background and Aim: Kawasaki disease (KD) is a paediatric vasculitis with an unknown aetiology. The aim of this study was to assess the clinical course, treatment and cardiovascular outcomes in children with KD. The secondary purpose of this study was to make a comparison with the Kawasaki-like disease Multisystem Inflammatory Syndrome in Children (MIS-C), which is triggered by SARS-CoV-2.
Methods: In this observational cohort study, clinical information from KD and MIS-C patients was collected. Data were described and a multivariate analysis was performed to identify risk factors for coronary artery aneurysms (CAAs). Clinical characteristics between KD and MIS-C were compared using chi-squared and Mann-Whitney U tests.
Results: 1003 KD patients were included. The male-to-female ratio was 3:2, a majority of the patients were <5 years old (78.3%), treated with a single dose of intravenous immunoglobulin (IVIG) (90.8%) and treated promptly (<10 days) (81.7%). A second dose of IVIG was needed in minority of the patients (24.7%). The main complication of KD were CAAs and known risk factors (i.e., male, young age, delayed treatment) for CAAs were confirmed. A total of 35 MIS-C patients were included for the comparison. These patients were older than the KD patients ($P<0.0005$), more often had an incomplete KD presentation ($P<0.0005$). MIS-C patients mainly presented with acute cardiac dysfunction, with complete recovery after treatment.
Conclusion: KD and MIS-C are severe post-infectious inflammatory diseases. Due to the risk of cardiovascular complications, vigilance and prompt treatment are advised to reduce risk of cardiovascular complications.

Keywords: Kawasaki disease, Multisystem Inflammatory Syndrome In Children, coronary artery aneurysms, COVID-19

Clinical characteristics

Features	KD (n = 1003)	MIS-C (n = 35)	Significance (*p*-value) *
Male sex, n (%)	603 (60.5%)	22 (61.9%)	0.8
Age at disease presentation in years, median (ranges)	2.7 (0-19.7)	11.8 (0.2-19.4)	**< 0.0005**
Nationality, n (%)			
Dutch	408 (60.4%)	3 (20.0%)	
At least one Dutch grandparent	137 (20.3%)	3 (20.0%)	
Other (e.g., Surinamese, Moroccan, Chinese)	126 (19.3%)	9 (60.0%)	**< 0.0005**
Symptoms			
Fever ≥ 5 days (%)	854 (96.7%)	34 (97.1%)	0.9
Polymorphic rash, n (%)	769 (88.0%)	23 (65.7%)	**< 0.0005**
Conjunctivitis, n (%)	746 (85.3%)	25 (71.4%)	**0.03**
Cervical lymphadenopathy, n (%)	672 (78.6%)	20 (57.1%)	**0.003**
Mucosal changes, n (%)	756 (86.7%)	22 (62.9%)	**< 0.0005**
Extremities changes, n (%)	706 (81.4%)	14 (40.0%)	**< 0.0005**
Respiratory symptoms, n (%)		9 (25.7%)	N/A
Abdominal symptoms, n (%)		33 (94.3%)	N/A
Disease Presentation			
Complete, n (%)	705 (77.4%)	13 (41.9%)	
Incomplete, n (%)	206 (22.6%)	18 (58.1%)	**< 0.0005**
Laboratory values			
Hemoglobin, average (SD)	6.8 (0.9)	6.4 (1.0)	**0.01**
CRP, median (ranges)	120 (1-508)	212 (27-404)	**< 0.0005**
Leukocytes, median (ranges)	15.7 (2.4-46.0)	15.4 (4.4-38.4)	1.0
Thrombocytes, median (ranges)	392 (26-1403)	369 (61-836)	0.3
Albumin, average (SD)	32.5 (5.9)	28.9 (1.0)	**0.03**
Admissions to the ICU, n (%)	28 (3.2%)	20 (57.1%)	**< 0.0005**
Treatment			
IVIG, n (%)	842 (90.8%)	32 (91.4%)	0.9
< 10 days after symptoms, n (%)	624 (81.7%)	30 (93.8%)	0.08
>1 administration, n (%)	200 (24.7%)	7 (21.8%)	0.7
Corticosteroids, n (%)	78 (8.9%)	24 (68.6%)	**< 0.0005**
CAA in acute phase, n (%)			
No enlargement	671 (78.6%)	28 (82.4%)	
Z score ≥2.5-5.0	82 (9.6%)	5 (14.7%)	
Z score ≥5.0-10	40 (4.7%)	1 (2.9%)	
Z score ≥10	60 (7.1%)	0	0.3
Second disease presentation	7 (0.7%)	0 (0%)	0.6
Death due to Kawasaki disease /MIS-C	4 (0.4%)	0 (0%)	N/A
*χ^2-test for the categorical variables, t-test for continuous variables with a normal distribution and Mann-Whitney test for continuous values with a non-normal distribution			

Clinical characteristics of KD versus MIS-C patients

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Basic science, Genetics

O-034

The critical role of ER selective autophagy in response to doxorubicin-induced myocardial injury

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Background and Aim: Recent studies have suggested that endoplasmic reticulum-selective autophagy (ER-phagy) plays a cell-protective role by mediating degradation of damaged ER. However, the function of ER-phagy in cardiomyocytes and its molecular mechanisms remain to be elucidated. Given that various cardiac loads cause ER damage, known as ER stress, which contributes to the development of heart failure, we aimed to investigate whether ER-phagy acts as a compensatory system for protecting cardiomyocytes from ER stress-mediated injury.

Methods: ER-phagy activity was assessed using ss-RFP-GFP-KDEL, an ER-phagy reporter protein possessing an N-terminal ER signal sequence followed by tandem fluorescent proteins and ER retention signal. The H9c2 cells and cardiomyocyte-specific transgenic mice, both expressing ss-RFP-GFP-KDEL proteins, were utilized as ER-phagy reporter models. RNA interference with short harpin RNA (shRNA) or Gene-trap mutagenesis technique was performed to generate loss-of-function models of target genes. Assessment of phenotypes and molecular signaling pathways were carried out using immunoblotting, quantitative PCR, cell viability assays, immunocytochemical and histopathological analyses and cardiac ultrasonography.

Results: Fluorescent microscopic analyses and immunoblotting demonstrated that amino acid deprivation of ss-RFP-GFP-KDEL-transduced H9c2 (tfH9c2) cells markedly elevated the amount of RFP fragments, suggesting the increase in ER-phagy activity. 48 hours of food starvation induced ER-phagy in Tg-mice hearts, as well. Treatment with Doxorubicin (Dox), a cardiotoxic compound inducing ER stress, activated ER-phagy in tfH9c2 cells. Likewise, treatment with Dox (20 mg/kg) significantly induced ER-phagy in myocardium of Tg-mice compared with those of saline-treated ones. qPCR analyses revealed that treatment with Dox enhanced the expression of CCPG1, one of the ER-phagy receptors, in tfH9c2 cells (Control vs. Dox: 1.0 ± 0.05a.u. vs. 2.1 ± 0.2a.u.; $p<0.05$). shRNA-mediated silencing of CCPG1 in tfH9c2 cells resulted in the reduction of ER-phagy activity, accumulation of pro-apoptotic proteins including cleaved caspase 3 and deterioration of cell survival (Cell viability: Control vs. Knockdown: 79.0 ± 3.3% vs. 62.4 ± 2.7%; $p<0.05$) in response to Dox treatment. Consistently, CCPG1-hypomorphic mice developed more severe deterioration in systolic function against Dox treatment, than wild-type mice did (Ejection fraction: Wild-type vs. CCPG1-hypomorphic: 54.4 ± 5.0% vs. 47.2 ± 4.1%; $p<0.05$).

Conclusion: Our results suggest that ER-phagy plays a protective role in cardiomyocytes against Dox toxicity, possibly through CCPG1-mediated signaling (Figure).

Keywords: ER-phagy, ER stress, CCPG1, Doxorubicin cardiomyopathy

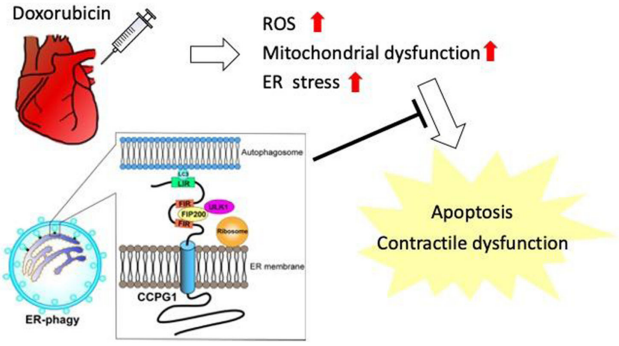

Figure 1 ER-phagy ameliorates Doxorubicin-induced myocardial injury

Interventional Cardiology

O-035

Percutaneous transhepatic periportal embolization of hepato-duodenal lymphatic fistulae as treatment for protein losing enteropathy after fontan

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Background and Aim: To determine early and medium term results of selective embolization of hepato-duodenal lymph vessels in Fontan patients with protein losing enteropathy (PLE).

Methods: Using ultrasound guidance, dilated lymph vessels in periportal position were percutaneously punctured with a 22G Chiba needle. Intralymphatic position was confirmed by water soluble contrast injection with drainage to hepatoduodenal fistulae. After flushing with 10% glucose solution, occlusion of hepato-duodenal lymphatics was effected by injection of 1-4 cc mixture 4-6/1 of Lipiodol/n-butyl cyanoacrylate (Histoacryl®).

Results: 16 patients with proven PLE were treated at age 16.8 ± 7.1 (range 6.0-38.8, median 15.2) years, 12.4 ± 5.6 (range: 2.7-28.8, median 12.0) years after the Fontan. Fontan palliation was performed at 4.6 ± 2.1 (range: 1.4-10.0, median 3.9) years; clinical PLE started 5.2 ± 3.6 (range: 0.9–35, median 3.5) years later. Procedural complications were limited (portal thrombus (4), abdominal discomfort in all for 24 up to 48 hours, transient cholangitis (1) and caustic duodenal bleeding/melena (1)). Five patients had after the initial procedure a lasting normalization of albumin with an ongoing follow-up of 2.0 ± 1.2 (range 0.2-3.7) years; no effect in 3 patients, some transient improvement in 8. A second procedure was performed in 8 patients 5.8 ± 4.7 (range 1.4-16.3) months later; a third procedure in 2 patients and a fourth

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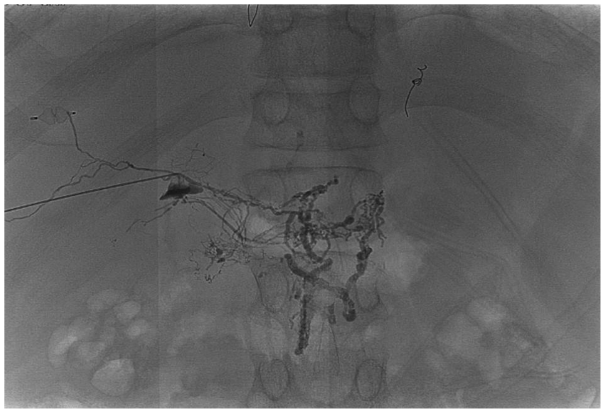
in 1 patient. Some additional procedures were performed in the failing group (fenestration (2), Fontan take-down (1), plication diaphragm (1)). When last seen 2.1 ± 1.1 (range 0.1-3.7) years after the initial procedure, 10/16 (62%) had "cured" PLE without treatment and significant improvement in quality of life (after 1-4 procedures), 2 patients had improved but still some PLE, 2 no effect, 1 take-down, 1 died. The procedure tended to be unsuccessful in cachectic patients, long standing advanced disease, and when ascites was present.

Conclusion: Embolization of hepatico-duodenal lymphatics is a promising technique in Fontan patients with PLE and already outclasses current medical strategies. However in 40% the embolization did not result in clinical result; the presumed leaks could not be reached from the periportal region and will require another diagnostic and therapeutic approach, especially when ascites is present. Larger series with longer follow-up are needed.

Keywords: protein losing enteropathy, lymphangiography, lymphatic embolization, n-butyl cyanoacrylate, Fontan

RX of hepato-duodenal region at end procedure

RX of hepato-duodenal region at end procedure



Other

O-036

Low- versus high-concentration intravenous immunoglobulin for children with kawasaki disease in the acute phase

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Background and Aim: Few studies have compared the effects of low-concentration (5%) and high-concentration (10%) intravenous immunoglobulin (IVIG) preparations for patients with Kawasaki disease (KD) in the acute phase. The purpose of this study was to compare outcomes between low- and high-concentration IVIG preparations in children with KD, using a national inpatient database in Japan.

Methods: We used the Diagnostic Procedure Combination database to identify patients with KD treated with IVIG from April 2012 to March 2020. We identified those receiving high- and low-concentration IVIG preparations as an initial treatment. The outcomes included the proportions of patients with coronary artery abnormalities (CAAs) and IVIG resistance, length of stay, and medical costs. Propensity score-matched analyses were conducted to compare the outcomes between the two groups. Instrumental variable analyses were performed to confirm the results.

Results: We identified 48,046 patients with KD and created 4:1 propensity score-matched pairs between the low- and high-concentration IVIG groups. There was a significant difference in the percentage with IVIG resistance between the two groups (20.6% vs 24.1%; risk difference, 3.5% [95% confidence interval, 2.3-4.7]; p <.001). However, there was no significant difference in CAAs (1.6% vs 1.6%; risk difference, 0.013% [95% confidence interval, −0.34 to 0.37]; p = .953). The instrumental variable analyses showed similar results.

Conclusion: The proportion of CAAs did not differ significantly between those receiving low- and high-concentration IVIG. To confirm the results of this study, prospective studies adjusting for duration of IVIG administration and duration of observation are needed.

Keywords: Kawasaki disease

Fetal Cardiology

O-037

Fetal complications associated with fetal cardiac interventions: experience of 205 procedures

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Background and Aim: Some cardiac defects may progress during fetal life and fetal cardiac interventions (FCI) were introduced to change the natural history of fetuses with critical aortic stenosis (CAS), critical pulmonary stenosis (CPS), pulmonary atresia with intact ventricular septum (PAIVS), or HLHS with restrictive or intact atrial septum (IAS). The aim of this study is to assess the overall success rates and risks of FCI at our institution.

Methods: The local FCI database was retrospectively reviewed for all fetuses who underwent fetal cardiac interventions (FCI) in our center since 2000. Patient's charts were analyzed with regards to complications and fetal outcome including intrauterine death (IUD) or prematurity.

Results: 205 fetal cardiac interventions in 160 patients have been performed since October 2000: 59 fetal pulmonary valvuloplasties (FPV), 4 atrial septostomies and 6 atrial septum stents, and 136 fetal aortic valvuloplasties (FAV). The median gestational age at the time of intervention was 27+2 weeks (21+4 to 38+3) for all interventions. Technical success-rates per fetus improved significantly for patients undergoing FCI for CAS (78% (39/50) early era vs. 97% (60/62) recent era; p = 0.0026), whereas FCI success-rates for patients with CPS/PAIVS did not show significant changes (71% (10/14) early era vs. 89% (25/28) recent era; p = 0.1967). The most common complications needing treatment for all FCI were bradycardias (36%) and pericardial effusions (11.7%). Bradycardias were more likely to occur during FPV than during FAV (47% vs. 32%, p = 0.0531), whereas thrombus-formation occurred more often during FAV (14% vs. 1.7%, p = 0.0087). FAV procedure related (p.r.) mortality improved over time (12% (6/50) vs. 5.9% (5/85), p = 0.3282), however not

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significantly. There were no significant maternal complications during the entire study period.
Premature deliveries occurred in 22.8% percent of all patients with 7.3% of patients being born before 34+0 weeks, thus being comparable with the natural prematurity rate of 26% (shown for patients with CAS).
Conclusion: Complications during fetal cardiac interventions must be expected. Therefore, an experienced team is required for an adequate management and acceptable low mortality rates.

Keywords: Fetal cardiac interventions, critical aortic stenosis, PAIVS, HLHS

Overview of complications of 205 procedures

Table	Overview of complications of 205 procedures				
Complications	IUD P.r.	IUD	Thrombus-formation	Bradycardia*	Pericardial-effusion*
Total (N=205)	16 (7.8%)	6 (2.9%)	20 (9.7%)	74 (36%)	24 (11.7%)
FAV (N=136)	11 (8%)	4 (2.9%)	19 (14%)	44 (32%)	14 (10%)
FPV (N=59)	3 (5%)	1 (1.7%)	1 (1.7%)	28 (47%)	8 (13%)
IAS Stenting (N=6)	1	0	0	1	1
Atrioseptosomies (N=4)	1	1	0	1	1

FAV, fetal aortic valvuloplasty; FPV, fetal pulmonary valvuloplasty; IAS, interatrial septum; IUD, intrauterine death; P.r., procedure related;

*Only bradycardias and pericardial effusions which required treatment are listed

O-038
Short term outcomes after prenatal detection of right aortic arch

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Background and Aim: The detection rate of a fetal right aortic arch (RAA) has increased after the introduction of the three-vessel view (3VV) and three-vessel trachea view (3VTV). RAA in the absence of other intra-cardiac and extra-cardiac abnormalities may be an innocent finding, but may sometimes require surgery. Which children will require surgery and at what age after a prenatal diagnosis of RAA is unknown.
Methods: A retrospective multicentre cohort study between 2007 and 2020 in three academic fetal medicine units in the Netherlands. All fetuses diagnosed with isolated RAA and one year follow-up were included. Cases with major intra-cardiac anomalies, associated extra-cardiac anomalies and genetic defects were excluded.
Results: We included 96 fetuses, 83 were suspected for RAA and 13 for a double aortic arch (DAA). A left-sided arterial ductus was observed in 53(64%), a right-sided duct in 7(8%) and not described in 23(28%) cases. In 39(41%) an abnormal origin of the left subclavian artery (ALSA) was described. Two pregnancies were terminated.
Postnatal echocardiography was concordant with the prenatal diagnosis in 84(87%) cases. Discordant findings were: Left aortic arch (n = 4); DAA not RAA (n = 4); RAA not DAA (n = 4). The median duration of follow-up was 4 years [1-11]. Respiratory symptoms or dysphagia occurred in 22% of the cases with a median onset at 9 months. 41(49%) children underwent a bronchoscopy and/or CT-scan (n = 25 and 28 respectively, of which 12 had both). Surgery was performed in 19(20%) children at a median age of 14 months [0-43], of which 6/19 (32%) had a DAA. The remaining 13/19 (68%) all had a left sided duct and ALSA. Only one child, with a DAA, required surgery in the first week of life due to an obstructive stridor.
Conclusion: The prognosis of children with a prenatal diagnosis of RAA is good with a low risk of acute postnatal problems. A delivery in an hospital with paediatric cardiothoracic surgery service may only be indicated in cases with suspected DAA. Expectant parents may be informed that presently 20% of the children needed surgery in the first 3 years of life, and rarely due to acute respiratory distress.

Keywords: prenatal screening, fetal echocardiography, congenital heart defect, right aortic arch

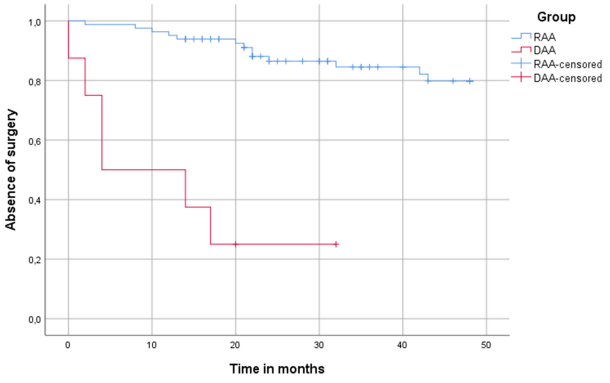

Figure 1. Time until surgery presented in a Kaplan-Meier curve

O-039
The contemporary surgical outcomes of congenital heart disease in the UK and its influence on the delivery of counselling and patient experience

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Background and Aim: Surgical outcomes of congenital heart diseases have been improving. Published evidence is outdated and misleading. We aimed to develop a reliable, and understandable information resource on the contemporary surgical outcomes of Congenital Heart Disease (CHD) on 10 selected anomalies in the United Kingdom (UK).
Methods: The 30-day and one-year surgical outcomes of 10 major CHD from the National Institute of Clinical Outcomes and Research UK (NICOR) database were translated into individual survival trendlines. User-friendly folders were created, distributed to healthcare professionals and patients. A feedback survey was conducted to prospectively evaluate the impact of the information on the efficacy of counselling and patient satisfaction.

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Results: 30-Day and 1-Year post-surgical survival rates of 10 selected CHDs have improved significantly over the past 16 years. 30-Day survival rates increased from an average of 93.91% between 2000-2005 to 96.68% between 2010-2015, whilst the 1-year survival rates increased from an average of 87.87% between 2000-2005 to 90.94% between 2010-2015. When the survival information was presented, 84.2% of healthcare professionals and 75% of patients reported an improvement in their knowledge. 100% of patients found the information pack useful, improving their overall experience. 94.2% of healthcare professionals and 100% of patients would recommend this to others. Feedback for improvement included display of numerical data, inclusion of additional anomalies and uploading the information pack onto online platforms.
Conclusion: Availability of this online national database on the surgical outcomes of CHD would allow medical practitioners and patients to make more informed, shared decisions regarding pregnancy and the treatments available. Provision of such information could better equip healthcare professionals and generate better patient experience.

Keywords: Counselling, Surgical Outcome, Congenital Heart Disease

Surgical Outcomes of CHD from 2000-2015

Congenital Heart Disease	2000-2005			2010-2015			Comparison between (2000-2005) vs (2010-2015)		All Years (2000-2016)		
	N (number of procedures)	30-Day Survival Rate (%)	1-Year Survival Rate (%)	N (number of procedures)	30-Day Survival Rate (%)	1-Year Survival Rate (%)	Statistical Significance using Wilcoxon Signed-Ranks Test		N (number of procedures)	30-Day Survival Rates	1-Year Survival Rate (%)
Vascular Ring Procedure	156	99.36%	96.30%	356	98.88%	94.89%	*30-Day Survival (α<0.05)* P=0.009	*1-Year Survival (α<0.05)* P=0.009	847	99.06%	94.80%
AVSD (complete) repair	772	97.41%	90.21%	1090	98.90%	93.02%			2643	98.15%	92.11%
AVSD (partial) repair	443	98.65%	96.42%	474	99.58%	98.37%			1268	99.21%	97.65%
Fontan Procedure	1077	95.45%	91.33%	1249	98.80%	96.15%			3118	97.75%	94.77%
Norwood Stage 1 Procedure	517	79.69%	62.20%	690	88.41%	67.77%			1749	84.96%	66.16%
Arterial Switch (for isolated transposition)	878	96.47%	94.42%	839	98.69%	96.07%			2432	97.86%	95.64%
Arterial Switch + VSD closure	224	93.30%	87.80%	349	97.99%	95.49%			844	96.56%	93.05%
VSD repair	1933	98.97%	96.81%	2091	99.76%	98.67%			5668	99.49%	97.81%
Truncus Arteriosus	162	92.60%	83.80%	205	93.66%	92.26%			515	93.40%	88.07%
Isolated Coarctation/Hypoplastic Aortic Arch Repair	1435	98.19%	94.82%	1773	98.38%	96.06%			4091	98.48%	95.37%

Surgical outcomes of 10 different chosen Congenital Heart Disease (CHD) from 2000-2015. Each CHD was separately analysed to show the change of its 30-day and 1-year surgical outcome between 2000-2005 and 2010-2015.

O-040

Prenatal detection rate of aortic coarctation in a well-organized screenings setting: are we there yet?

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Background and Aim: Aortic coarctation (CoA) is still one of the most frequently missed diagnosis in prenatal screening programs. Severe CoA requires urgent postnatal care with prostaglandins and therefore, a missed diagnosis can lead to severe neonatal morbidity or death. This study explores current prenatal detection rates (DR) in a well-organized screening setting.
Methods: All prenatally and postnatally diagnosed CoA cases, born between 2012-2021, were extracted from our regional PRECOR registry. Cases with additional cardiac malformations were excluded, with the exception of persistent left superior vena cava, bicuspid aortic valve, restrictive foramen ovale and ventricular septal defect. Data on cardiac diagnosis, extra-cardiac malformations (ECM) and neonatal outcome were retrieved from medical files. Annual DR was calculated with a focus on the trend over time, as we expected that the quality of the prenatal screening should have improved through continued training of the ultrasonographers and expansion of mandatory scanning planes.
Results: In total, 116 CoA cases were eligible for inclusion. In 42.2%, the diagnosis was made before birth, of which 75.5% before 24wks GA (median GA 20.8wks). The presence of an ECM significantly increased the chance of detection, as DR for non-isolated cases was 66.7% while DR for isolated cases was 25.0% ($p<0.001$). Time trend analysis showed no improvement of the DR over time ($p = 0.33$). Of the undetected cases, 20.9% presented with an acute cardiovascular event. Although no neonatal death occurred in these cases, a significant higher proportion showed severe lactate acidosis (32.1% vs 13.6%, $p = 0.03$). The detected cases demonstrated longer duration of NICU stay and postoperative mechanical ventilation, presumably attributable to a higher proportion of more severely affected cases in this group.
Conclusion: Even in a well-organized screening program with permanent quality control, the DR of CoA (42.2%) remains disappointingly low compared to other severe congenital heart defects. A major improvement is needed, especially for isolated cases. In the 20-weeks standard anomaly scan of the Netherlands, only the three-vessel view is mandatory as scanning plane for the great vessels in the upper mediastinum. Implementation of the three-vessel-trachea view and semilunar valve measurement in our national guidelines is essential to achieve a higher DR for CoA.

Keywords: Aortic coarctation, prenatal screening, detection rate, congenital heart disease.

O-041

Fetal aortic coarctation: a combination of echocardiographic parameters that improves the prediction of postnatal outcome. a single center experience

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Background and Aim: Prenatal detection of CoA is still challenging and affected by high rate of false-positive diagnoses especially during the third trimester.
To determine a combination of fetal echocardiographic parameters for improving the prediction of coarctation of the aorta (CoA) after birth.
Methods: Cardiology database was searched for fetuses undergoing echocardiography because of ventricular and/or great vessels disproportion between January 2010 and December 2020. All fetuses diagnosed with CoA suspicion at second or third trimester of pregnancy

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were included in the study population. Last prenatal echocardiography was reviewed. The following retrospective measurements were carried out: mitral valve (MV) z-score; mitral/tricuspid valve (MV/TV) ratio; left/right ventricle (LV/RV) ratio; aortic valve/ pulmonary valve (AV/PV) ratio; AV z-score; main pulmonary artery/ ascending aorta ratio (MPA/AA); aortic isthmus (AI) and transverse aortic arch (TAA) z-score on three vessel view; aortic isthmus/arterial duct (AI/AD) ratio. LV appearance was also considered ('borderline' or adequate size LV). Gestational age (GA) was used to calculate the z score. Color Doppler assessment of flow respectively at the aortic arch and at the foramen ovale (FO) was recorded. Associated minor cardiac anomalies as ventricular septal defect (VSD), persistent left superior vena cava (PLSCV) and redundant FO membrane were reported as well as the neonatal outcome. Left common carotid-to-left subclavian artery distance (LCSA) was measured only at neonatal echocardiography. Neonates were divided into two groups depending on the absence (group 1) or presence (group 2) of CoA at postnatal ultrasound examination.
Results: Among the 91 fetuses with CoA suspicion, 27 (30%) were postnatally confirmed with CoAo and underwent surgical repair. All cardiac parameters except redundant FO membrane and PLSCV showed significant correlation with CoA (Table 1). Lower LV/RV ratio, lower aortic valve annulus z score and diffusely hypoplastic aortic arch were more common in fetuses with CoA (Table 2).
Conclusion: Prenatal detection rate of CoA may improve when a multiparametric diagnostic model is adopted. In our experience, the presence of redundant FO membrane and of PLSCV may lead to a false positive CoA diagnosis during the third trimester of pregnancy. LCSA distance should be included among the cardiac parameters assessed prenatally.

Keywords: Coarctation, Aortic Arch, Fetal, Echocardiogram

Table 1 Univariate analysis

Variable N=91	All patients	No CoA N=64	CoA N=27	P value
GA at last fetal ECHO		35.29 (2.18)	35.48 (2.24)	0.72
Early CoA Diagnosis (≤ 28 weeks)		16 (25%)	23 (85.2%)	**0.0001**
Borderline LV		0	10 (37%)	**0.0001**
Inflow tracts				
z score MV		-2.67(1,02)	-3.93 (1,30)	**0.0001**
MV/TV ratio		0.65 (0.09)	0.51 (0.08)	**0.0001**
LV/RV ratio		0.64 (0.08)	0.53 (0.11)	**0.0001**
Outflow tract				
z score AV		-0,74(0.94)	-2.22 (1.27)	**0.0001**
AV/PV ratio		0,75 (0.30)	0,56 (0.09)	**0.0001**
MPA/AA ratio		1,52 (0.33)	1,88 (0.26)	**0.0001**
Aortic Arch (on three vessel view)				
AI z score		-2.26 (1.14)	-3.59 (1.12)	**0.0001**
TAA z score		0.71 (1.35)	-1.76 (0.94)	**0.0001**
AI/AD ratio		0.51 (0.13)	0.43 (0.1)	**0.009**
Functional Features				
Reversed or mixed flow at the Aortic Arch		17 (26.6%)	13 (48.1%)	**0.05**
Bidirectional flow at the foramen ovale		8 (12.5%)	17 (63%)	**0.0001**
Associated Cardiac Anomalies				
VSD		9 (14.1%)	9 (33.3%)	**0.05**
Redundant FO Membrane		26 (40.6%)	5 (18.8%)	**0.05**
PLSCV		7 (17.9%)	3 (15%)	1
Bicuspid Aortic Valve Suspicion		5 (8%)	10 (37%)	**0.0001**
LCSA on postnatal echo		4.4 (1.16)	6.4 (1.76)	**0.0001**

Quantitative results are expressed as mean (standard deviation)

Table 2 Multivariate logistic regression

	OR (95%CI)	*p-value*
AoVZScore	0.27 (0.09-0.80)	**0.02**
LVRV diameter	0 (0-0.06)	**0.01**
Trasverse ZScore	0.22 (0.10-0.48)	**0.0001**

OR=odds ratio; CI= confidence interval;

Statistical analysis: variables compared between the CoA and non CoA groups. Univariate and Multivariate analysis

O-042

Comparison of the arterial valve in prenatally diagnosed common arterial trunk and congenital heart defects with pulmonary atresia

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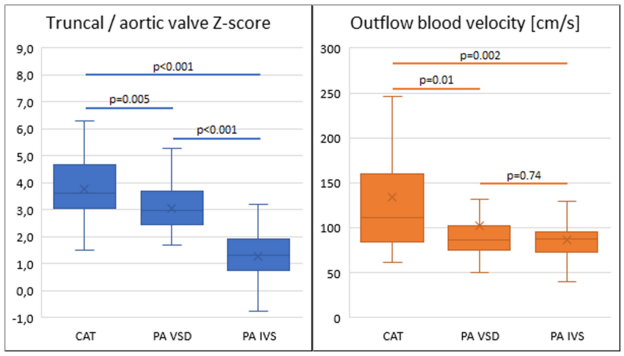
Background and Aim: Arterial valve in common arterial trunk (CAT) is usually dilated and dysplastic, due to both developmental and hemodynamic processes. To assess the relative contribution of both factors, we studied the arterial valve characteristics in fetuses with CAT and other congenital heart defects with pulmonary atresia.
Methods: We retrospectively reviewed echocardiographic examinations of fetuses diagnosed with common arterial trunk (CAT), pulmonary atresia with ventricular septal defect (PAVSD), and pulmonary atresia with intact ventricular septum (PAIVS) between 2011 and 2021 at our institution. The baseline echocardiographic examination performed between 20-25th week of pregnancy and the last prenatal examination were analyzed. Diameter of the aortic (or truncal) valve was measured together with the blood velocity across the valve. Z-score of the valve was calculated using the normal values for the aortic valve. Growth of the arterial valve was expressed as a change in its diameter over time [mm/week]. Groups were compared using T-Student test or UMW test, when appropriate.
Results: 34 fetuses with CAT, 40 with PAVSD and 34 with PAIVS were included in the study. Mean age at diagnosis was 22 weeks in all groups. Mean truncal valve Z-score was significantly bigger than the aortic valve Z-score in both PAIVS (3.76 vs. 1.26, $p<0.001$) and PAVSD (3.76 vs. 3.05, $p = 0.005$). Growth of the truncal valve was also faster than the aortic valve in PAIVS (0.39 vs. 0.24 mm/week, $p<0.001$), and, not significantly, in PAVSD (0.39 vs. 0.32 mm/week, $p = 0.07$). Velocity across the arterial valve was the highest in CAT patients (mean 134 cm/s vs. 86 cm/s in PAIVS and 102 cm/s in PAVSD, $p = 0.002$ and 0.01, respectively), reflecting frequently met truncal valve dysplasia; the aortic valve was morphologically abnormal only in single cases of PAIVS or PAVSD.
Conclusion: Disturbed outflow tract septation seems to be the main reason for increased arterial valve size in both CAT and PAVSD. Larger valve diameter and faster valve dilation in CAT may be due to its abnormal morphology and higher flow velocity, which are also important clues for making the prenatal diagnosis. Aortic valve size in PAIVS is usually within normal values despite increased output throughout a single valve.

Keywords: Fetal Cardiology, prenatal diagnosis, common arterial trunk, truncal valve, aortic valve, pulmonary atresia



Comparison of the arterial valve Z-score and blood velocity across the valve in the baseline echocardiographic examination

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Interventional Cardiology

O-043

Transcatheter creation of a restrictive atrial communication for treatment of patients with heart failure and hypertensive left atrial pressures

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Background and Aim: Left atrial decompression is considered in patients with symptomatic heart failure with hypertensive left atrium. We retrospectively evaluated our patients who underwent interventional creation of a restrictive atrial septum communication in terms of clinical improvement and as palliative therapy for future treatment as well as heart transplantation.

Methods: From February 2018 to December 2021, all patients with high left atrial hypertension, who underwent transcatheter creation of restrictive atrial septum communication, were included in the study.

Results: A total of 12 patients (female, n = 7;10/12 (83.3%) with an age ≤ 13 years; median weight 25.5 kg (range 2.8-80)) with heart failure and high left atrial pressure were intervened. The median systemic ventricular ejection fraction was 53.5% (range 25–76). The diagnoses of patients were as following: restrictive cardiomyopathy (n = 2), dilated cardiomyopathy (n = 2), hypertrophic cardiomyopathy (n = 1), non-compaction cardiomyopathy (n = 1), mitral valve stenosis (n = 2), aortic stenosis (n = 2), shone complex (n = 1) and left ventricular failure after myocardial infarction due to Kawasaki disease (n = 1). The median pulmonary arterial pressure (mean) was 33 mmHg (range 14-40), the median left and right atrial pressure (mean) were 18.5 mmHg (range 9-26) and 7 mmHg (range 4-22), respectively. Transseptal puncture was performed in 11/12 patients. The median balloon size, procedure time and fluoroscopy time were 12 mm (range 8–18), 110 min (range 82-189) and 15.9 min (range 8.6-30.1), respectively. After balloon dilatation, the median left atrial pressure (mean) decreased significantly to 13.5 mmHg (range 9-24) ($p<0.05$) and the mean right atrial pressure increased to 9.1 mmHg (range 3-17) ($p>0.05$), probably also caused by additional volume load due to contrast medium. All patients have improved clinically. The median follow-up was 31 months (range 4-45). One patient diagnosed with myocardial infarction after Kawasaki disease received orthotopic heart transplantation. No cardiac assist device was necessary in the other pateints. No procedural death or complications occurred.

Conclusion: Transcatheter left atrial decompression appears as a safe and efficient approach in children with symptomatic heart failure due to left atrial hypertension.

Keywords: interventional, creation, restrictive atrial septum communication

Other

O-044

The preponderance of genetic involvement in paediatric pulmonary hypertension

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Background and Aim: Heritable forms of paediatric pulmonary arterial hypertension (hPAH) include familial forms with or without identified mutations, and sporadic forms carrying a mutation. So far, 17 genes have been identified as playing a causal role in PAH with variable level of evidence. Children with PAH or pulmonary hypertension (PH) may also present other genetic diseases or chromosomal anomalies.

Here, we describe the prevalence and phenotypes of children with hPAH and PH with associated genetic disorder.

Methods: Children with idiopathic PAH, hPAH, PAH associated with congenital heart disease (closed shunts, coincidental shunts (APAH-CHD)), PH/PAH associated with genetic disorders diagnosed at our institution between January 2000 and October 2021.

Results: A hundred and nineteen patients had confirmed PH at right heart catheterization (mean pulmonary artery pressure > 25 mmHg, PVRi > 3 WU.m2).

Forty-one patients (34.4%) had hPAH, involving various genes mutations: 11 BMPR2, 7 TBX4, 6 ACVRL1, 3 Sox 17, 3 EIF2AK4, 2 BMP9, 1 SMAD 4, 1 variant of unknown significance (VUS) in the CAV1 gene, 1 KCNK3, 6 patients with familial PAH had no identified mutations.

Seventeen patients (14.3%) had a genetic disorder associated with PH: 2 VHL mutations, 1 incontinentia pigmenti, 1 neurofibromatosis type 1, 3 Down syndromes, 1 PTPN11 mutation, 1 ABCC9 gain of function mutation, 1 JAG1 mutation, 1 TSC1 mutation, 1 SARS II mutation, 1 cyanocobalamin deficiency, 1 NOD2 mutation, and 3 chromosomal rearrangement (table 1). Twenty-four had sporadic idiopathic PAH (20%) without mutation in the known PAH genes, and 20 had APAH-CHD (16%).

Conclusion: Prevalence of genetic disorders and hPAH in children with PH/PAH appears higher than previously described, being almost observed in half of the children. Systematic genetic testing in apparently idiopathic PAH as well as in newly diagnosed patients with PH should not be solely limited to the current panel

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of known genes but may include larger sets of genes as well as exome/genome sequencing to increase detection of new genes in this condition.

Keywords: Pulmonary hypertension, genetics, children.

Genes involved and genetic disorders associated PH

Genes involved and genetic disorders associated PH

Heritable PAH	N (%)	N Associated CHD	Type of CHD
BMPR2	11	0	
TBX 4	7	2	1 ASD 1 PDA
ACVLR1	6	1	1 ASD
Sox 17	3	1	1 ASD
EIF2AK4	3	0	
BMP9	2	0	
SMAD4	1	0	
KCNK3	1	1	1 ASD
CAV1 (VUS)	1	0	
No mutation	6	2	2 ASD
Total (%)	41 (100)	7 (17)	
PH associated genetic disorder	**N (%)**	**N associated CHD**	**Type of CHD**
Down Syndrome	3	2	2 PDA
PTPN11 Mutation	1	1	1 ASD
VHL mutation	2	0	
NF1 mutation	1	0	
ABCC9 gain of function	1	0	
STB1	1	0	
7q11.22 deletion	1	1	1 ASD and 1 VSD
JAG1 mutation	1	1	TOF
CHRNA7 mutation	1	0	
Incontinentia pigmenti	1	0	
Chromosome 2 microdeletion	1	0	
1p36 deletion	1	0	
TEK gene mutation	1	0	
NOD 2 mutation	1	0	
Total (%)	17 (100)	5 (29)	

CHD: Congenital heart disease, ASD: Atrial septum defect, VSD: Ventricular septal defect, TOF: Tetralogy of Fallot, PDA: Patent Ductus arteriosus, VUS: Variant of unknown significance.

Heart failure

O-045

Rimeporide, a first in class na/h exchanger-1 inhibitor, ameliorates right ventricular dysfunction and pulmonary arterial hypertension in a suhx model

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Background and Aim: Pulmonary arterial hypertension (PAH) is characterized by pulmonary vasoconstriction and vascular remodeling leading to right ventricular (RV) failure. Although RV function is the major prognostic factor in PAH, no RV-specific therapies exist. The Na+/H+ exchanger type 1 (NHE1) regulates the intra- and extracellular pH balance mainly through the 1:1 exchange of intracellular H+ with extracellular Na+. Increased NHE-1 activity is involved in several diseases, including hypoxic PAH. Inhibition of NHE-1 activity attenuates pulmonary vascular remodeling in rodents exposed to chronic hypoxia but its potential direct contribution in RV remodeling remains unclear.

AIM: To investigate the cardio-pulmonary protective effects of Rimeporide, a first in class NHE-1 inhibitor, in rats with PAH and RV dysfunction.

Methods: PAH was induced in adult Sprague-Dawley rats with a single injection of SU5416 (20 mg/kg) followed by 3 weeks of chronic hypoxia (10% O2; Sugen/Hypoxia model: SuHx) and then return to normoxia. Control rats were injected with equal volume of vehicle. At week 5 post-injection, control and SuHx rats were treated with rimeporide (100 mg/Kg daily in drinking water, n = 10/group) or placebo (n = 11/group) for 3 weeks. RV phenotype was monitored by echocardiography and hemodynamic analysis. Lung and heart were collected for histological and protein expression analysis, such as NHE-1 and hypoxia-inducible factor-1 (HIF-1 α).

Results: As reported in Table I, SuHx caused severe PAH and RV dysfunction. RV dysfunction in SuHx rats was accompanied by an increase in RV hypertrophy and RV fibrosis and PA remodeling (increase in the arteriolar wall thickness and fibrosis). A marked increase in macrophages was found in lung and RV tissues but not in LV. Moreover, Western blot analysis showed that RV protein levels of NHE-1 and HIF-1α were higher in SuHx rats than Normoxia rats (2-fold and 4-fold increase, respectively). Treatment with Rimeporide reversed SuHx-induced PAH as well as RV dysfunction and blunted pulmonary and RV inflammation, RV NHE-1 and HIF-1α expression.

Conclusion: Our results indicate that NHE-1 and HIF-1α levels are increased in RV tissues from SuHx-induced PAH rats. Rimeporide, through NHE-1 inhibition, might be a potential new target for treatment of RV dysfunction and PAH.

Keywords: Pulmonary arterial hypertension; right ventricular dysfunction; chronic hypoxic

Table: Effects of Rimeporide on cardio-pulmonary function.

Definition of abbreviations: PAT/ET= pulmonary acceleration time/ejection time; VTI= velocity time integral; mPAP= mean pulmonary arterial pressure; RVFWTd= RV free wall thickness at diastole; RVIDd= RV internal diameter at diastole; RVEF= RV ejection fraction; RV+dp/dt= RV dp/dt maximum; RV-dp/dt= RV dp/dt minimum; RV and LV CSA= RV and LV cross-sectional area.

* vs Normoxia and Normoxia + Rimeporide rats
# vs SuHx rats.

Parameters	Normoxia		Normoxia + Rimeporide		SuHx		SuHx + Rimeporide		P ANOVA One-way
Echocardiography									
PAT/ET, ratio	0.36 ±0.02	11	0.35±0.02	10	0.18±0.01*	11	0.24±0.01*#	10	<0.0001
VTI, mm	51.2±1.66	11	52.7±1.21	10	35.0±1.75*	11	42.2±2.25*#	10	<0.0001
mPAP, mmHg	19.9±1.85	11	22.2±2.11	10	39.4±2.23*	11	32.4±1.45*#	10	<0.0001
RV FWT d, mm	1.05±0.05	11	1.10±0.03	10	2.17±0.05*	11	1.77±0.12*#	10	<0.0001
RV ID d, mm	2.41±0.10	11	2.24±0.07	10	3.54±0.02*	11	3.01±0.12*#	10	<0.0001
RV EF, %	93.1±1.33	11	88.2±2.63	10	54.2±3.45*	11	77.9±1.30*#	10	<0.0001
Hemodynamics									
RV end-systolic pressure, mmHg	21.9±0.10	9	20.9±1.01	9	56.5±3.61*	7	43.3±3.51*#	9	<0.0001
LV end-diastolic pressure, mmHg	98.7±3.39	11	104.3±5.19	10	94.0±4.90	9	103.0±3.30	10	NS
RV end-diastolic pressure, mmHg	1.40±0.22	9	1.09±0.20	9	2.82±0.16*	7	1.84±0.31#	9	<0.0001
RV +dp/dt, mmHg/s	1039±110	9	956±53	9	1838±79*	7	1864±141*	9	<0.0001
RV -dp/dt, mmHg/s	-607±52	9	-681±48	9	-1319±79*	7	-1353±97*	9	<0.0001
RV Tau, ms	20.3±0.99	9	19.6±0.97	9	24.5±1.42*	7	19.0±0.93*#	9	<0.001
RV remodeling									
Body weight, g	487.1±14.7	11	481.1±9.1	10	444.1±12.6	11	455.0±13.7	10	NS
RV/LV+Septum, ratio	0.25±0.02	5	0.26±0.01	4	0.67±0.09*	4	0.53±0.04*#	4	<0.0001
RV CSA, mm2	335.8±16.9	6	340.4±22.2	6	1062.3±54.0*	7	827.2±79.30*#	6	<0.0001
LV CSA, mm2	385.2±33.1	6	420.2±24.7	6	421.8±29.3	7	419.3±32.4	6	NS
RV collagen, %	5.46±0.68	6	5.52±0.59	6	22.25±1.88*	7	14.88±1.59*#	6	<0.0001
LV collagen, %	4.56±0.46	6	5.15±0.76	6	7.81±0.53*	7	4.77±0.45*#	6	<0.001

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Imaging/Functional assessment

O-046
Validation of the new eppvdn pediatric pulmonary hypertension risk score by cardiac magnetic resonance imaging and speckle tracking echocardiography

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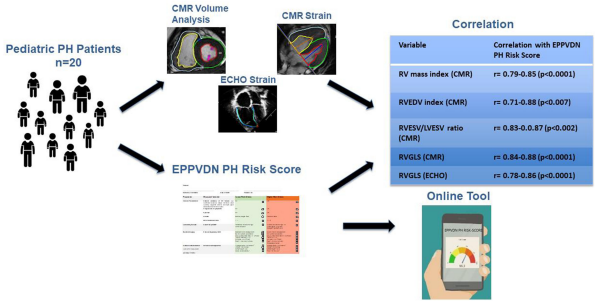
Background and Aim: The European Pediatric Pulmonary Vascular Disease Network (EPPVDN) developed in 2019 a pediatric pulmonary hypertension (PH) risk score to assess the risk and severity of PH in children and young adults. This prospective observational study seeks to validate the EPPVDN pediatric PH risk score by correlating suitable variables from cardiac magnetic resonance imaging (CMR) and speckle tracking echocardiography.
Methods: We enrolled 20 children with PAH (age: 10.8 ± 1.0, range 4.0-17.6 years). During the same inpatient stay, invasive and non-invasive EPPVDN PH risk scores were determined and a protocol-driven CMR study was performed. Subsequently, we correlated the risk scores with conventional and strain CMR- and advanced echocardiographic variables, including strain (speckle-tracking). In addition, we applied the risk score to 9 children with PAH who received add-on selexipag therapy. Before and approximately 6 months after selexipag start, the risk score and echocardiographic 2D RV free wall longitudinal strain (RVFWLS) were determined and delta-changes of both were correlated. For a broad application, we digitized the risk score as a web-based online tool using the Flutter-Programming-Kit version 2.5.3 developed by Google.
Results: We observed significant ($p<0.006$) and strong correlations of conventional CMR ($r = 0.69$-0.88), CMR strain ($r = 0.71$-0.88), advanced echocardiographic ($r = 0.65$-0.88) and echocardiographic strain variables ($r = 0.67$-0.86), with the EPPVDN PH risk scores. The EPPVDN higher risk scores correlated stronger with the CMR-, ECHO-, and CMR-strain variables, when compared to the lower risk scores. In the add-on selexipag cohort, the change in ECHO-derived RV free wall strain correlated well with the change in the invasive higher risk score ($r = 0.72$, $p<0.03$).
Conclusion: We found strong correlations of outcome-relevant CMR and ECHO variables, including strain, with the EPPVDN PH risk scores, and thus validated the score via independent methods. The novel web-based tool enables a quick and easy usage of the digitized PH risk score for clinicians and researchers. The novel EPPVDN PH risk score will be useful in routine clinical care, and can now be applied in larger prospective, pharmacological-interventional PH studies in children and young adults.

Keywords: pulmonary hypertension – risk score – right ventricle – children –cardiac magnetic resonance – strain analysis

Central Illustration Validation of the New EPPVDN Pediatric Pulmonary Hypertension Risk Score



Central Illustration

Interventional Cardiology

O-047
Long-term outcomes of transcatheter potts shunt in children with suprasystemic pulmonary arterial hypertension

Raymond Nagi Haddad, Maryline Levy, Isabelle Szezepanski, Sophie Malekzadeh Milani, Damien Bonnet; Centre de Référence Malformations Cardiaques Congénitales Complexes - M3C, Hôpital Universitaire Necker-Enfants malades, AP-HP, Paris, France.

Background and Aim: Transcatheter Potts shunt (TPS) is identified as effective palliation for children with severe pulmonary arterial hypertension (PAH). Debates on the long-term outcomes remain unsolved. We comprehensively evaluate the long-term outcomes of the largest single-center experience with TPS with a particular focus on the durability of the implanted material.
Methods: Transcatheter Potts shunt (TPS) is identified as effective palliation for children with severe pulmonary arterial hypertension (PAH). Debates on the long-term outcomes remain unsolved. We comprehensively evaluate the long-term outcomes of the largest single-center experience with TPS with a particular focus on the durability of the implanted material.
Results: Thirteen patients (53.8% males) were identified. TPS was performed with bare-metal stenting of restrictive or probe arterial duct ($n = 7$) or created by radiofrequency vessel perforation and covered stenting ($n = 6$). Improvement in overall clinical condition was significant at discharge ($p<0.001$), inconsistent across follow-up, but remained significantly improved at the last visit ($p<0.05$). Improvement in functional status was significant ($p<0.001$). There was no significant gradual improvement in other disease markers (TPASE, 6MWD z-scores, and NT-proBNP levels). 57.1% of patients initially under prostanoid therapy were weaned but reduction in the overall need for

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PAH medications was not significant. Median follow-up was 62.7 months (IQR, 32.2-96.5 months). One patient died 28.5 months postoperative after a severe viral infection. Survival was 100% at 1 year and 92.3% at 5 years. Freedom from reinterventions was 77% and 17% at 1 year and 5 years, respectively. Stent malfunctioning indicated balloon dilatation (46.1%) and re-stenting (38.5%). Stent malfunctioning was present in 46.2% of patients at the latest follow-up. Independently of the anatomical indication, the need for reinterventions was quite similar in both techniques applied for TPS. One patient is listed for heart-lung transplantation.

Conclusion: Survivors of TPS procedures experience significant improvement in functional class that can be durable and potentially free from prostanoid. Clinical worsening and stent malfunctioning are frequent morbid events indicating recurrent transcatheter reinterventions throughout follow-up. Five-year survival is however satisfactory. Room for innovation is existent and the challenges can be addressed.

Keywords: Children; outcomes; pulmonary arterial hypertension; Potts shunt.

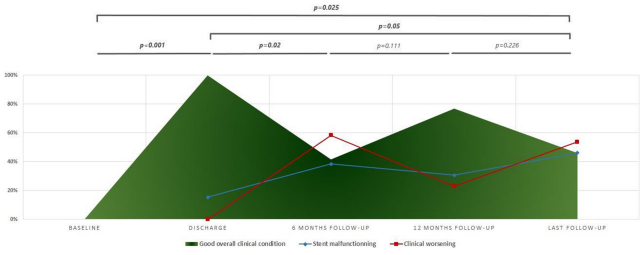

Figure 1. Progression of overall clinical status across follow-up *Clinical worsening is defined by the presence of at least one of the following: 1) worsening in world health organization- Functional class (WHO-FC); 2) NT-pro BNP ≥1400 pg/ml; 3) tricuspid annular plane systolic excursion (TAPSE) ≤15mm; 4) increase or adjunction of diuretics; and 5) adjunction of new pulmonary arterial hypertension specific medical therapy. Stent malfunctioning is defined as maximum velocity on stent >2 m/s*

Imaging/Functional assessment

O-048

Full recovery of right ventricular systolic function in children undergoing bilateral lung ttansplantation for severe pulmonary arterial hypertension

Georg Hansmann[1], Franziska Diekmann[1], Philippe Chouvarine[1], Fabio Ius[2], Julia Carlens[3], Nicolaus Schwerk[3], Gregor Warnecke[4], Jens Vogel Claussen[5], Dagmar Hohmann[1], Tim Alten[5], Thomas Jack[1]
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Background and Aim: We investigated whether RV function recovers in children with pulmonary arterial hypertension (PAH) and RV failure undergoing bilateral lung transplantation (LuTx).

Methods: Prospective, observational study of 15 consecutive children, 1.9-17.6 years old, with PAH receiving bilateral LuTx (December 2013 - January 2021). We performed advanced echocardiography (Echo) and cardiac magnetic resonance imaging (MRI), followed by conventional and strain analysis, pre and ~6 weeks post LuTx (Table 1).

Results: Nine patients were under 12 years old (LAS exemption), two of which had body surface area and weight below 0.5 m2 and 9 kg, respectively. About half of the patients had a body weight below the 10th (n = 8) or even below the 1st (n = 4) percentile (cachexia). After LuTx, RV/LV endsystolic diameter ratio (Echo), RV volumes and systolic RV function (RVEF 63 vs. 30 %; $p<0.05$) by MRI completely normalized, even in children with severe RV failure (RVEF < 40%). The echocardiographic endsystolic LV eccentricity index nearly normalized post LuTx (1.0 vs. 2.0, $p<0.0001$) while RV hypertrophy regressed more slowly and was still evident. We found especially the end-systolic RV/LV ratios by Echo (diameter; 0.6 vs. 2.6) or MRI (volumes: 0.8 vs. 4.2) to be excellent diagnostic tools ($p<0.05$): Together with RVEF by MRI, these ratios were superior to tricuspid annular plane systolic excursion (TAPSE; $p>0.05$) in assessing global systolic RV dysfunction. Moreover, children with severe PAH had reduced RV 2D longitudinal strain (Echo, MRI; $p<0.05$) and decreased RV 2D radial and circumferential strain (MRI; $p<0.01$), all of which greatly improved following LuTx. One of the 15 PH-LuTx patients had to be re-transplanted 9 months after the initial LuTx because of cellular rejection. As of December 1, 2021, all 15 pediatric PAH-LuTx patients are alive and in clinical follow up (mean 39 months; range 11 months – 8 years).

Conclusion: We demonstrate full recovery of RV systolic function in children within two months after LuTx for severe PAH, independently of the patients' age, weight, and hemodynamic

Table 1. Characteristics of all 15 PH Patients Undergoing Bilateral Lung Transplantation

Patients #1-15	Pre LuTx n = 15	Post LuTx n = 15
Demographics		
Age – years	10.7±1.3 (1.9 – 17.6)	10.9±1.3 (2.1 – 17.8)
Sex, Female – n (%)	12 (80%)	12 (80%)
Height – m	1.4±0.1	1.4±0.1
Weight – kg	32.3±4.0	31.5±3.9
BSA – m^2	1.1±0.1	1.1±0.1
Clinical Diagnosis		
PH Group – n		
PH Group 1		
1.1 IPAH	6	
1.2 HPAH	4	
1.4.4 PAH-CHD	2	
1.6 PVOD/PCH	3	
Co-morbidities – n		
Hereditary thrombophilia	1	
HHT (Osler's disease)	1	
Type 1 diabetes	1	
von Willebrand disease	6 confirmed	
Functional status		
WHO Functional Class	3.7±0.1	
6-min. walk distance – m, n = 13	213±46	
NTproBNP – ng/l, n = 7	3093.7±1647.3	945.6±262.2
Key Hemodynamics		
mRAP – mm Hg, n = 12	9.3±1.1	
RVEDP – mm Hg, n = 11	12.7±0.9	
mPAP/mSAP, n = 12	1.2±0.04	
PVRi – WU·m^2, n = 12	26.2±2.5	
PVR/SVR, n = 12	1.4±0.1	
Qsi – L/min/m^2, n = 12	2.9±0.3	
Risk stratification (EPPVDN)		
Patients total – n	15	
Noninvasive Risk – n	Higher Risk – 9 Intermediate Risk – 6	
Higher Risk Score, max. 15 (decimal)	10.2/15 (0.68±0.04)	
Lower Risk Score, max. 14 (decimal)	1.9/14 (0.14±0.02)	
Patients with cath 0-12 months pre LuTx – n	12	
Invasive Risk – n	Higher Risk – 8 Intermediate Risk – 4	
Higher Risk Score, max. 21 (decimal)	13.3/21 (0.63±0.05)	
Lower Risk Score, max. 20 (decimal)	3.3/20 (0.16±0.03)	
Pre/Post LuTx Imaging Intervals		
Interval Echo to Tx / Tx to Echo – days (range), n = 15	28±6 (0 – 75)	41±5 (9 – 76)
Interval MRI to Tx / Tx to MRI – days (range), n = 6	61±31 (15 – 203)	43±4 (31 – 56)

Table 1. *Characteristics of all 15 PH Patients Undergoing Bilateral Lung Transplantation.* Values are presented as mean ± SEM. Last NTproBNP levels pre LuTx and NTproBNP levels ± 14 days around the post LuTx echo are shown. For risk stratification, see the new 2019 EPPVDN risk score.

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compromise preceding the LuTx. Even in end-stage pediatric PAH with poor RV function and low cardiac output, LuTx should be preferred over heart-lung transplantation.

Keywords: right heart failure, pulmonary arterial hypertension, bilateral lung transplantation, echocardiographic 2D speckle tracking, cardiac magnetic resonance, CMR tissue tracking

O-049

MRI lymphangiography in children: a single centre experience

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Background and Aim: Refractory chylothorax/chyloperitoneum, plastic bronchitis and protein losing enteropathy can be a consequence of functional (high right atrial pressure, anatomical obstruction to the systemic venous drainage) or anatomical disorder/disruption of the lymphatic system drainage.
Our aim was to demonstrate the role of dynamic contrast-enhanced MRI-lymphangiograms (MRL) in identifying problems with the lymphatic drainage system and guide appropriate further management (medical, interventional or surgical).
Methods: Sixteen MRL studies were performed at the Evelina London Children's Hospital between April 2017 and August 2021 in patients referred for pleural effusion (6), plastic bronchitis (5), protein losing enteropathy (4) or pulmonary lymphangiectasia (1).
Mean age was 9 years and 4 months (range 6 month – 22 year), 11 were male and 5 were females. Fourteen patients had operated congenital heart disease, including 8 with a Fontan circulation. 15 studies were performed under general anaesthesia and one under local anaesthesia. Bilateral inguinal lymph node cannulation was performed under ultrasound guidance, before intranodal injection of 0.1ml/kg Gadovist per site. Serial 3D isotropic T1 weighted images were acquired with coverage of the groin, abdomen and thorax to fully visualize the lymphatic drainage to the systemic venous system over 30-60mins.
Results: All cases were diagnostic with no procedural complications. Abnormalities of the lymphatic system were identified in 9 out of 16 cases with associated lymphatic leaks in 2 cases. Four patients underwent a targeted intervention which were all successful in resolving the presenting lymphatic issue. One patient received selective embolization of an abnormal leaking lymphatic vessel. Three patients had interventions to treat functional lymphatic obstruction: 2 underwent successful systemic venous recanalization, and 1 had cardiac surgery with pulmonary valve replacement and TV repair to improve central venous pressures. One patient is currently awaiting selective embolization of an abnormal leaking vessel. All remaining patients had medical or conservative treatment and remained stable at last follow up.
Conclusion: MRL is a safe diagnostic tool in children and young adults which helps aid clinical decision making and target treatment of refractory chylothorax/chyloperitoneum, plastic bronchitis and protein losing enteropathy.

Keywords: lymphangiography, lymphangiogram, chylothorax, plastic bronchitis, protein-losing-enteropathy

O-050

Non-invasive assessment of myocardial work in adolescents and young adults with effectively corrected versus residual aortic coarctation

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Background and Aim: Echocardiographic evaluation of left ventricular (LV) systolic function using ejection fraction (EF) and global longitudinal strain (GLS) has low sensitivity to evaluate results and stratify risks in corrected aortic coarctation (AoCo) patients. Non-invasive myocardial work (MW) is a novel parameter that is obtained by dividing LV pressure by GLS (units: mmHg%). This study aims to explore the use of this parameter in aortic coarctation.
Methods: 53 AoCo patients and 31 healthy controls, aged 12-40 years, were evaluated. Effectively corrected AoCo (cAoCo) was defined as transisthmic Doppler gradient ≤20mmHg (Dgrad) (n = 36), and residual AoCo (rAoCo) as Dgrad > 20 mmHg (n = 17). Dependent variables were: biplane EF; GLS; global work index (GWI) – total work; global constructive work (GCW – work performed during shortening in systole); global wasted work (GWW – energy not contributing to blood ejection); global work efficiency (GWE – GCW divided by the sum of GCW and GWW). T-tests were used to compare these variables among groups. Multivariable regression was used to test their correlation with LV indexed mass (LVimass), Dgrad and the ratio between the narrowest diameter of aortic arch and aorta at diaphragm level (AoRatio).
Results: LVimass was significantly higher in patients with rAoCo when comparing with both controls ($p = 0.005$) and cAoCo ($p = 0.028$). EF and GLS were not different among groups. GWI, GCC and GWW were significantly higher in rAoCo than controls, but not in patients with cAoCo (table 1). GWE was lower in both cAoCo and rAoCo. Measures of MW were significantly determined by LVimass (GCW - $p = 0.005$; GWW- $p<0.001$; GWE – $p = 0.009$), even when controlling for Dgrad or AoRatio.
Conclusion: Evaluation of MW parameters provide a better discrimination of LV systolic function in patient with AoCo, and should be considered for routine follow up assessment. Patients with effectively corrected AoCo have mild LV systolic disfunction when assessed by these novel parameters, which is more severe in patients with residual gradient. This provides rational for a stronger control of risk factors that raise LV afterload, and, ultimately, LV mass.

Keywords: aortic coarctation, echocardiography, systolic function, myocardial work

O-051

Cardiac catheter examination in kawasaki disease: initial data from the central european registry

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Background and Aim: Patients with a history of Kawasaki disease (KD), especially those with diagnosed coronary artery involvement, require long-term cardiac care. Cardiac catherization (CC) remains the gold-standard for diagnosing coronary pathology. However, there is no evidence-based diagnostic and therapeutic strategy for this patient population. We conducted an international survey to document and evaluate the clinical impact of CC performed in KD patients. Here is our first preliminary data.
Methods: We used a standardized questionnaire to retrospectively analyze CCs performed in KD children from the year 2010 until today. Acute phase clinical data, detailed morphology, distribution and development of coronary artery pathologies and therapeutic strategies pursued in these patients are covered in this registry.
Results: A total of 159 patients (65% male) undergoing 233 CCs have been included so far. Median age during KD's acute phase was 1,7 years with a mean follow-up interval to the first CC of 27-month (range 0 month - 21 years). Almost 60% of the patients had at least one coronary artery aneurysm and up to 8 aneurysms in one patient have been detected. The segment involved most often was the right coronary artery's (RCA) proximal segment (50/159 patients) and it displayed the largest aneurysmatic diameter and length (mean/range of the CAA diameter x length): 6mm/1,9-31 mm x 10,7mm/2,8 – 62 mm). Cross-table analysis revealed that the distal RCA and LAD segments exhibit CAAs mainly if the respective proximal segments were also affected; the ratio was 2/14 for the RCA ($p<0,0001$) and 0/9 for the LAD (; $p<0,0001$). In addition there was a positive correlation between the maximum diameter (and respective Z-Score) and the total number of aneurysms per patient ($RP = 0,54$; $p < 0,0001$; see figure 1).
Conclusion: According to our first data-analysis, the proximal right coronary artery was the most affected vessel. There was a significant positive correlation between the number of CAAs and the maximum CAA diameter. Distal CAAs occurring more often when proximally-located CAAs existed, may influence the long-term cardiac follow-up in KD patients.

Keywords: Kawasaki disease, Coronary artery aneurysms, Cardiac catherization

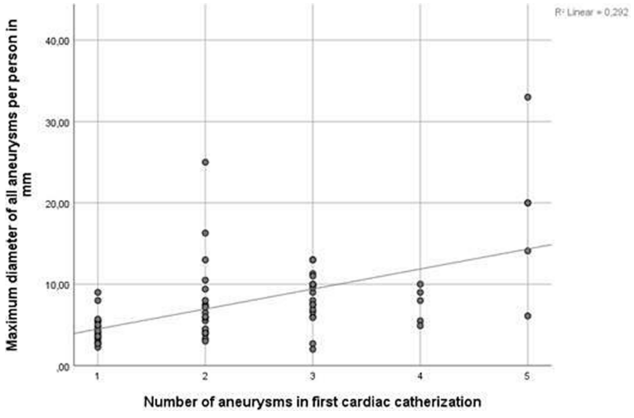

Figure 1. Significant positive correlation between maximum diameter of the largest aneurysm and the total number of aneurysms present ($RP = 0,54$; $p < 0,0001$).

O-052

Use of speckle tracking echocardiography to detect anthracycline-induced cardiotoxicity in childhood cancer: a prospective controlled study

Marie Vincenti[1], Hamouda Abassi[1], Nicolas Lanot[1], Grégoire De La Villéon[1], Sophie Guillaumont[1], Thibault Mura[6], Laurence Vergely[7], Olivier Cazorla[4], Alain Lacampagne[4], Nicolas Sirvent[3], Laure Saumet[3], Martina Avenasi[5], Pascal Pascal[2]
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Background and Aim: This study aimed to detect sub-clinical patterns of cardiac dysfunction using speckle tracking echocardiography (STE) in children with cancer remission more than 12 months after the end of anthracycline treatment.
Methods: This prospective controlled study enrolled 196 children, 98 of which had been treated with anthracyclines (mean age 10.8 ± 3.6 years; 51% female) and 98 were age- and gender-matched healthy subjects in a 1:1 case-control design. Conventional echocardiographic variables were collected for left ventricle (LV) and right ventricle (RV). STE analyses were performed in the LV longitudinal, radial, and circumferential displacements and in the RV free wall longitudinal displacement. The association between LV global longitudinal strain (GLS) and the main clinical and biological parameters was evaluated.
Results: After a mean time interval of 5.1 ± 3.2 years since the end of chemotherapy (mean cumulative anthracycline dose of 192 ± 96 mg/m2), conventional echocardiographic measures were normal. GLS was significantly decreased in the anthracycline group (-19.1% vs. -21.5%, $P<0.0001$), with a higher proportion of children with abnormal values (Z-score <-2 in 18.6% vs. 1.0%, $P<0.0001$). No association was found between GLS and clinical or biological parameters. Circumferential strain was significantly worse in the anthracycline group (-16.8% vs. -19.4%, $P<0.0001$), and radial strain significantly better (+51.4% vs. +35.9%, $P<0.0001$). RV conventional echocardiography and STE parameters were normal and not different between anthracycline and control groups.
Conclusion: The existence of a modified LV strain despite normal LV function in children treated with anthracyclines represents an important perspective for cardiomyopathy surveillance in childhood cancer survivors.

Keywords: chemotherapy, 2D strain, echocardiography, pediatrics, cardiomyopathy, heart failure

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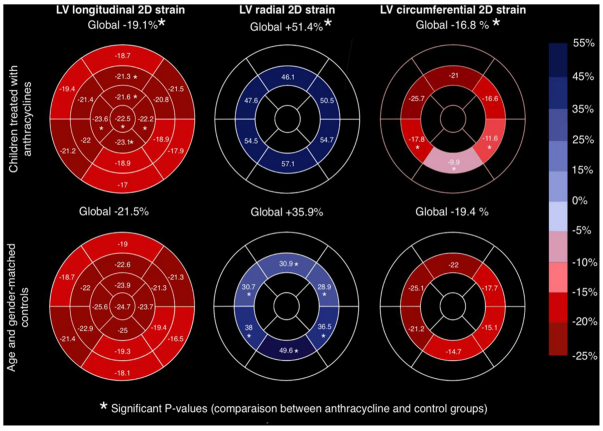
Left ventricle global and segmental 2D strain in children treated with anthracyclines and control subjects



These 6 figures represent, for each group, the longitudinal, radial and circumferential 2D strain, using the classical blue (positive displacement) and red (negative displacement) color-coded bull's eye plot. The anthracycline group is represented on the top 3 figures and the controls on the 3 bottom ones. In each strain segment of the anthracycline group, an asterisk indicates the existence of a significant difference with the corresponding segment of the control group.

O-053

Diagnostic value of 18F-fluorodeoxyglucose positron emission tomography computed tomography in prosthetic pulmonary valve infective endocarditis

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Background and Aim: Pulmonary prosthetic valve or conduit endocarditis (PPVE) is a major issue in the growing congenital heart disease (CHD) population. Diagnosis is challenging and usual imaging tools are not always efficient or validated in this specific population. Particularly, the diagnostic yield of 18F-fluorodeoxyglucose (18F-FDG) positron emission tomography (PET) / computed tomography (CT) remains poorly studied in PPVE. The aim of this study was to assess the diagnostic performances of 18F-FDG PET/CT in CHD patients with PPVE suspicion.
Methods: A retrospective multicenter study was conducted in 8 French tertiary centers. Children and adult CHD patients who underwent 18F-FDG PET/CT in the setting of PPVE suspicion between January 2010 and May 2020 were included. The cases were initially classified as definite, possible, or rejected PPVE regarding the modified Duke criteria and finally by the Endocarditis Team consensus. The result of 18F-FDG PET/CT had been compared with final diagnosis consensus used as gold-standard in our study.
Results: A total of 66 cases of PPVE suspicion involving 59 patients (median age 23 years, 73% men) were included. Sensitivity, specificity, positive predictive value, and negative predictive value of 18F-FDG PET/CT in PPVE suspicion were respectively: 79.1% (95% CI: 68.4%-91.4%), 72.7% (95% CI: 60.4%-85.0%), 91.9% (95% CI: 79.6%-100.0%), and 47.1% (95% CI: 34.8%-59.4%). 18F-FDG PET/CT findings would help to correctly reclassify 57% (4 of 7) of possible PPVE to definite PPVE.
Conclusion: Using 18F-FDG PET/CT improves the diagnostic accuracy of the Duke criteria in CHD patients with suspected PPVE. Its high positive predictive value could be helpful in routine to shorten diagnosis and treatment delays and improve clinical outcomes.

Keywords: 18F-FDG PET/CT, congenital heart disease, infective endocarditis, pulmonary valve

O-054

Magnetic resonance vessel wall imaging is a reliable technique for determining vessel wall thickening in coronary lesions of kawasaki disease

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Background and Aim: Two-dimensional cross-sectional Double-Inversion-Recovery black-blood vessel wall imaging (VWI) of the coronary arteries is a non-invasive tool for evaluating vessel wall thickening. However, there is a paucity of views on the relationship between the maximum diameter of the coronary lesion in the acute phase of Kawasaki disease (KD) and stenotic lesions in the remote phase and wall thickening evaluated by VWI.
Methods: This retrospective study included fifty-four patients with seventy-four coronary lesions (median age at KD onset: 2.1 years [IQR; 0.6-4.5], median total evaluation period from KD onset: 7.1 years [IQR; 3.5-14.0]) in whom cardiovascular magnetic resonance was examined more than 100 days after KD onset from May 2007 to December 2020. All examinations were performed on 1.5-Tesla MR clinical imagers (Intera Achieva and Ingenia, Philips Healthcare, Best, The Netherlands). VWI were obtained by a free-breathing navigator-gated and cardiac-triggered Black Blood T1-weighted Turbo Field Echo positioned using coronary MR angiography. The lesions were divided into two groups in the final evaluation, Group P (n = 40) with wall thickening and Group N (n = 34) without wall thickening. Five wall thickening lesions were also evaluated by optical coherence tomography (OCT) in Group P. The maximum diameter of the coronary lesion obtained by echocardiogram in the acute phase of KD was compared between the two groups.
Results: All five Group P lesions evaluated by OCT showed remarkable intimal thickening in the areas with thickened vessel

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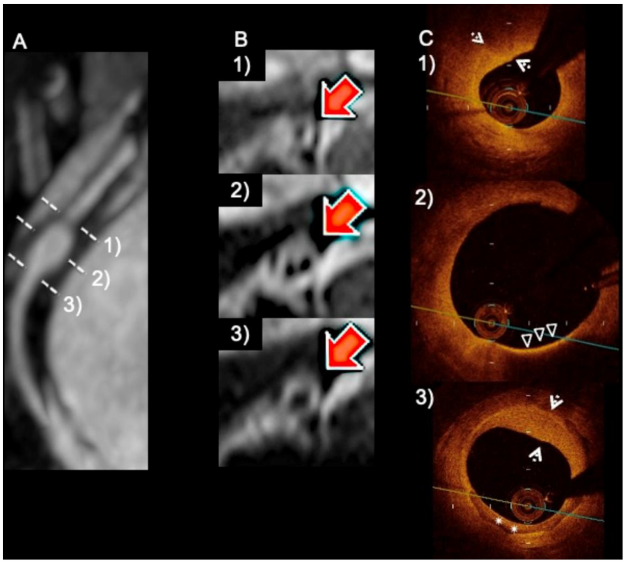
walls using VWI. The maximum diameter of the coronary lesion in the acute phase was significantly greater in Group P than Group N (median [IQR]; P: 5.9 mm [5.0-7.0] vs. N: 4.0 mm [3.5-4.6], p<.0001). Six lesions (15%) in Group P developed stenotic or occlusive lesions in the remote phase, whereas no lesions in Group N developed stenotic lesions. According to ROC curve analysis, a 5.2 mm diameter of the coronary lesion in the acute phase had maximum sensitivity and specificity for separating lesions with vessel wall thickening evaluated by VWI (AUC: 0.879, Sensitivity: 0.70, Specificity: 0.91, p<.0001).
Conclusion: VWI is a reliable technique for evaluating wall thickening in the coronary artery lesions of KD in the remote period.

Keywords: Coronary Vessel Wall Imaging, Coronary artery lesions, Kawasaki Disease, Cardiac Magnetic Resonance, Optical Coherence Tomography

Imaging findings of an 8-year-old boy with stenotic lesions and aneurysm in the RCA.



On MRCA, the proximal part of the aneurysm showed localized stenosis (A). VWI showed thickening in lesions (B, arrows). On OCT(C), intimal thickening with fibrosis (dashed arrows), a lipid-laden intima (open triangles), and calcified intima (asterisks) were evident.

Interventional Cardiology

O-055

Ductus arteriosus stenting versus blalock-taussig shunt for infants with ductal-dependent pulmonary circulation: a comparative international study

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Background and Aim: New-borns with congenital heart disease with ductal-dependent pulmonary blood flow (PBF) require an early palliative procedure, either a patent ductus arteriosus stenting (PDAs) or a modified Blalock-Taussig shunt (mBTS). Outcomes with mBTS and those with PDAs have been compared but available data are scarce. An international multicentre comparison of these 2 techniques would be of major interest.
Methods: Infants<1 month with ductal-dependent PBF palliated with either a PDAs or a mBTS between Jan 2000 and Dec 2019 were reviewed from 10 European participating centres (France n = 4, Spain n = 5, UK n = 1). Peri-procedural and mid-term outcomes were compared between the 2 groups, using a propensity score adjustment calculated with potential confounding variables.
Results: The study cohort was composed of 179 infants who underwent PDAs and 456 infants with mBTS. Expected biventricular physiology and presence of additional PBF were in greater proportion in PDAs group (66% vs 57%, p = 0.03 and 57% vs 39%, p<0.001, respectively). Median follow-up was 79 months (IQR 39.50-138.00). After propensity score adjustment, no significant differences were found in relation to the primary outcome of survival before next-stage surgery or at 18 months, (mBTS 91.7%, [95% CI 86.4-97.4] vs PDAs 88%, [95%CI 80-96.8]; p = 0.69). Patients of PDAs group had a reduced the risk of death before repair compared to mBTS group (hazard ratio, 0.76; [95% CI, 0.55-1.06]; p = 0.10). However, the incidence of reintervention (either planned or unplanned) before next-stage surgery was more common in PDAs group (28.3% versus 14.1%, p = 0.024). In addition, the PDAs group had a lower adjusted intensive care unit length of stay (4 days [95% CI, 2–8] versus 6 days [95% CI, 4–14]; p<0.001), a lower duration of inotropic support (39 vs 89 hours, p<0.001) and invasive respiratory support (55 vs 91 hours, p = 0.013) and a lower proportion of diuretic use at discharge (39.5% vs 70.7%; p<0.001).
Conclusion: In this large international comparative study, there was no difference in the primary end point between PDAs and mBTS. Although re interventions rate was higher in PDAs group, other markers of perioperative morbidity support PDAs as a reliable therapeutic strategy in selected patients with ductal-dependent PBF.

Keywords: Ductus arteriosus stent, Blalock-Taussig shunt, Congenital heart diseases

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O-056

A new self expandable valve; pulsta thv in patients with enlarged rvot and severe pulmonary regurgitation due to transannular patch

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Background and Aim: Severe pulmonary valve regurgitation after tetralogy of Fallot (TOF) surgical repair is a serious condition threatening the patient life. Percutaneous pulmonary valve implantation has emerged as an alternative to surgery in recent years. However, due to variability of the right ventricular outflow tract (RVOT) morphology, balloon-expandable valves do not fulfill the all requirements. Since the implantation of a balloon-expandable valve in a large RVOT has some technical difficulties, new alternatives have been investigated. Although investigations of the self-expandable valve are promising, there is no approved valve so far for the pulmonic position. A new self-expandable valve Pulsta(R) (Taewoong company, South Korea) provides a new opportunity for pulmonary valve implantation. Herein we presented early results of our experiences with Pulsta(R) valve in native RVOT's.

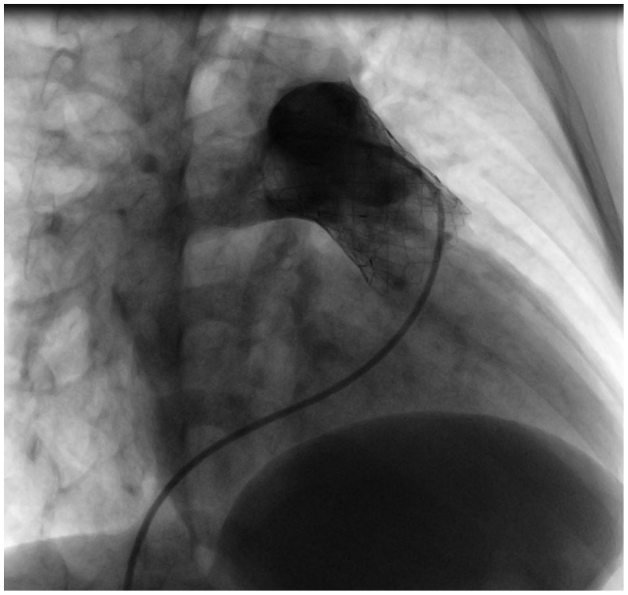
Methods: The patients with Tetralogy of Fallot who underwent total repair using transannular patch were recruited to the study. Previously defined criteria for the pulmonary valve implantation were also used for the inclusion criteria. The patient screening was performed by using transthoracic echocardiography. Main pulmonary 2 D diameter measurements smaller or equal than 30 mm is accepted as a prerequisite for inclusion. The patients with severe pulmonary regurgitation and dilatation of the right ventricle were evaluated in terms oft he main pulmonary artery diameters, flow velocity throughout the RVOT, Pulmonary artery branch peak velocity, and tricuspid valve functions. Ten patients were screened and considered candidates for the Pulsta(R) valve implantation. Cardiac Magnetic Resonance Imaging Angiography (MRI) was performed on all patients. MRI is used for the evaluation of the detailed anatomy of the right ventricle, RVOT, pulmonary artery, and branches. PPVI indication also was based on the MRI findings previously defined criteria.

Results: Pulsta(R) valve implantation was successfully performed in ten cases. No major complication such as migration, embolization, vascular problems, hemorrhage, coronary compression, and dysrhythmia was observed. Patients were discharged 48 hours after procedure without any event. No valvular regurgitation was detected more than mild in any case.

Conclusion: Pulsta (R) is a new, effective and safe alternative to surgery, in patients with severe regurgitation and enlarged native RVOT's. Long-term follow-up is necessary for the durability of the valve.

Keywords: Pulsta Self expandable valve Pulmonary Regurgitation Tetralogy of Fallot

Pulsta valve after implantation



No valvular regurgitation at control angiogram

O-057

Percutaneous edge to edge repair for systemic tricuspid valve regurgitation in patients with congenital heart disease: the first descriptive cohort

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Background and Aim: In adults with congenital heart diseases (ACHD) such as congenitally corrected transposition of the great arteries (ccTGA), transposition of the great arteries with surgical atrial switch (asTGA) or functionally single ventricles (SV), a systemic tricuspid regurgitation (STR) is frequently encountered.

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We report our experience of PETER in a series of 12 ACHD patients with STR using the MitraClip system.
Methods: Between June 2019 and September 2021, all french ACHD presenting symptomatic and severe systemic tricuspid valve regurgitation were discussed in a national heart team and refered for PETER in case of high surgical risk.
Results: At baseline, 7/12 patients were ≥ NYHA III. Standard femoral venous access in 11/12 patients. The systemic tricuspid valve was access directly in unpalliated SV though the RA (n = 3), while a SV patient with a Fontan circulation underwent a Fontan conduit puncture to reach the systemic atrium. Moreover, in TGA patients, either transseptal (ccTGA patients, n = 4) or transbaffle (asTGA patients, n = 3) puncture was performed (4). In one patient with situs inversus, levocardia and ccTGA, we used a direct left atrium surgical access via right thoracotomy
Mitraclip XTR devices were successfully implanted in patients 10/12 using standard technique (3,4) (Figure 1). Procedural failures included 1) one patient with ccTGA in whom the left atrium anatomy and size did not allow an appropriate positioning of the Mitraclip and 2) one patient with SV in whom a septal leaflet rupture occurred following clip release, leading to a severe STR; the patient died from refractory cardiac failure 1 week later. After a median follow-up of 12-months (range 1-25), no death occurred. Patients had significant reduction of STR and were clinically improved (9/10 patients were NYHA I or II). Compared to the pre-operative assessment, cardiac magnetic resonance performed in 6 patients after 6 months of follow-up showed a decrease in median RV end-diastolic volumes as well as an improvement of RV ejection fraction.
Conclusion: Our series confirms the technical feasibility efficiency of PETER in ACHD with systemic tricuspid valve regurgitation

Keywords: Edge to edge repair, Systemic tricuspid valve regurgitation

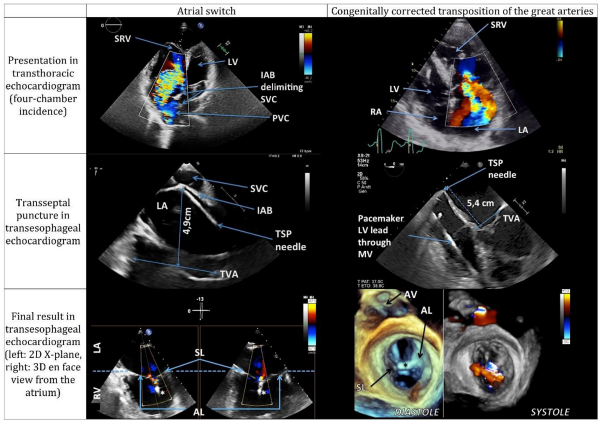

Figure 1 Per procedural views of PETER in different cases of systemic tricuspid valves regurgitation

O-058

Ductal stenting versus rvot stenting in infants with tetralogy of fallot and duct dependant pulmonary circulation

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Background and Aim: Infants born with Tetralogy of Fallot (ToF) and duct dependant pulmonary circulation require early intervention to secure stable pulmonary blood flow. In the recent years, transcatheter palliation with right ventricular outflow tract (RVOT) stenting or arterial ductal stenting have been used as a primary palliative approach. The aim of this study is to compare safety and outcome of ductal stenting vs RVOT stenting as an initial palliative approach in a cohort of patients diagnosed with ToF spectrum and compromised pulmonary blood flow.
Methods: Retrospective review of newborns with ToF who had undergone RVOT stenting vs PDA stenting at Evelina London and Royal Brompton Hospitals (London) from March 2010 to December 2021. Medical charts, echocardiography, catheterization, surgical data and follow-up were reviewed. Post-procedural clinical data including mechanical ventilation, need for ECMO, intensive care need and surgical technique was recorded.
Results: The review included 50 patients (21 PDA stenting, 29 RVOT stenting). There were no significant differences in sex, birth weight or gestational age. There was a trend of a higher proportion of associated cardiac malformations in patients undergoing RVOT stenting (62.5% vs 31.2%, p = 0.053).
The main indication for intervention was desaturation (100% in the PDA stent group vs 58.3% in the RVOT stent group, p = 0.003). The use of drug-eluting stents (DES) was significantly higher in the RVOT stent group (91.3% in the RVOT stent group vs 46.1% in the PDA stent group, p = 0.003). The rate of success was higher in those patients undergoing RVOT stenting (95.8%) when compared to PDA stenting (68.7%) (p = 0.02). The rate of complications was higher in the RVOT group (23% vs 11%, p = 0.03). The rate of reintervention was higher in the RVOT group (63% vs 36%, p = 0.01). Pulmonary arteries had grown better in the RVOT group prior surgical repair according to echoNakata index (p = 0.02). There were no major differences in pulmonary arteries size post-surgical repair.
Conclusion: RVOT stenting accounts higher rate of success when compared toPDA stenting and better pulmonary arteries growth. However, these patients require more reintervention prior surgical repair. Nomajor differences were found in both groups after surgical repair.

Keywords: RVOT stenting, PDA stenting, reintervention, pulmonary arteries growth

O-059

Transcatheter closure of ventricular septal defects with lifetech multifunctional occluder device (MF- konar)- a single-center experience

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Background and Aim: We aim to evaluate the safety and efficacy of the LifeTech multifunctional occluder device (MF- Konar) for the transcatheter closure of ventricular septal defects (VSD).
Methods: Clinical features, demographic characteristics and follow-up data of patients who underwent transcatheter VSD closure with MF-Konar between 2016 and 2021 were reviewed retrospectively.
Results: Transcatheter VSD closure with MF-Konar device was performed successfully in 56 out of 57 procedures. One patient had two different VSD closure procedures. The median age and weight of the patients were 15, 33 years (range 12 months– 60

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years) and 50 kg (range 6.5–78 kg), respectively. Twenty-four of the patients were boys, and 31 were girls. VSD was perimembranous in 50, muscular in 3, and postoperative residual in 3 patients. The mean Qp/Qs was 2.3 (range 1.5-2,8), mean pulmonary artery pressure was 18 mmHg (range 11-43). The narrowest VSD diameter on the angiogram was 6.5 (range 3-12) mm. The most commonly used device size was 10/8 mm (16 patients). The other devices were 12/10mm, 9/7mm, 14/12mm, 8/6 mm, 6/4 mm (12,12, 8,6 and 2 patients, respectively). The procedure and fluoroscopy times were 41 minutes (range 20-100) and 778 seconds (range 202 -2056). The procedure was performed via retrograde, antegrade, jugular, and hybrid routes in 28, 25, 2 and 1 patients, respectively. One patient was referred to surgery because of evolving complete atrioventricular block during the procedure. Mortality and major complications were not observed. Minor complications occurred in four patients (residual shunt, nodal rhythm, pericardial effusion and second-degree atrioventricular block). The mean follow-up duration was 216 days (33–537).
Conclusion: Transcatheter VSD closure with MF-Konar is an effective and safe treatment option in selected patients. This flexible, low profile and user-friendly device could prevent one of the most frustrating problems with transcatheter VSD closure, namely new-onset complete heart block.

Keywords: LifeTech multifunctional occluder, MF- Konar, transcatheter, ventricular septal defect closure

O-060

Pulmonary artery stenting after bidirectional cavopulmonary connection (BDCPC) in single ventricle patients: necessity and benefit

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Background and Aim: Comparison of single ventricle patients with and without PA-stent implantation post BDCPC regarding risk factors and pulmonary arterial growth.
Methods: Single centre, retrospective (2006-2021), longitudinal study on 144 patients (exclusion criteria: congenital abnormalities of the PAs, pulsatile partial CPC or temporary stent implantation). PA growth was assessed comparing angiography data pre BDCPC and pre-TCPC.
Results: A total of 44/144 (28%) patients received a PA-stent at median (IQR) 12 (1-136) days post BDCPC. Indications for cardiac catheterization after BDCPC surgery were suspected PA stenosis on echocardiography or haemodynamic instability. Main intraprocedural findings were suspected LPA compression from the ascending aorta (16/42, 38%); LPA hypoplasia (13/42, 31%); stenosis of surgical anastomosis (12/42, 27%); RPA torsion (1/40, 3%). 39/42 (93%) underwent LPA stenting, 3/42 (7%) (2 of which with right aortic arch) underwent RPA stenting. Two patients (2/42, 5%) suffered from acute LPA dissection at the site of surgical anastomosis during stent implantation but, after surgical repair, recovered well. Multiple regression analysis showed single LVs to receive less likely PA-stents than single-RVs (OR 0.41; $p = 0.05$), while reduced LPA/BSA (mm/m2) and higher diameter of aorta/DKS anastomosis prior to BDCPC were associated with increased likelihood (resp. OR 0.89, $p = 0.03$; OR 1.05, $p = 0.001$). Early BDCPC ($p = 0.003$) related with higher incidence of PA-stenting but did not add significantly in the model. Collective pulmonary artery diameters pre-BDCPC were lower in the PA-stent group: McGoon ($p<0.001$), Nakata ($p<0.001$). Lower-lobe-index was lower, but not significantly ($p = 0.08$). Pulmonary artery diameters increased equally in both groups but remained lower pre TCPC in the PA-stent group: McGoon ($p<0.001$), Nakata ($p = 0.009$), and Lower-lobe-index ($p = 0.003$). LPA and RPA growth was symmetrically in both groups. Stent re-dilatation was performed in 37/42 (90%) after 1 (0.8-1.6) year.
Conclusion: PA-stent implantation early after BDCPC is feasible and safe. Single RV, significantly larger ascending aorta / DKS anastomosis and small LPA prior to BDCPC are independent risk factors for secondary PA-stent implantation. Early BDCPC seems to affect only marginally PA growth. Fortunately, pulmonary artery diameters after PA-stent and stent dilatation showed significant growth together with the contralateral side, but the PA-system remained symmetrically smaller in the stent group.

Keywords: Single ventricle, pulmonary artery stenting, bidirectional cavopulmonary connection

Arrhythmia / Electrophysiology

O-061

Electrophsiologic study characteristics and risk assessment in patients with intermittent vs persistent ventricular preexcitation

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Background and Aim: Patients with ventricular pre-excitation (VPE) can be at increased risk for sudden cardiac death (SCD), and risk assessment of intermittent preexcitation is still can be challenging. The aim of this study was to share our experience about the electrophysiological characteristics of pediatric and young patients with intermittent and perisistent VPE and comparing their potential risk of SCD.
Methods: We retrospectively investigated 543 consecutive pediatric and young patients with VPE who underwent transcatheter EPS ± ablation between 2010-2020 in our center. Shortest preexcited R-R interval during inducted atrial fibrillation (Afib-SPERRI) was used for risk determination and a value of ≤ 250 ms indicated risk for SCD. If any doubt in assessing Afib-SPERRI, Accessory Pathway (AP) efective refractory period (APERP) was used for risk stratification, ≤250 ms indicating risk for SCD.
Results: Intermitant AP was seen in 97/543(17.9%) patients and 54(55.7%) were male. Their mean age was 12.07 ± 4.17 years and mean body weight 45.87 ± 20.20 kg. Persistant AP was seen in 446/543 (82.1%) patients and 315 (58%) were male. Their mean age was 12.33 ± 3,90 years and mean body weight 47.11 ± 19.40 kg. There was no statistically significant difference according to sex, age and weight of the patients. When comparing the ratio of symptomatic patients between two groups (56/97[58.3%] vs 297[66.4%], $p = 0.131$) or concommitant congenital heart defects between two groups (19/97[19.5%] vs 65/446[14.5%], $p = 0.243$) there was also no difference (Table-1). While APERP tended to be higher in intermittent VPE group ($p = 0.001$) there was no statistically significant difference in SPERRI during AFib between two groups ($p = 0.176$). 10/97(11.4%) patients with intermittant VPE were found having risk for SCD compared to 159/446(36.2%) patients with persistent VPE($p = 0,001$). While intermittent AP's tended to be more left sided (44/97; 45.4%), persistent AP's tended to be more septal (224/446; 52.1%) in location($p = 0.024$). Ablation results were satisfying with an acute sucess rate of 95.2% overall.

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Conclusion: Although statistically risk assessment of intermittent cases show a low risk for SCD, there are considerable cases with high risk measures and even history of cardiac arrest in one patient. For the treatment of the high risk patients, catheter ablation results are satisfactory.

Keywords: ventricular pre-excitation, WPW syndrome, intermittent preexcitation, risk assessment, pediatric ablation

Table 1: Patient and EPS characteristics compared between persistent and intermittent AP groups

	Total Patients n = 543[100%]	Patients with PersistentWPW n = 446 [82.1%]	Patients with Intermittent WPW n = 97 [17.9%]	*P*-value
Male-sex (*n*) [%]	315 [58%]	261[58.5%]	54[55.7%]	*0.606*
mean Age ± SD (years)	12.23 ± 3.86	12.33 ± 3,90	12.07 ± 4.17	*0.878*
mean Body Weight ± SD (kg)	46.89 ± 19.53	47.11 ± 19.40	45.87 ± 20.20	*0.635*
Symptomatic patients (*n*) [%]	353 [65%]	297 [66.4%]	56 [58.3%]	*0.131*
Concomittant CHD (*n*) [%]	84[15.4%]	65[14.5%]	19[19.5%]	*0.243*
Patients w multiple AP (*n*) [%]	**16 [2.9%]**	**16 [3.6%]**	**0 [0%]**	***0.041***
APERP at basal EPS ± SD (ms)	**295.38 ± 64.06**	**286.94 ± 55.19**	**344.4 ± 86.76**	***0.001***
Orthodromic AVRT induction(*n*) [%]	**371 [68.3%]**	**315[70.6%]**	**56[57.7%]**	***0.013***
Antidromic AVRT induction (*n*) [%]	38 [7%]	32[7.2%]	6[6.3%]	*0.729*
Sustained Afib induction (*n*) [%]	35 [6.4%]	25[5.6%]	10[10.3%]	*0.087*
mean Afib SPERRI ± SD (ms)	283.10 ± 60.55	281.81±59.96	297.38 ± 66.12	*0.176*
Patients with Afib SPERRI≤250ms(*n*) [%]	**146 [29.1%]**	**136[32.9%]**	**10 [11.4%]**	***0.001***
Adenosin Responsive AP (*n*) [%]	**38 [7%]**	**20[4.5%]**	**18[18.6%]**	***0.001***
Patients with risk of sudden death (*n*) [%]	**169 [32.3%]**	**159 [36.2%]**	**10 [11.9%]**	***0.001***
AP localization — **Right sided**	**87 [16.5%]**	**73 [17%]**	**14 [14.4%]**	***0.024***
AP localization — **Left sided**	**177 [33.6%]**	**133 [30.9%]**	**44 [45.4%]**	
AP localization — **Septal**	**263 [49.9%]**	**224 [52.1%]**	**39 [40.2%]**	

O-062

Assessment in a geographical cohort of primacy performance for risk stratification for sudden cardiac death in childhood hypertrophic cardiomyopathy

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Background and Aim: A new risk assessment algorithm for assessing risk for sudden cardiac death (SCD) in childhood hypertrophic cardiomyopathy (HCM), PRIMaCY, has been published with a C-statistic of 0.71 in a validation cohort from a tertiary centre registry, but no data on sensitivity and specificity were given. The value suggesting tertile with highest risk was >8.3% in both validation and derivation cohorts. We aimed to assess the performance of PRIMaCY in a Swedish national cohort of HCM diagnosed <19y age.

Methods: Freedom from SCD, resuscitated cardiac arrest or appropriate ICD discharge (SCD/CA) were analysed with Kaplan-Meier analysis, and C-statistic of ROC-curves analysed with SPSS statistical software. PRIMaCY scores were calculated via the on-line calculator and results with and without including results of genetic testing were compared.

Results: The cohort contained 138 cases of HCM without syndrome-association with sufficient data for PRIMaCY, male = 67%, age at diagnosis median 10.6y [IQR 4.0-14.7], and maximal wall thickness at diagnosis 14mm [11-19]. Follow-up was 10.6y [4.0-14.7], 135 had at least five year of follow-up and were used for performance-calculations, with 11 SCD/CA-events during the first five years. PRIMaCY without genetic information as a continuous function had a C-statistic of 0.71 (95%CI 0.51-0.90), p = 0.024, the value delineating lowest tertile was <5.4, whereas the upper tertile was >9.0. PRIMaCY with genetic information had C-statistic of 0.67 (0.45-0.89), p = 0.07, and upper tertile had values >11.3. A potential cut-off of >8.3% had a C-statistic of 0.57, p = 0.42, whereas a higher cut-off of >9 had 0.66 (0.49-0.84), p = 0.07, a relative risk of 4.1 (1.1-18.2), sensitivity of 82% (52-97), specificity of 52% (41-63), positive predictive value of 20% (11-35), and negative predictive value of 95% (84-99). Even in the tertile with lowest risk assessment, <5.41%, there were two SCD/CA.

Conclusion: Our results align with those of the original validation cohort, but suggest a slightly higher cut-off for high-risk, however even with that higher cut-off specificity is low in our cohort. Adding genetic information does not improve performance The PRIMaCY algorithm appears not specific enough to be used for decision of ICD-implantation on its own, but needs to be considered together with other clinical parameters.

Keywords: sudden cardiac death, risk stratification, hypertrophic cardiomyopathy, PRIMaCY

O-063

Arrhythmogenic cardiomyopathy in childhood

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Background and Aim: Arrhythmogenic cardiomyopathy (AC) is considered a disease most relevant for the adult population. However, there is lacking data on both the incidence of events in children with AC and the penetrance of disease in AC genotype positive pediatric family members. Current guidelines recommend that first-degree relatives of an individual with confirmed AC undergo diagnostic screening starting at 10-12 years of age, but the clinical value of this approach has not been systematically evaluated.

We aimed to explore the incidence of events in pediatric patients with AC and to estimate the penetrance of AC disease in genotype positive pediatric family members.

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Methods: We performed a retrospective cohort study including all pediatric AC patients (age ≤18 years) followed at the Department of Pediatric Cardiology, Oslo University Hospital, Rikshospitalet, Norway, between 2007 and 2021. Both probands and genotype positive family members identified by family screening were included. Penetrance of AC disease was defined as fulfilling definite AC diagnosis by the 2010 Task Force Criteria, encompassing electrocardiographic, structural, histological, and arrhythmic characteristics. Major adverse cardiac events were defined as cardiac death, heart transplantation or severe ventricular arrhythmic events.
Results: We included 61 pediatric AC patients (10 probands, 51 family members, age 10 ± 5 years). Definite disease criteria were fulfilled in 19 patients (31%). Mean age at time of definite diagnosis was 14 ± 4 years, and 7 patients (11%) were ≤12 years. Thirteen patients (21%) undervent major adverse cardiac events, 6 of them ≤12 years of age (Figure 1). Nine patients experienced a severe ventricular arrhythmic event and 5 patients developed end-stage heart failure necessitating heart transplantation.
At genetic diagnosis, 5 (10%) of pediatric family members had AC disease. Cardiac penetrance was 18% at 6 ± 4 years of follow-up at a mean age of 15 ± 6 years. Three (33%) of the family members meeting diagnostic criteria for AC experienced a major adverse cardiac event at a mean age of 13 ± 6 years.
Conclusion: Our results indicate that AC is a significant pediatric disease. Cardiac penetrance was high in genotype positive pediatric family members with frequent major adverse cardiac events, suggesting that AC screening should begin at a younger age than currently recommended.
Keywords: Arrhythmogenic cardiomyopathy, sudden cardiac death, family screening, penetrance

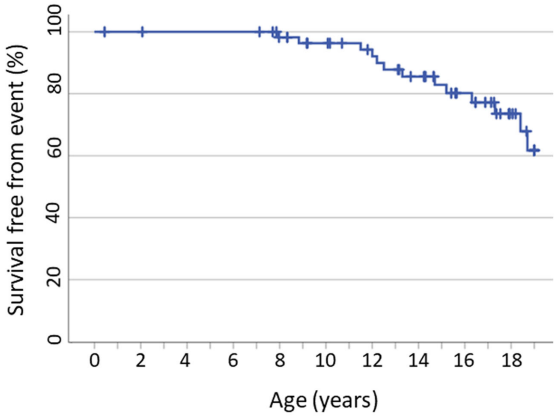

Figure 1: Curves of survival free from major adverse cardiac event in 61 pediatric arrhythmogenic cardiomyopathy patients. Thirteen (21%) of the patients had experienced event at ≤18 years of age. Mean age at time of event was 13.8±3.4 years.

O-064

Electrophysiologic characteristics and catheter ablation results of fascicular ventricular tachycardia in children

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Background and Aim: Fascicular ventricular tachycardia (FVT) is 10–15% of total pediatric ventricular arrhythmias. The majority of FVTs originate from the posterior fascicle.
Methods: We studied 31 patients in whom FVT was successfully eliminated by ablation. Sinus ECGs before ablation were compared with those after ablation. VT ECGs of the patients were evaluated. Electrophysiological data and ablation details were also noted. Follow-up data were reviewed.
Results: Thirty-one patients with a mean age of 13 and a weight of 52.5 were included (22M/9F). Thirty-four substrate ablations were performed in these 31 patients. (30 posterior FVT, 3 anterior FVT and 1 upper-septal FVT). Only 5 patients were referred to our center with a diagnosis of FVT. Muscular bands were detected in a total of 14 patients (45.2%). When sinus ECGs were compared before and after the procedure, no significant changes in axis, interval and morphology were detected. Left ventricular stimulation was required for VT stimulation in 3 patients. Ablation was performed during VT in 21 patients (67.8%). The mean procedure time and fluoroscopy time were 150+/-43 minutes and 3.8+/-2.4 minutes, respectively. In 4 patients, a different VT morphology was induced after primary substrate ablation. Typical AVNRT was stimulated in 5 patients. Recurrence was observed in only 3 patients during a mean follow-up of 56 months.
Conclusion: Fascicular tachycardia is a distinct subgroup of idiopathic VT that may be confused with SVT with or without aberrancy. The ECG diagnosis may remain challenging. AVNRT was the most common type of SVT accompanying FVT. Both can be stimulated by atrial induction. During EPS, AVNRT can be stimulated before FVT. If the tachycardia in the clinic cannot be documented the procedure can be terminated by simply performing AVNRT ablation. Morphology change in QRS during or after ablation of FVT has been reported in the literature between 3.6 and 5.1%. These changes may explain with theories such as the stimulation of tachycardia over other fascicles, the presence of more than one bystander connection, and the change in the spread of the existing tachycardia.

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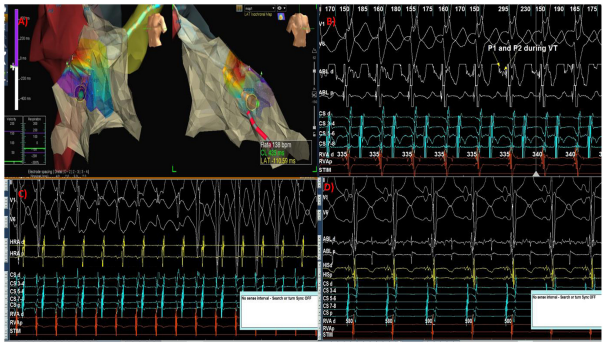
Ablation is a safe and effective option for the treatment of FVT.

Keywords: Fascicular ventricular tachycardia, ablation, posterior fascicle

Ablation Images



A) Image of the classic ablation location for posterior FVT in the Left Anterior Oblique and Right Anterior Oblique positions B) P1 and P2 signals at the distal electrode of the ablation catheter during VT C) QRS morphology change while VT continues D) Appearance of the diastolic potentials during mapping

O-065

Retrospective single-centre study of paediatric catheter ablation in the ERA of 3D navigation and mapping

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Background and Aim: Catheter ablation is effectively used to treat paediatric arrhythmias. We performed a retrospective study on ablation outcomes using non-fluoroscopic 3D navigation and mapping systems and compared them to the previous fluoroscopy era.
Methods: Data were retrieved from the institutional patient database, outpatient medical reports, referring paediatric cardiologists and by structured phone calls to patients/their legal guardians. Long-term follow-up (>10, median 39 months) was available in 70.4% of patients. Results were compared to our previously published data from two fluoroscopic navigation eras (1993-2005 and 2006-2010, doi:10.1093/europace/euu087). In all three ablation periods fluoroscopy was used at the discretion of the operating physician according to the ALARA principle (As Low As Reasonably Achievable).
Results: From 2010 to 2020 (3D navigation and mapping era) 772 ablation procedures were performed in 696 patients (girls 48%, structural heart disease in 5%) to treat 739 arrhythmogenic substrates (accessory pathways 59%, AVNRT 32%). Radiofrequency (96%) or cryo ablation (4%) were used along with the LocaLisa (88%), Ensite/NAVx (7%, see Picture) and Carto (5%) navigation/mapping systems, resp. Multiple procedures were necessary in 8% of patients. Long-term success rate was 85.5% for accessory pathways (right sided 73.2%, septal 81.2% and left-sided 92.8%, P = 0.002) and 89.5% for AVNRT. Both the procedure and fluoroscopy times were significantly lower as compared to previous fluoroscopy eras. Acute success rate was, however, not improved (Table).
Conclusion: Procedure duration decreased significantly over time. Use of non-fluoroscopic 3D navigation and mapping systems led to a dramatic reduction in fluoroscopy time. Long-term procedure success is, however, still limited to 85 – 90 % for the two most common substrates – accessory pathways and AVNRT. Major differences in efficacy exist between different pathway locations. Such information is important for patient counselling and follow-up.

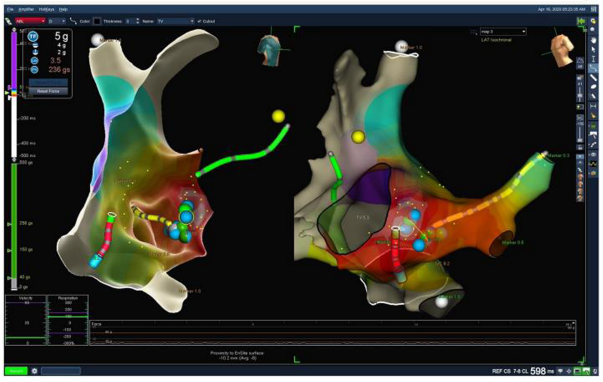
Keywords: ablation, 3D navigation and mapping

Procedure, Fluoroscopy time and Acute success through different ablation eras.

	Median (IQR) time [min.]		Acute success [%]	
Era	Procedure	Fluoroscopy	AP	AVNRT
1993-2005*	154 (120-210)	24 (14.4-42.3)	84.6	98.2
2006-2010**	110 (80-145)	14 (7.0-20.7)	91.1	98.9
2010-2020†	90 (69-130)	1.1 (0.5-2.6)	90.2	98.8
P	<0.001	<0.001	NS	NS

*Early/**Late fluoroscopy era; †3D navigation era;

Mapping and ablation of right posteroseptal pathway using the EnSite/NAVx system



O-066

Long-term fate of an unselected cohort of congenital long QT syndrome patients diagnosed in childhood

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Background and Aim: Congenital long QT syndrome (LQTS) is a genetically heterogeneous disorder with type-specific risk of major arrhythmic event (MAE). We aimed to perform a single-centre retrospective analysis of LQTS patients diagnosed in childhood.
Methods: All paediatric patients (N = 224, female 119, 53 %) diagnosed with LQTS (Schwartz score ≥1.5 points and/or presence of a pathogenic or likely pathogenic genetic variant) in the absence of structural heart disease between July 1985 and December 2021 at median (IQR) age 11.9 (6.8-14.6) years were included. Data were retrieved from the institutional database and medical records and cross-mapped with the National Death Registry. Patients were followed-up for a median (IQR) of 7.7 (2.7-16.4) years.
Results: Reasons for presentation were positive family history of LQTS (N = 66, 29.5%), syncope (N = 53 cases, 23.7%), incidental finding of prolonged QTc (N = 37 children, 16.5%), positive pre-participation screening (N = 30, 13.4%), sudden cardiac arrest (N = 26,11.6%) and palpitations/bradycardia (N = 6, 2.7% each). The longest individual QTc interval was median (IQR) 482ms (460-516ms), the median (IQR) Schwartz score was 4.0 (3.0–5.0) points. Likely pathogenic or pathogenic variants were found in 119/159 tested patients (74.8%): KCNQ1 mutations in 63 patients (39.6%), KCNH2 in 33 (20.8%), SCN5A in 12 (7.5%), KCNE1 in 5 (3.1%), KCNJ2 in 4 (2.5%) and SNTA1 in 2 patients

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(1.3%). Betablockers (BB) were administered in 202 patients (90.2 %) with proportion of non-selective BB increasing from 16.7 % to 58.5 % comparing periods 1985-2015 and 2016 onwards (p<0.001). Pacemaker was implanted in 24 patients (10.7%) and ICD in 30 (13.4%, primary prevention in 10/30). Eleven patients died from cardiovascular cause (4.9%, sudden death in 10/11) yielding a 5/10/20 years survival probability of 97.7/95.1/91.7%. Freedom from MAE defined as either sudden cardiac death/arrest or appropriate ICD shock after diagnosis of LQTS was 93.5/88.9/85.5%. MAE was independently predicted by QTc duration (HR 1.018, p<0.001) and presence of LQT3 (HR 2.816, p = 0.040).

Conclusion: Patients with LQTS diagnosed in childhood had a long-term survival probability over 90%. QTc duration was a major predictor of MAE. BB therapy has shifted to non-selective BB over time. Genetic testing had a high diagnostic yield.

(Supported by: MHCZ–DRO, Motol University Hospital, Prague, Czech Republic 00064203)

Keywords: arrhythmias, long QT syndrome, sudden cardiac death

Preventive

O-067

Physical fitness, bmi, health-related quality of life and mental concentration – cross-sectional cardiovascular risk data of a large school cohort

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Background and Aim: Low physical fitness (PF) levels and elevated BMI values already in young age are known cardiovascular risks and may contribute to impairment of health-related quality of life (HRQOL) and mental health. The aim of this investigation was to evaluate the impact of BMI on PF levels in a large cohort of primary school children in a rural region of Bavaria (Berchtesgadener Land). Furthermore, we wanted to determine the association between PF, concentration level and HRQOL.

Methods: Cross-sectional data of a prospective study of 6533 healthy primary school children (8.73 ± 1.18 years, 3285 ♀) were analyzed. PF was assessed by a standardized test battery, including PF tests of the FITNESSGRAM (push-ups, curl-ups, PACER shuttle run), standing long jump and handgrip strength. Children were categorized into 4 weight classes, according to German reference values: underweight (< 10. percentile (PZ)), normal weight (≥ 10. ≤ 90. PZ), overweight (> 90. ≤ 97. PZ) and obesity (> 97. PZ). In a sub-cohort, concentration was measured with the d2-R test, and KINDL questionnaires were used to determine HRQOL.

Results: 13% of all primary school children were categorized as overweight or obese, boys had a higher incidence for elevated BMI values (p <.001). Overweight and obese children performed significantly worse in all of the PF tests when compared to children with normal weight or underweight (p <.001). In both boys and girls, HRQOL was lower in the obese group and d2-R total score was significantly lower in obese boys (p <.001). Multiple linear regression models showed a strong impact of VO2max on mental concentration levels (ß = 0.10, p <.001) and HRQOL (ß = 0.21, p <.001).

Conclusion: Even in a rural child cohort, cardiovascular risk factors play an important role. Moreover, HRQOL and mental abilities are influenced by these risk factors, contributing to school performance. Thus, cardiovascular fitness should be more promoted, e.g., in schools, to reduce cardiovascular risk in children, to improve HRQOL and possibly also mental health.

Keywords: cardiovascular health; physical fitness; primary school; concentration; psychosocial health

O-068

Impact of a transition education program on health-related quality of life in adolescents and young adults with congenital heart disease

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Background and Aim: Recent advances in the field of congenital heart disease (CHD) led to an improved prognosis of the patients and in consequence the growth of a new population: the grown up with congenital heart disease. Until recently, more than 50% of these patients were lost to follow up because of the lack of specialized structures. The critical moment is the transition between paediatric and adult unit. Therapeutic education is crucial to solve this issue by helping patients to become independent and responsible. The TRANSITION randomized trial aims to assess the impact of a transition education program on health-related quality of life (HRQoL) of adolescents and young adults with CHD.

Methods: Multicentre, randomised, controlled, parallel arm study in patients with CHD aged from 13 to 25 years old. Patients were randomised into 2 groups (transition education program vs. no intervention). The primary outcome was the change in self-reported HRQoL (PedsQL generic questionnaire) between baseline and 12-month follow-up. Using an intention-to-treat analysis, a total of 200 patients were required to observe a significant increase of the overall self-reported HRQoL score (80% power, 5% α-risk). The secondary outcomes were proxy-reported HRQoL scores, clinical outcomes, cardiopulmonary exercise test parameters (VO2max, VE/VCO2 slope, anaerobic threshold, oxygen pulse), the level of physical activity, the level of knowledge of the disease using the Leuven knowledge questionnaire for CHD, physical status and psychological status. Trial registration: NCT03005626. Study design, DOI: 10.1186/s12955-021-01668-1.

Results: A total of 189 patients were enrolled in the trial (mean age 18.6 ± 3.6 years, sex ratio = 1). We observed a significant difference of 3.9 ± 1.1 points (P = 0.04) in the primary outcome, i.e., the self-reported PedsQL total score, in favour of the transition education group. Significant differences were also observed in the self-reported PedsQL psychosocial score (4.2 ± 1.2 points, P = 0.049), the proxy-reported physical score (6 ± 2.3 points, P = 0.01), and the disease knowledge score (4.9 ± 0.7 points, P<0.001). No significant group differences were observed in the clinical or CPET parameters.

Conclusion: The HRQoL of adolescents and young adults with CHD was improved by our transition education program. The

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supervision of the program by a specialist nurse as"transition care manager" is probably an interesting operating model to follow.

Keywords: transition; therapeutic education; congenital heart disease; quality of life; adolescent.

Transition group session

O-069

Reference values for cardiorespiratory fitness in children and adolescents with congenital heart disease

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Background and Aim: Cardiopulmonary Exercise Testing (CPET) is considered the gold standard assessment of peak oxygen uptake (VO_2peak), and thereby cardiorespiratory fitness, also for children with congenital heart disease (CHD). The common practice is to compare the results with reference values from healthy children. Unfortunately, this practice does not account for complexity of the heart defect, and it may also be demotivating for the children. Against this background, we aimed to describe VO_2peak in relation to age and gender in the most prevalent types of CHD, and to establish reference values for VO_2peak.

Methods: From 1996 to 2020, a total of 1470 CPETs until exhaustion were performed by 988 patients (40 % female) at two hospitals in Norway. The patients were subsequently divided into three subgroups: simple defects (VSD, ASD, CoA, LVOTO; n = 494, 40 % female), moderate complex defects (TGA, Fallot; n = 308, 41 % female) and univentricular defects with a Fontan circulation (n = 186, 42 % female). A two-sample t-test was performed to compare VO_2peak between male and female within the subgroups, and VO_2peak between the different subgroups.

Results: The preliminary results for mean VO_2peak ($mL \cdot kg^{-1} \cdot min^{-1}$) with increasing age are presented in Figure 1. There was a significant gender difference in VO_2peak in all subgroups (male>female): simple defects (mean difference: 7.7, p<.001), moderate complex defects (mean difference: 6.1, p<.001), and univentricular defects (mean difference: 3.6, p<.001). In addition, VO_2peak was significantly lower in the patients with univentricular defects with a Fontan circulation compared to those with simple defects (mean difference: 11.1, p<.001), and moderate complex defects (mean difference: 7.5, p<.001).

Conclusion: We present reference values for VO_2peak in the most prevalent types of CHD. Using reference values related to disease complexity might improve clinical decision-making of the children.

Keywords: Cardiopulmonary Exercise Test, Congenital Heart Disease, Cardiorespiratory Fitness

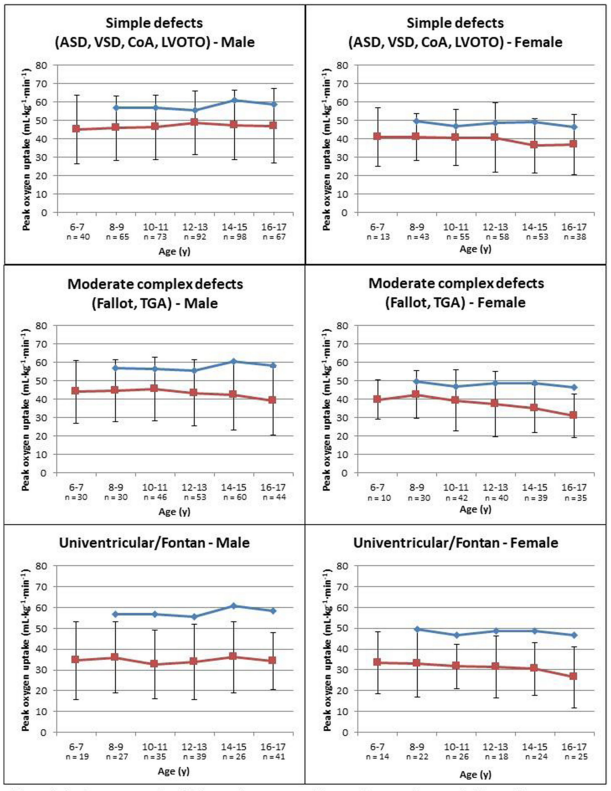

Figure 1: Peak oxygen uptake with increasing age according to subgroup of congenital heart disease presented as **mean (±2SD)**. Blue line represents mean peak oxygen uptake of Norwegian reference data in healthy children (Fredriksen P.M et al. (1999) Eur J Appl Physiol 80;409-416).

O-070

Return to play after SARS-COV-2 infection: safe sport for children and adolescents

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Background and Aim: With the progressive spread of the coronavirus among the youngest and the need for a safe resumption of physical activity, several protocols have been proposed for healed athletes. The aim of the present study is to evaluate the presence of cardio-respiratory complications in the pediatric population after mild or asymptomatic SARS-CoV-2 infection.

Methods: From January 2021 the protocols of the Italian Sports Medical Federation were applied to all the children and adolescents who came to our observation for return-to- play after covid infection. The protocols take into account the severity of the

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infection. In case of mild or asymptomatic infection echocardiogram, electrocardiogram, treadmill ECG test and pulmonary function tests were performed.
Results: From January 2021 to November 2021, 169 children and adolescents (mean age 14+3 ys; male = 92) with previous SARS-CoV-2 infection were evaluated according to the protocols in force after an average time of 48 + 13 days from SARS-CoV-2 swab negativity. 60,3% of the subjects (N = 102) reported an asymptomatic infection; 39,7 % reported a mild symptomatic infection. Results of lung function test have exceeded the limit of 80% of the theoretical value in all patients. No subject presented evidence of cardiovascular function impairment.
Conclusion: The data obtained showed that, in the pediatric population, mild coronavirus infection does not cause cardiorespiratory complications in the short and medium term. This reinforce the evidence from preliminary studies that return to play after Coronavirus infection seems to be safe and must be strongly recommended. It will therefore be possible to assess the possibility of lightening or even suspending these evaluations in the cases of mild coronavirus infection in the pediatric age groups.

Keywords: SARS-CoC-2 infection, functional capacity, pediatrics

O-071
Cognitive function in children with CHD impaired in a direct twin comparison

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Background and Aim: In children with congenital heart defect (CHD), it seems reasonable to assume that cyanosis or abnormalities in blood flow may have a potential impact on cognitive function as well as surgical or drug treatment. Besides this, cognitive function is dependent on various external influences such as socioeconomic aspects, environmental and family influences the children live. Therefore, to exclude these external influences we compared cognitive function of twin siblings with and without CHD.
Methods: Cognitive function was measured as part of the nationwide ongoing twin study "SameSame", investigating neurodevelopment (cognitive function, motor function, mental health and health-related quality of life). 43 pairs of twins (one with CHD and the other without, 48 girls (55.8%), 8.1 ± 3.6 years) have completed the Wechsler Intelligence Scale for cognitive function so far (WISC V, WPPSI IV). The tests consist of 10 age-specific parts covering the functions verbal comprehension, visual-spatial reasoning, fluid reasoning, working memory and processing performance as well as the overall intelligence quotient (IQ). The data of the twin siblings among each other were compared using the student t-test for paired samples. In addition median and percentiles was used.
Results: Children with CHD (7.0% simple/44.2% moderate/48.8% complex CHD severity) showed significantly reduced IQ scores in visual-spatial reasoning with a mean difference (MD) of -6.8 points (p>0.001), in processing performance (MD -5.9, p = 0.010) and overall IQ (MD -5.4, p = 0.006). The reduced parts in children with CHD in median (M) overall IQ -4.0 points with percentile 25. of -7.0 points, in visual-spatial reasoning (M -8.0; percentile 25.: -11.75) and processing performance (M -3.0; percentile 25.: -11.25) showed the differences significant clear.
Conclusion: Children with CHD showed impaired cognitive function in overall IQ, visual-spatial reasoning and processing performance compared to their healthy twin siblings. The impairments are particularly evident in the 25th percentile. These findings underline the importance of early developmental assessment to identify the 25% of children with CHD who have impairments to intervene with targeted early intervention.

Keywords: neurodevelopment, cognitive function, twins, children with CHD, prevention

O-072
Applying swiss watch high tech and precision to serve high unmet needs of neonates born with congenital heart defects (CHD)

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Background and Aim: INTRODUCTION: Congenital heart disease (CHD) affects 1 in 100 newborns each year. In 5% of cases, the severity of CHD is life threatening and responsible for the largest proportion of mortality caused by birth defects. The majority of malformations are treated by complete corrective cardiac surgery but due to size and/or type of malformations, open-heart surgery at birth may not be possible. Pulmonary Artery Banding (PAB) is used as a palliative technique to control pulmonary blood flow. Maintaining a proper pulmonary flow over time is essential to avoid long term complication and remains an area of high unmet need in particular in neonates. With the current traditional banding techniques using fixed band, there is a need for multiple serial adjustments with age, meaning several re-operation(s) which raises morbi/mortality.
EspeRare, using philanthropic and public funding, has committed to develop NeoCare, an active implanted medical device (AIMD) which allows a remote regulation of blood flow non-invasively.
Methods: NeoCare geometry was designed to achieve a safe and efficient banding of vessels in order to remotely control pulmonary flow and to match neonatal anatomy. Its key attributes and specifications were set by a worldwide consortium of experts combining pediatric cardiac surgeons, pediatric cardiologists, and engineers from HEIG-VD/IICT.
Results: NeoCare functional prototypes and control units have been produced. The system includes a miniature actuated component composed of a banding system linked to a subcutaneous antenna by an electronic cable, a control unit connected to an external antenna to remotely regulate the occlusion of the vessel. A software was developed to collect information on the piston 's position which precisely defines the degree of occlusion.
NeoCare technology includes unique know-how allowing non-invasive adjustments of blood flow applying the Swiss watch technology to medical needs. The precision of the piston is of 0,1 mm and the device can be applied to babies' arteries as little as 11 mm of diameter.

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Conclusion: Additional efforts are needed to transform these technological breakthroughs to industrial scale. Pending funding, EspeRare is committed to continue the development of NeoCare so that patients with CHD can benefit from this technology.

Keywords: Congenital Heart Disease, Pulmonary artery Banding, NeoCare, Remote Control, Pulmonary Flow

Other

O-073

Congenital heart defects and their association with maternal diabetes and overweight during pregnancy: a register-based study from finland

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Background and Aim: Congenital heart defects (CHDs) are the most common congenital malformations affecting 1% of newborns. Two significant environmental risk factors related to CHDs are maternal overweight and diabetes. The aim of this study was to analyze the role of maternal diabetes and overweight as risk factors for CHDs in a register-based setting in Finland.
Methods: This national register-based study included all 644 073 children born 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. Logistic regression analyses were used to analyze the association between maternal overweight and diabetes and offspring's CHDs when adjusted to maternal diabetes, maternal BMI, parity, maternal smoking status, infants' sex, gestational age, and maternal age. P-value <0.05 was considered as statistically significant.
Results: A total of 640 217 children were included in the analysis. At least one isolated CHD was detected in 1.6% of children. Maternal type I (OR 2.6 (95% CI 2.2-3.0)) and gestational diabetes (OR 1.1 (95% CI 1.0-1.1)) and the morbid obesity (BMI>40) (OR 1.1 (95% CI 0.96-1.3)) were associated with an increased risk of any CHD. In a subgroup analysis of 8 different heart defect types, of which septal anomalies were most common. Type I diabetes was most significantly associated with transposition of great arteries (OR 5.9 (95% CI 2.3-15), left and right outflow tract obstruction (OR 3.0 (95% CI 2.0-4.5) and OR 2.4 (95% CI 1.6-3.7) respectively) and septal anomalies (OR 2.6 (95% CI 2.2-3.1). Obesity was associated with left and right outflow tract obstruction (OR 1.8 (95% CI 1.2-2.8) and OR 1.4 (95% CI 1.1-1.7)) respectively.
Conclusion: In this nationwide Finnish register-based study, maternal type 1 diabetes, gestational diabetes, and obesity were identified as significant risk factors for CHD in the offspring. Knowledge about the increased risk in specific subgroups can aid in implementing targeted prevention strategies and may be of help in further research on the pathophysiological developmental effects underlying these lesions.

Keywords: congenital, heart, defect, diabetes, obesity, overweight

O-074

The congenital cardiology cloud – proof of feasibility of the first telemedical network for pediatric cardiology in germany

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Background and Aim: Recent technological developments offer a multitude of new options for innovative approaches in patient care. Especially during COVID-19-pandemic, use of telemedical infrastructure has worldwide become a crucial part of pandemic containment. For an optimate interplay based on data secure exchange of diagnostical data (DD) between patients with complex congenital heart disease, ambulatory care and hospital care, we successfully implemented the first telemedical network for pediatric cardiology in Germany, the Congenital Cardiology Cloud (CCC). This study proofs the CCC´s feasibility and analyses its technical characteristics as well as its implementation in routine clinical work.
Methods: Analysis of implementation and technical characteristics comprised numbers of incoming/outgoing data, related file types, treatment options for tele medically processed patients and patient classification with respect to severity of disease. Proof of feasibility was made by the analysis of successful telemedical transmissions of discharge documents at the end of observation period (03/2020-10/2020).
Results: Analysis of bilateral telemedical traffic showed a number of 1178 files for a total of 349 patients, favouring transmissions towards the clinic (782 files). Incoming traffic was predominantly characterised by diagnostical data (88%), consisting of a multitude of file types, whereas 94% of the dispatched data corresponded to discharge letters. Number of teleconsultations counted up to 61 during observation period, with a necessary subsequent treatment in 90% of the presented cases. Tele medically processed patients generally showed to be more complex (severe chronic heart disease 42% vs. 24%). From a total number of 422 discharged patients, 323 had a successful telemedical transmission of their discharge documents, resulting in a rate of success of 97,6% at the end of observation period (pic 1).
Conclusion: Implementation of the first telemedical network for pediatric cardiology in Germany proofs recent technological developments to successfully enable innovative patient care, connecting the ambulatory and hospital sector for a joint patient advice. Transferred diagnostical data facilitates mutual assessment and predominantly involves more complex cases, resulting in a subsequent necessary hospitalization. The introduction of possible governmentally guided refinancing concepts will show its long-term feasibility.

Keywords: pediatric cardiology, telemedicine, tele medical network, telecardiology, congenital heart disease, cloud

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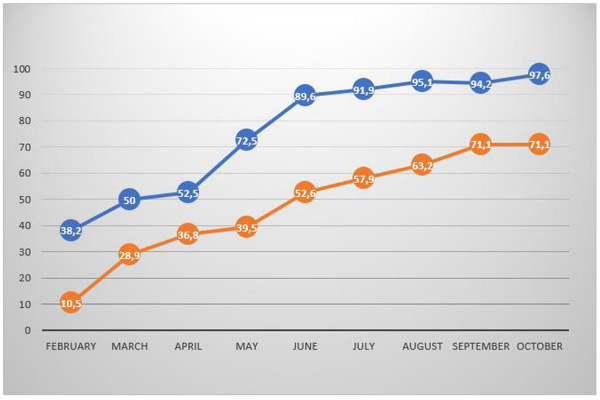
Comparison of successful tele medical transmissions to conncetion of referring physicians



Picture 1: Successful tele medical transmission of discharge documents (blue) compared to tele medically connected referring physicians of the University Competence Network for Congenital Heart Disease (orange)

O-075

National paediatric mortality secondary to cardiac causes: an irish experience

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Background and Aim: This study aims to describe the characteristics of paediatric mortalities due to all cardiac causes, from 2011 to 2018 in the Republic of Ireland.
Methods: A retrospective descriptive study of all cardiac causes contributing to Paediatric Mortality was conducted. Mortality data were obtained from National Paediatric Mortality Registry (NPMR), The National Institute for Cardiovascular Outcomes Research (NICOR) database and Cardiac Nurse Specialist patient records, from 2011 to 2018. Medical records were reviewed to obtain relevant clinical demographic and outcome data.
Results: A total of 304 paediatric deaths secondary to cardiac causes were recorded from 2011 to 2018, with an average mortality rate of 0.56 per 1000 live births per year during the study period. There were 162 male and 142 female, with a median age of death of 1.7months old, ranging from fourteen minutes to 16 years old (Figure 1). The leading cardiac cause of death among the paediatric cohort was congenital structural heart diseases (Figure 2), accounted for 83% (252/304). Inherited cardiac conditions including cardiomyopathies and cardiac arrhythmias accounted for 10% (30/304), while acquired cardiac conditions including myocarditis and complete heart block secondary to autoimmune disease accounted for 7.2% (22/304).
Conclusion: This is the first contemporaneous descriptive study of all cardiac causes contributing to Paediatric deaths at National level.

Keywords: Paediatric mortality, Congenital heart disease, Inherited cardiac conditions, Acquired cardiac conditions.

Figure 1: Paediatic Cardiac Mortality in the Republic of Ireland by Age Group, from 2011 to 2018.

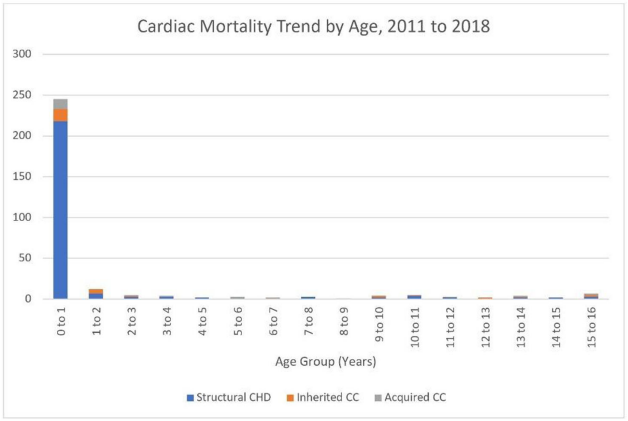
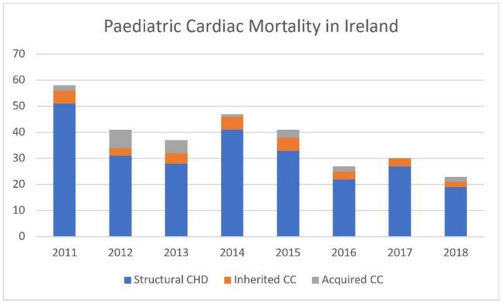


Figure 2: Annual summary of Paediatric Cardiac Mortality in the Republic of Ireland, from 2011 to 2018.



Morphology

O-076

Outflow anatomy in double-outlet right ventricle

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Background and Aim: In double-outlet right ventricle (DORV) both great arteries arise completely or nearly completely from the RV. Two-ventricle outcome is possible in most patients and the anatomy of the outflow is an important determinant of the surgical approach.
Methods: We examined 15 DORV heart specimens with emphasis on the outflow anatomy. The position/orientation of the infundibular septum (IS), position of arterial roots, ventricular septal defect (VSD)-arterial root relationship (if present) and outflow obstruction were assessed.
Results: 3/15 hearts were not amenable to a 2-ventricle repair (mitral atresia with intact septum/muscular VSD -2, mitral stenosis, LV hypoplasia, membranous VSD -1) while 12/15 hearts were candidates and are described. 8 had isolated DORV, 2 straddling

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mitral valve (SMV) and 2 complete common atrioventricular canal (CAVC). The VSD was between the limbs of septal band (outlet) in 10 hearts (isolated, SMV) and AV canal type plus outlet extension in 2 with CAVC. The VSD was subaortic in 8, including CAVC, subpulmonary in 3 (including SMV) and doubly committed in 1 case. The IS joined the superior ramus of septal band in all subaortic VSDs with D-aorta, being more horizontal with a posterior aorta and more vertical with side-by-side or anterior aorta. The IS was rightward over the RV cavity and vertical in the subpulmonary VSDs and absent in the doubly committed VSD. With L-aorta the IS was vertical and above the RV when the VSD was subaortic. Outflow obstruction in 7 (subaortic - 1, subpulmonary - 6) was due to uneven outflow division with deviation of IS in 6 cases and due to SMV attachments in 1 case. *Conclusion:* The orientation of the IS and position of the arterial roots determine the outlet VSD-arterial root relationship in DORV. The orientation (vertical/horizontal) of the IS is closely linked with the relative positions of the arterial roots (side-by-side, anterior-posterior). Uneven division of the outflow with IS deviation is associated with outflow obstruction. Straddling AV valve leaflets can be associated with obstruction of the VSD and/or outflow. Thus, identification of IS orientation and AV valve morphology is of utmost importance for adequate planning of therapeutic interventions.

Keywords: Double Outlet Right Ventricle, infundibulum septum, clinical implications

Other

O-077

Improved endothelialization in the regenerative small diameter vascular grafts with SNAP loaded nanoparticles

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Background and Aim: Synthetic small diameter vascular grafts (SDVGs, <6 mm internal diameter) exhibit a high failure rate after in vivo implantation. Therefore, in situ regeneration of vascular grafts is an option to reduce undesired complications and improve long-term patency. Nitric oxide (NO) is an endogenous signaling molecule encouraging endothelial cell migration and proliferation while preventing bacterial infection and thrombus formation.
Methods: In this study, S-nitroso-N-acetyl-D-penicillamine (SNAP) as an NO donor was loaded on carbon nanotubes (CNTs) to eliminate the initial burst release and to prolong the NO release profile. The morphology, the bandings and amount of SNAP on loaded CNTs were characterized by transmission electron microscopy, Fourier-transform infrared spectroscopy and CHNS elemental analysis. Effects of NO on endothelial cell migration and proliferation, platelet activation and bacterial growth were tested.
Results: A concentration of 5 wt% SNAP was identified and selected as the optimal condition to be coated on the lumen of 3D-printed SDVGs out of a biodegradable polymer.
The in vitro studies revealed 20 days of NO release in the physiological range of 0.5-4 × 10-10 mol.cm-2.min-1 with significant enhancement of endothelial cell migration and proliferation. Moreover, the grafts significantly inhibited platelet adhesion/activation and bacterial growth of both Gram-positive and Gram-negative strains.
Conclusion: Overall, using NO-loaded CNTs as a nanocarrier seems a promising approach to be combined with 3D printing of biodegradable SDVGs to promote in-situ endothelialization and tissue integration in vivo in an approach of personalized medicine. The prolongation of NO release profile and encouragement of endothelial cells to migrate and proliferate, in parallel with bacterial eradication and platelet activation inhibition, is a step forward for further development of a next generation of biodegradable 3D-printed SDVGs with tissue regeneration and in situ re-endothelialization capacities.

Keywords: Nanocarriers, Nitric oxide, small diameter vascular grafts, 3D printing, Controlled release system

O-078

The value of galectin-3 in pediatric acute rheumatic fever

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Background and Aim: Galectin-3 -a unique member of galectins- is associated with acute and chronic inflammation, oxidative stress, and fibrosis and is associated with increased risk of cardiovascular diseases. In this single-center prospective study, we aimed to evaluate the diagnostic significance of galectin-3 in children with acute rheumatic fever.
Methods: The study group included 30 patients with acute rheumatic fever and the control group included 26 healthy children. In addition to routine laboratory tests used in the diagnosis of acute rheumatic fever, Galectin-3 was measured at the time of diagnosis and at the end of the treatment from the study group and from the control group.
Results: At the time of diagnosis 12 (40%) cases had mild, 11 had (36.7%) moderate and 7 (23.3%) had severe carditis. After treatment, none of the patients had severe carditis and 23 (76.7%) cases had mild carditis. The mean Galectin-3 level of the study group at the time of diagnosis (7.18 ± 8.31 ng/ml) was significantly lower than the mean post-treatment level (22.29 ± 19 ng/ml) ($p<0.001$). When the study group was further classified according to the severity of carditis, there was a significant difference in mean galectin-3 levels between subgroups defined according to the severity of carditis ($p = 0.06$). Cases with mild carditis had significantly higher mean galectin-3 level compared to cases with moderate and severe carditis. ($p = 0.028$, $p = 0.012$ respectively). Galectin-3 levels were also negatively correlated with the severity of carditis (r:-539, $p = 0.02$). The mean Galectin-3 level of the whole study group was significantly lower than the control group at the time of diagnosis ($p = 0.02$), whereas it was similar after the treatment ($p = 0.714$). Galectin-3 levels were also negatively correlated with the severity of carditis ($r = -539$, $p = 0.02$).
Conclusion: Galectin-3 levels are significantly decreased in children with acute rheumatic fever. Galectin-3 levels are negatively correlated with the severity of the carditis. To the best of our knowledge, this is the first study evaluating the role of galectin-3 in acute rheumatic fever. Multi-center studies with larger sample size would provide more reliable information may highlight the role of galectin-3 in acute rheumatic fever.

Keywords: Galectin-3, acute rheumatic fever, carditis, arthritis.

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The comparison of mean galectin-3 levels of the study group and the control group

		Patient Group (n:30)	Control Group (n:26)	p
Before Treatment	*Mild carditis*	12.6 ± 10.03	20.09 ± 19.59	0.222
	Moderate carditis	4.66 ± 5.05		***0.015***
	Severe carditis	4.41 ± 1.81		***0.021***
	All	7.18 ± 8.31		***0.002***
	p value: ***0.006***			
After Treatment	*Mild carditis*	22.73 ± 15.22		0.718
	Moderate carditis	21.99 ± 13.83		0.808
	Severe carditis	21.99 ± 31.62		0.862
	All	22.29 ± 18.99		0.714
	p value: 0.995			

Student T test, Galectin levels are expressed as mean ± Standard deviation and ng/ml

Surgery and Intensive Care

O-079

A bicentric propensity matched analysis of 158 patients comparing porcine versus bovine stented bioprosthetic valves in pulmonary position

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Background and Aim: Pulmonary valve replacement is the most common operation in adults with congenital heart disease. There is controversy regarding the best bioprosthesis. We compare the performance of stented bioprosthetic valves [the Mosaic (Medtronic™) porcine pericardial against Carpentier Perimount Magna Ease (Edwards™) bovine] in pulmonary position in patients with congenital heart disease.

Methods: Between January 1999 and December 2019, all the pulmonary valve replacements were identified from hospital databases in two congenital heart centres in Spain. Valve performance was evaluated using clinical and echocardiographic criteria. Propensity score matching was used to balance the two treatment groups. Propensity score was calculated for each group using multivariate logistic regression according to the following clinical covariates: Age, body surface area, size of the prosthesis, sex, base diagnosis, year of procedure and the presence of concomitant procedures

Results: 319 patients were retrospectively identified. After statistical adjustment, 79 propensity-matched pairs were available for comparison Freedom from reintervention for the porcine cohort was 98.3%, 96.1% and 91.9% at 3, 5 and 10 years and 100%, 98% and 90.8% for the bovine cohort($p = 0.88$). Freedom from structural valve degeneration for the porcine cohort was 96.9%, 92.8% and 88.7% at 3, 5 and 10 years and 100%, 98% and 79.1% for the bovine cohort ($p = 0.38$). Bovine prosthesis was associated with a reintervention hazard ratio (HR) = 1.12, 95% confidence intervals (CI) (0.24-5.26); $p = 0.89$ and structural valve degeneration HR = 1.69(0.52-5.58); $p = 0.38$. In the first 5 years, there was no difference in outcomes. After 5 years, the recipients of the bovine bioprosthesis were at higher risk for structural valve degeneration [reintervention HR = 2.08(0.27-16.0); $p = 0.49$; structural valve degeneration HR = 6.99(1.23-39.8); $p = 0.03$].

Conclusion: Both bioprosthesis have similar outcomes up to 5 years, afterwards, porcine bioprosthesis seem to have less structural valve degeneration.

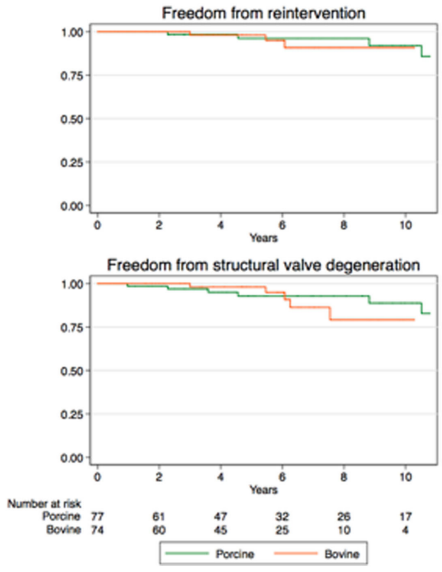
Keywords: Pulmonary valve replacement, bioprosthesis

Graphical abstract:

Key Question: Which bioprosthesis is better in pulmonary position; porcine or bovine?

Key findings: Both bioprosthesis have similar outcomes during the first 5 years, afterwards porcine bioprosthesis degenerate less.

Take home message: Porcine bioprosthesis seem to degenerate less beyond 5 years.



Kaplan Meier curves for the propensity score matched cohort

O-080

Surgical repair of obstructed total anomalous pulmonary venous connection in infants: results and risk factors for mortality in a single centre cohort

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Background and Aim: Advances in modern cardiac imaging has enabled the diagnosis of obstructed total anomalous venous connection (TAPVC) with ease and precision. However, in countries with suboptimal pediatric referral systems and developing infrastructure and especially with lack of routine antenatal diagnosis and pediatric extra corporeal membrane oxygenation (ECMO), the surgical repair continue to be challenging. Aim of this study was to evaluate the perioperative and postoperative surgical risk factors and outcomes among infants operated in a single tertiary center from East Indian population.

Methods: Data was collected prospectively for 39 infants with TAPVCs who were operated at Rabindranath Tagore International Institute of Cardiac Sciences, Kolkata, India between

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January 2015 to March 2018. All patients underwent 2D echocardiography and 6(16%) had a pre-operative computed tomography (CT) scan for difficult pulmonary venous anatomy. Statistical analysis was performed by using SPSS 20 software.
Results: 29 (74.63%) were male and 10 (25.37%) female. 21(54%) were between ages 1-3 months, 11(28%) between 3-6 months and 7(18%) were neonates. Supra-cardiac 17 (44%), cardiac 14 (36%), and mixed in 8 (20%) were the various diagnostic subgroups. Significant pulmonary hypertension was present in 36 (92%) cases. Most common associated cardiac defect excluding an atrial septal defect (ASD), were patent ductus arteriosus (PDA) 8 (20.5%), left superior vena cava (LSVC) 1(2.5%), double outlet right ventricle (DORV) 1(2.5%). Overall early mortality was 18% with 71% long term event free survival. 3% early and 6% late pulmonary venous obstructive disease was observed. Younger age, lower weight and cardiac type of obstructed TAPVC were preoperative risk factors whereas post operatively, reintubation, sepsis, acute kidney injury, delayed sternal closure and pulmonary artery hypertensive crisis were statistically significant risk factors for mortality. The mean clinical follow up was for 896 days and echocardiographic follow up was for 592 days. 87% patients remained in NYHA I/II postoperatively.
Conclusion: Although faced with many logistical challenges in population where antenatal diagnosis is rarely available, reasonable outcomes of surgical repair is feasible amongst infants with obstructive TAPVC. However, younger age, lower body weight and pulmonary hypertensive crisis remain significant predictors of mortality and residual pulmonary venous disease continue to contribute towards repeat interventions.

Keywords: TAPVC, Total Anomalous Pulmonary Venous Connection, Obstructed TAPVC, TAPVC in Infants

Patient Demographics and Perioperative data

	Supracardiac (n=17)	Cardiac (n=14)	Mixed (n=8)	Total (n=39)
Male	13	11	5	29
Female	4	3	3	10
Median Age (days)	78	77	53	
No of Survivors	15	9	8	32
No of Death	2	5	0	7
CPB Time (mins)	130	139	88	127
Range	(60-1490)	(113-183)	(60-120)	(60-1490)
AxC Time (mins)	84	115	57	86
Range	(36-135)	(57-137)	(31-118)	(31-137)
PAH				
Mild	1	1	1	3
Moderate	5	2	1	8
severe	11	11	6	28
Weight (kg)	4 (2.4-6.5)	4(2.5-6.5)	4.5(3.5-6)	4.2(2.4-6.5)
Age				
<30 days	2	1	4	7
31-90 days	9	6	6	21
>90 days	6	1	4	11
Pre Op Ventilation	8	6	6	20
--Death in pre op ventilation	2	4	-	6
Other CHD				
PDA	5	3 (2)	-	8(2)
DORV	0	1	-	1
VSD				
PS				
LSVC	1(1)	0	-	1(1)

CPB- Cardio Pulmonary Bypass , AxC – Aortic Cross Clamp time, PAH – pulmonary arterial hypertension , CHD- Congenital Heart Defects, PDA – Patent Ductus Arteriosus , DORV- Double Outlet Right Ventricle, PS – Pulmonary Stenosis, LSVC-Left Superior Vena cava . Values in Bracket in RED indicate Mortality

The image contains the patient demographics as well as Cardiopulmonary Bypass data of each subgroup of obstructed TAPVCs

Other

O-081
Aortic root dilatation after ross operation

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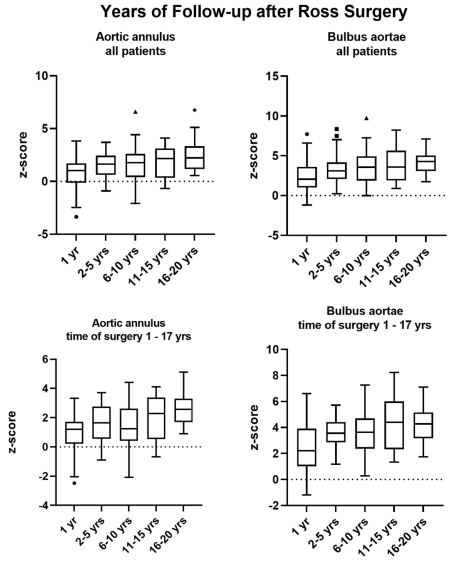
Background and Aim: The Ross procedure is a recognized cardiac surgery to treat left ventricular outflow tract diseases. A major disadvantage of Ross surgery is the progressive dilatation of the neo-aortic root, which is one of the main causes of reoperation, both in children and adults.
This study aims to provide new insights into the progression of neo-aortic root dilation and to assess the natural history after Ross surgery.
Methods: 73 patients (10 infants with a mean age of 1.9 months ± 1.3 months, 39 children mean age 8.6 ± 4.1 years, 24 adults mean age 34.3 ± 10.0 years at the time of the Ross surgery), who underwent Ross procedure and had regular follow-up visits, were included retrospectively by measuring the diameters at the level of the aortic annulus and aortic bulb by echocardiography and cardiovascular magnetic resonance over an observation period up to 20 years after Ross surgery. The increase of the aortic diameters in the long-term course was evaluated. Z-scores of the aortic root were used for comparison.
Results: Aortic annulus´ and aortic bulb´s diameter increased significantly only in the first five years after Ross surgery, $p = 0.0164$ and $p = 0.0006$ respectively, in the entire study group.
Subgroup analysis showed that this increase of aortic annulus diameter ($p = 0.0155$) and of aortic bulb (($p = 0.0022$) was seen only in children (age 1 to 17 years at the time of surgery)
No significant aortic annulus and bulb growth difference was seen in patients who received Ross surgery in infancy and in adulthood.
Conclusion: Early surgery before 1 year of age, or beyond 18 years of age preserved z-scores of the aortic annulus and aortic bulb diameter and could prevent excessive growth of the aortic root and thus reoperation and further complications.

Keywords: Ross operation, Aorta, Bulbus, Anulus, Dilatation

Follow up



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Surgery and Intensive Care

O-082

Effect of platelet transfusions on extracorporeal life support oxygenators' function

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Background and Aim: Bleeding is a common complication of extracorporeal membrane oxygenation (ECMO), leading to increased mortality. Since one of its main complications is bleeding, platelet transfusions are frequently prescribed for children on ECMO. However, there is currently very little information on the effect of platelet transfusions on the function of the ECMO oxygenator. Our objective was to describe the effect of platelet transfusions on oxygenator function.
Methods: In this retrospective study, we included all children (< 18 years) who received ECMO support in our pediatric intensive care unit (PICU) between January 2017 and December 2019. Oxygenator function, measured before and after platelet transfusion, was assessed by post-oxygenator PECMOO2 and the gradient in pre- post-oxygenator pressures (Delta Pressure).
Results: Over 3 years, we analyzed 235 platelet transfusions from 55 children who received ECMO support. Thirty-two (80%) of children were on veno-arterial ECMO and majority of them were peripherally cannulated. When looking at all transfusions, the post-transfusion change in delta-pressure was 0.1mmHg ($p = 0.69$) and post-membrane PECMOO2 was 6 mmHg ($p = 0.49$). However, in the subgroup with the lowest quartile of pre-transfusion oxygenator function, the post-transfusion change in delta-pressure was -5.2 ± 2.7 mmHg ($p<0.001$) and the post-transfusion change in PECMOO2 was -118 ± 49 ($p<0.001$, Figure). The area under the ROC curve for the pre-transfusion delta-pressure and PECMOO2 to predict a worsening of the oxygenator function were 0.72 (95%CI 0.63-0.81) and 0.71 (95%CI 0.64-0.78) respectively. Using regression models, pre-transfusion delta-pressure and PECMOO2 were the only independent factors associated with oxygenator function worsening ($p<0.001$).
Conclusion: Our study suggests that overall, platelet transfusions do not seem to impact the ECMO oxygenator's function. However, in the subgroup of patients with the lowest pre-transfusion oxygenator function, platelet transfusions were independently associated with a worsening function. Future studies should investigate if this warrants adjustments of the anticoagulation strategy around the platelet transfusion, especially among patients with lower oxygenator function.

Keywords: Platelet Transfusion, Thrombosis, Children, Extracorporeal Membrane Oxygenation, Hemostasis

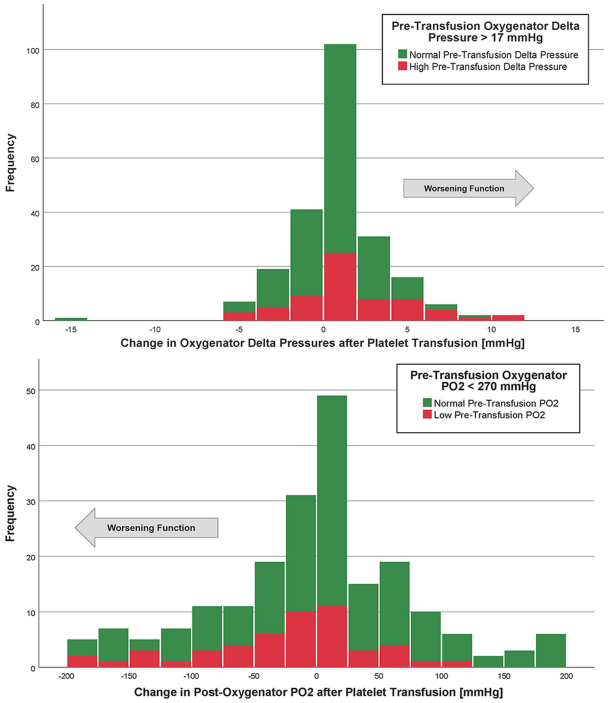

Figure 1 Histogram of the change in oxygenator delta-pressures (top) and post-oxygenator PO2 (bottom) after platelet transfusion. Patients with the lowest pre-transfusion oxygenator function are in red.

O-083

Renal and cardiac effects of remote ischemic preconditioning in children undergoing cardiopulmonary bypass surgery

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Background and Aim: Children undergoing cardiac surgery are at risk for AKI and cardiac dysfunction affecting outcome. Improved surgical techniques and post-operative care allow better outcomes in complex repairs and at-risk patients. Opportunity exists in further protecting multiple end organ function with remote ischemic preconditioning (RIPC) given promising but mixed data in adults and children. Hypothesis: A non-invasive technique of inducing reversible lower extremity ischemia with inflation of pressure cuff lessens renal and myocardial injury.

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Methods: A single center randomized, placebo controlled, double blinded trial of RIPC in children undergoing cardiac surgery of RACHS-I category 2 or greater. RIPC was performed in the operating room after anesthesia and before sternotomy. Pre-specified end points are change in creatinine, eGFR, development of AKI, B-type natriuretic peptide and Troponin I at 6, 12, 241 48, 72 hours post separation from bypass. Secondary end points included select clinical outcomes.
Results: There were 45 in the RIPC and 39 patients in the control group: age was 3.5 and 3.8 years, respectively; 57 patients below 1 year of age; 35 patients below 1 month of age. There was no difference between groups in creatinine, cystatin C, eGFR at each time points. There was a trend for a larger rate of decrease, especially for cystatin C ($p = 0.042$) in the RIPC group but the magnitude was small. AKI was observed in 21 (54%) of control and 16 (36%) of RIPC group ($p = 0.094$). Adjusting for baseline creatinine, the odds ratio for AKI in in RIPC compared to control group was 0.31 ($p = 0.037$). Adjusted for clinical characteristics, the odds ratio was 0.34 ($p = 0\ 056$). Peak troponin occurred at 6 hrs. Compared to control, the RIPC group had a lower troponin at 6 hrs ($p = 0.140$). However, no difference in other analyses of troponin and B*type natriuretic peptides between the groups. Length of stay, all-cause mortality, systolic function by echocardiography and composite clinical end points of advanced renal and heart failure were not different between groups.
Conclusion: There is suggestion of RIPC delivering renal protection in an at-risk pediatric population. Additional larger, higher risk population studies will be required to fully determine its efficacy.

Keywords: Children, congenital heart defect, heart surgery, ischemic preconditioning, renal injury, myocardial protection

O-084
Necrotizing enterocolitis and congenital heart disease

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Background and Aim: Congenital heart disease represents a risk factor for necrotizing enterocolitis
The objectives of the study were to describe and compare the population of infants with congenital heart disease and digestive symptom complicated or not by necrotizing enterocolitis, and assess the risk factors.
Methods: Between January 2016 and December 2020, retrospective analysis of data in infants with congenital heart disease under 3 months of age diagnosed with digestive symptom complicated or not by a necrotizing enterocolitis. The population was divided into: "NEC group" (infants with necrotizing enterocolitis) and "No-NEC group" (infants with an isolated digestive symptom, but no necrotizing enterocolitis)
Results: Overall 67 patients were included in the study: 36 in the NEC group and 31 in No-NEC. The prevalence of enterocolitis during the study period was 4%. Underlying congenital heart disease included coarctation of the aorta, pulmonary atresia and transposition of the great arteries. Fourty-five patients (67%) were administered prostaglandins infusion, 49 (67%) had an umbilical venous catheter, while 67,2% underwent cardiac surgery, 38,8% percutaneous interventional catheterization and 11,9% had no intervention. Digestive events occurred more often within the pre-interventional period ($p < 0.05$) but necrotizing enterocolitis was not more frequent than benign digestive symptoms, although all the infants were fed. Patients were older and cardiac intervention was performed later in the NEC group than No-NEC group ($p < 0.05$). Mother's own milk was less frequently used in NEC patients than No-NEC (40.7% versus 60%, ns). The incidence of hypotroph newborns, high-calorie milk formula and use of antibiotics was greater in NEC than No-NEC group ($p < 0.05$). There was no significant difference in gestational age and transfusion between groups. Twenty-eight cases (48%) occurred in early course after surgical or percutaneous procedure: necrotizing enterocolitis tended to be more frequent in cases (43.3%) who underwent "palliative" surgery or prolonged discontinuation of prostaglandin than in those who had complete surgical repair (61.5% vs 47%, ns).
Conclusion: Cardiac procedure should not be delayed, in order to prevent preoperative enterocolitis. Feeding does not increase the incidence of enterocolitis infants and the administration of mother's own milk or donor human milk should be promoted in this population.

Keywords: necrotizing enterocolitis, neonates, congenital heart disease, risk factors

Arrhythmia/Electrophysiology

O-085
Typical AVNRT - underestimated arrhythmia after atrial redirection surgery for d-transposition of the great arteries? 20 years of catheter ablation

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Background and Aim: Atrial tachycardia represent the most dominant arrhythmic complication during long-term follow-up after Mustard- and Senning-type (M/S) atrial redirection surgery for dextro transposition of the great arteries (dTGA). In contrast, reports of typical AVNRT in this context are scarce. With this analysis we aimed to ascertain the prevalence of typical AVNRT in our large single center cohort.
Methods: All pts with dTGA after M/S repair who underwent an electrophysiologic study and catheter ablation for any tachyarrhythmia at our tertiary referral center between 2001 and 2021 were included retrospectively.
Results: 138 catheter ablations were performed in 92 pts (female 33 (35.8 %), mean age 33.6 [13.3 - 53.3 yrs]). Atrial macro reentrant tachycardia was found in 77/92, non-automatic focal atrial tachycardia in 8/92, atrial fibrillation in 2/92, focal atrial automaticity in 2/92, and ventricular tachycardia in 1/92.
Fourteen pts had typical (slow/fast) AVNRT (12.6 %, mean age 29.5 yrs [18.6 - 45.4 yrs]; female 8; M/S-operation 8/6). Slow pathway modification/ablation was targeted from the pulmonary venous atrial (PVA) aspect of the interatrial septum in 10 cases and from both PVA and systemic venous atrial aspects in the remaining 4 cases. The PVA was accessed retrogradely via transaortic route in 12 and via trans-baffle approach in 2 cases. Ablation was successful in 12 of 14 cases (86%) according to standard endpoints established for slow pathway modification/ablation in normal hearts. Three recurrences of the targeted AVNRT were observed after 2 to 12 weeks. One major complication (femoral artery aneurysm) occurred. Neither procedure related AV blocks emerged nor pacemaker implantation was necessary.
Conclusion: In contrast to published series our analysis of two decades of catheter ablation therapy in dTGA after M/S repair revealed an unexpectedly high prevalence of typical AVNRT. Catheter ablation has shown to be feasible with high success

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and extremely low complication rates despite the demanding post-operative anatomy and subsequent challenges to access relevant ablation sites. Keeping in mind the preoperatively normal atrial anatomy in dTGA we speculate that baffle-surgery, involving inferior regions of the triangle of Koch, may add relevant prerequisites for the development of AVNRT and thus could explain the high incidence in our cohort.

Keywords: Typical AVNRT, d-transposition of the great arteries, d-TGA, Mustard, Senning, atrial switch

O-086

Action potential shape in right ventricular myocardium of patients with congenital heart defects is related to fibrosis and disease severity

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Background and Aim: While for many years surgical scars introduced during repair operation were assumed to be responsible for ventricular arrhythmias in tetralogy of Fallot (TOF) patients, more recent data point towards pro-arrhythmic perturbations originating from surgically unaffected right ventricular (RV) myocardium. The aim of this study was to investigate action potential (AP) remodelling, arrhythmia susceptibility, and possible links of electrophysiological alterations to increased collagen deposition, so-called fibrosis, in repaired and unrepaired TOF patients, compared to patients with a less severe congenital heart defect, the atrial septal defect (ASD).

Methods: RV outflow tract (RVOT) samples from 22 TOF patients and 3 ASD patients were collected during operative repair or re-operation. The study was approved by the ethics committee of the Albert-Ludwigs University Freiburg, and informed consent was given by all patients. Intracellular AP were recorded from the live tissue samples at physiological temperature, and arrhythmias provoked by perfusion with solutions containing reduced potassium and barium chloride, or isoprenaline. Subsequently, collagen content (fibrosis) was quantified in fixed tissue based on Sirius red staining.

Results: Arrhythmias, AP duration (APD) alternans and/or impaired APD restitution manifested frequently in TOF tissue, while they were rare or absent in ASD samples. Longer APD and/or larger AP were positively associated with increased fibrosis and with higher disease severity, i.e. patients requiring re-operation, TOF patients, prolonged QRS duration, proBNP elevation, pre-operative ß-blocker treatment, normal and severely elevated (but not moderate) echocardiographic RV to main pulmonary artery (PA) pressure gradient, and occurrence of tissue arrhythmias. In contrast, shorter and smaller AP with slower upstroke were positively related to pre-operative cyanosis and moderate RV to PA gradient. Higher extent of fibrosis was in itself associated with repaired patients and low RV to PA gradient, but not with cyanosis or tissue arrhythmias.

Conclusion: In conclusion, TOF patients exhibited arrhythmic activity in RVOT tissue already at a very young age and before manifestation of clinical ventricular arrhythmias. Patients with severe acyanotic disease and arrhythmic tissue showed longer and larger AP, associated with increased fibrosis. The combination of electrophysiological and structural characteristics may present a potential diagnostic target to identify patients threatened by a more severe clinical course.

Keywords: tetralogy of Fallot, congenital heart defects, action potential, electrophysiology, fibrosis

O-087

Coronary sinus ablation in pediatric patients with supraventricular arrhythmias; eight years single-center experience

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Background and Aim: Intracoronary sinus ablations have been performed for various epicardial arrhythmical substrates. This study reveals our experience with supraventricular tachyarrhythmia (SVT) ablations into coronary sinus.

Methods: This retrospective study includes all patients who underwent SVT ablation into coronary sinus from October 2013 to October 2021 in a single center. Clinical presentation, type of arrhythmia causing tachycardia, ablation procedure and outcomes were recorded.

Results: A total of 27 patients were included in the study. The total number of patients who underwent ablation in 8 years at the study center was 1450. The median age and weight of the patients were 11 years (range 4.3 to 18 years) and 43.5 kg (range16 to 110 kg). Nineteen of the cases (70,4%) were diagnosed with Wolff–Parkinson–White Syndrome (WPW) (9/19 cases had supraventricular tachycardia (SVT), 8/19 were asymptomatic WPW, 2/19 cases presented with the rapid transition of atrial fibrillation resulting with syncope). Four (14.8%) cases of SVT were diagnosed with concealed accessory pathway, 2 (7.4%) cases with focal atrial tachycardia (FAT), 2 (7.4%) cases with permanent junctional reciprocating tachycardia (PJRT). Ten (37%) patients had a previous history of unsuccessful ablation in other centers. A negative delta wave was noteworthy, especially in lead II in 11/19 (58%) cases, and coronary sinus diverticulum was detected in these cases. Among those with manifest AP (19 cases), 15 (79%) APs were risky. All those APs were adenosine unresponsive. Fluoroscopy was used in 25/27 (92,5%) cases. Median fluoro time was 9,45 minute (min 1 max 34,5). Radiofrequency catheter (RF) ablation was performed in 25/27 (92,5%) cases, and 16/25 (64%) of them were irrigated RF catheters. Cryoablation was established in 4/27(14,8 %) cases, and 2 of them were subsequently intervened with RF catheter ablation. The median procedure time was 186 min (min 105 max 290 min), and the procedure was successful in all (27/27, 100%). Complication or recurrence was not observed during follow-up.

Conclusion: Catheter ablation of supraventricular tachyarrhythmias can be accomplished effectively and safely within the coronary sinus when endocardial approaches are unsuccessful. CS diverticula should be suspected in patients with manifest posteroseptal accessory pathways with a previously failed ablation and typical electrocardiographic signs.

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Keywords: Coronary sinus ablation, supraventricular arrhythmias, radiofrequency catheter ablation, cryoablation

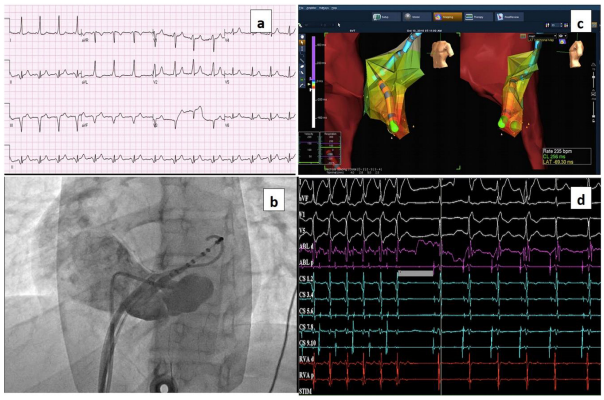

Figure 1 a: ECG of the patient with WPW b: diverticula of same patient c: anatomic mapping of this patient d: successful ablation of this patient

O-088

The effect of electro-anatomical mapping on the success rate and fluoroscopy time in supra-ventricular tachycardia ablation in children

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Background and Aim: Supra-ventricular tachycardia (SVT) is the most common arrhythmia in children, for which catheter ablation of the electro-anatomical substrate is standard of care. Electro-anatomical mapping (EAM) systems have been introduced to help guide the catheter position during ablations and therefore reduce the use of fluoroscopy time.

The aim of this single center study was to evaluate the effect of electro-anatomical mapping on success rate and fluoroscopy time in ablation of SVT in children.

Methods: Patients referred from different centers in the Netherlands and who received a first ablation for SVT in the Leiden University Medical Center between 2014 and 2020, were included in this retrospective cohort study. They were divided in two groups: group 1, procedures in patients who received an ablation between 2014 and 2016 with fluoroscopy; group 2, procedures in patients who received an ablation between 2018 and 2020 using EAM. Procedures in which only an electrophysiological study was performed were excluded.

Results: In total, outcomes of ablation of 371 electro-anatomical substrates were analysed. Mean age (14.0 vs 14.4 years) and mean weight (52 vs 55 kg) did not differ significantly between the two groups. Acute success rate in group 1 (n = 178) was 95.9% compared to 94.5% in group 2 (n = 193) (p = 0.539), with a recurrence rate of 6.1% in group 1 and 5.1% in group 2 (p = 0.731) after a 12-months follow up. Redo-ablations were performed in 12 cases in group 1 and 5 cases in group 2, with a success rate of 83.3% vs 100%, making the overall success rate after 12-months 95.9% in group 1 and 93.1% in group 2 (p = 0.270). Subgroup analysis according to type of SVT showed no significant difference between the groups. Fluoroscopy decreased significantly from 16.00 ± 18.00 minutes (median ± interquartile range) to 2.00 ± 4.00 minutes (p = 0.000), while mean procedure time and range remained stable or even decreased slightly. In group 1, 4 complications occurred (2.0%) and in group 2 zero complications occurred.

Conclusion: The results of this single center study demonstrate that ablations of SVT in children are a highly effective and safe treatment and the use of EAM can significantly reduce fluoroscopy time.

Keywords: Supra-ventricular tachycardia, ablation, electro-anatomical mapping, success rate, fluoroscopy time.

Results of ablation procedures

	Group 1	Group 2	Total	p-value
Acute success first ablations	163/170 (95.9%)	171/181 (94.5%)	334/351 (95.2%)	.539
AVNRT	54/56 (96.4%)	63/63 (100.0%)	117/119 (98.3%)	.219
AVRT	98/102 (96.1%)	93/102 (91.2%)	191/204 (93.6%)	.152
AT	8/9 (88.9%)	12/13 (92.3%)	20/22 (90.9%)	1.000
AFL	3/3 (100.0%)	3/3 (100.0%)	6/6 (100.0%)	
Recurrences within 12-months	10/163 (6.1%)	7/136 (5.1%)*	17/299 (5.7%)	.713
AVNRT	3/54 (5.6%)	5/47 (10.6%)*	8/101 (7.9%)	.467
AVRT	6/98 (6.1%)	1/78 (1.3%)*	7/176 (4.0%)	.135
AT	1/8 (12.5%)	1/9 (11.1%)*	2/17 (11.8%)	1.000
AFL	0/3 (0.0%)	0/2 (0.0%)	0/5 (0.0%)	
Success rate redo-ablations	10/12 (83.3%)	5/5 (100%)*	15/17 (88.2%)	
AVNRT	4/4	2/2*	6/6	
AVRT	5/6	2/2*	7/8	
AT	1/2	1/1*	2/3	
AFL	-	-	-	
Overall success	163/170 (95.9%)	134/144 (93.1%)*	297/314 (94.6%)	.270
AVNRT	55/56 (98.2%)	44/47 (93.6%)*	99/103 (96.1%)	.329
AVRT	97/102 (95.1%)	79/85 (92.9%)*	176/187 (94.1%)	.533
AT	8/9 (88.9%)	9/10 (90.0%)*	17/19 (89.5%)	1.000
AFL	3/3 (100.0%)	2/2 (100.0%)*	5/5 (100.0%)	
Procedure time (min) Median ± interquartile range	184.00 ± 80.00	180.00 ± 64.00	183.00 ± 70.00	.675
Fluoroscopy time (min) Median ± interquartile range	16.00 ± 18.00	2.00 ± 4.00	7.00 ± 15.00	.000
Major complications	4 (2.0%)	0 (0.0%)	4 (2.0%)	

* 12-month follow-up of procedures performed in last months of 2020 are not complete yet

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O-089

Cheek smear analysis as a prognostic tool in children with arrhythmogenic cardiomyopathy

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Background and Aim: Arrhythmogenic cardiomyopathy (ACM) is a major cause of sudden cardiac death (SCD) worldwide. Yet, its diagnosis and risk stratification remain highly challenging especially in children who do not show overt clinical disease. We have shown that ACM development correlates with myocardial cell-cell junction protein shifts in adult patients. This finding may have diagnostic and prognostic implications but its utility is limited by the need for a heart sample. More recently, we reported that these myocardial processes are mirrored by protein re-localization in the buccal epithelium. Herein we aimed to establish when protein distribution abnormalities first occur and whether they correlate with progression of clinically apparent disease.
Methods: To this end we analysed serial buccal mucosa samples of children and adolescents bearing ACM-causing mutations in desmosomal genes (n = 16). Smears obtained from age-matched healthy children with no clinical manifestations or family history of heart disease served as normal controls (n = 13). Samples were immunostained for plakoglobin, desmoplakin, plakophilin-1 (an isoform of plakophilin-2 expressed in the upper epithelia) and connexin43 (Cx43; the major ventricular gap junction protein) and analysed by confocal microscopy. All participants were swabbed at least twice with an average interval of 12-18 months between sampling.
Results: We found that junctional protein re-localization does not correlate with the presence of an ACM-causing variant but instead it correlates with the onset of disease. No changes are seen in buccal smears unless and until there is clinical evidence of disease. In addition, progressive shifts in the distribution of key proteins correlate with worsening of the disease phenotype. Finally, we observed restoration of junctional signal for Cx43 in a patient with a favourable response to anti-arrhythmic therapy.
Conclusion: Due to ethical concerns about obtaining heart biopsies from children with no apparent disease, it has not been possible to correlate molecular changes with the onset/progression of clinical disease. Using cheek smears as surrogate for the myocardium provides an opportunity to investigate these questions. Our results indicate that analysis of buccal cells, may be a totally safe and inexpensive tool for risk stratification and potentially monitoring response to treatment in children bearing ACM variants.

Keywords: Arrhythmogenic cardiomyopathy, sudden cardiac death, prognostic test, cheek smear, desmosome, Cx43

O-090

Electrocardiographic and electrophysiological characteristics of fasciculoventricular fibers in children: single-center 8-years experience

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Background and Aim: Fasciculoventricular fiber (FVF) is a rare cause of ventricular preexcitation. Although adenosine response and some electrocardiographic features are important in differentiate from Wolf Parkinson White (WPW), a clear distinction may not always be possible without an electrophysiological study (EPS). In this study, our aim is to evaluate the clinical and electrophysiological features of our pediatric patients with FVFs.
Methods: In this study, FVF was detected in 27 (4.7%) of 565 patients who underwent electrophysiological study due to ventricular preexcitation between 2013-2021. The demographic characteristics of the patients were obtained from the file records. In addition, delta wave amplitudes were measured from the surface ECG before and after ablation in patients with additional accessory pathways. Post-procedure values were included in the FVF data.
Results: The mean age of the patients was 11.47 ± 4.25 years. 70.4% of the reasons for admission were symptoms such as palpitations and syncope. Two patients had hypertrophic cardiomyopathy and one patient had ccTGA. In the EPS, additional manifest WPW was found in 9 (33%) patients (3 patients with high risk), focal atrial tachycardia in one patient, and atrioventricular nodal reentrant tachycardia in another patient (totally 11/27). While the delta wave amplitude in surface electrocardiography was 2.56 ± 1.38(1-5.5)mm in 9 patients with additional accessory pathway, it was 1.64 ± 0.67(0.5-3) mm in the FVF group. Delta amplitude >3.5mm was not detected in any patient with isolated FVF. Interestingly, delta amplitude was <3.5mm in 7 of 9 patients in whom additional accessory pathways were identified and ablated. Nineteen of the patients (59.3%) were adenosine-responsive (18 isolated FVF, 1 manifest AP+FVR responsive to adenosine).
Conclusion: Although the FVF are not the cause of tachyarrhythmia, the accessory pathway and other tachyarrhythmia substrate frequencies accompanying these cases are quite high (approximately 40%) in EPS. The delta wave characteristics of ablated patients are very similar to FVF patients. Although the adenosine response is important, is not definitively distinctive. Therefore, performing EPS in patients (especially symptomatic) with suspected FVF based on surface ECG features seems to be important for the detection of additional tachyarrhythmias and risky accessory pathways.

Other

O-091

Short to long-term outcome of patients implanted with a telemetric adjustable pulmonary artery banding device flowatch-PAB® in switzerland

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Background and Aim: Pulmonary artery banding (PAB) was developed treat children having cardiopathies with excessive pulmonary blood flow by inducing an artificial stenosis of the pulmonary artery (PA) with the aim to lower pulmonary artery pressure and protect the pulmonary vascular bed. Difficulties to optimally adjust the tightening during surgery may lead to multiple

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additional procedures, increasing the risk of damage to the PA. An innovative technology: a battery free, wireless telemetric PAB, called FloWatch-PAB, was developed to overcome these cand limitations.
Methods: We investigated the short and long-term outcomes of 69 patients (pts) implanted with the FloWatch-PAB in Switzerland between 2002 and 2016. Data were collected through a survey sent to three hospitals and included patient demographics, diagnostic, implantation data (patient data, mode of implantation, simultaneous cardiac surgery, complications), post implantation data (complications, hospital stay, intensive care unit stay, FloWatch-PAB adjustment(s)), patient explantation information (cardiac status, concomitant corrective surgery) reconstruction of pulmonary artery and long-term follow up.
Results: 83% of patient had no surgical complications and easy FloWatch implantation. For the remaining, the difficulties were mainly caused by inadequate PA condition, such as pulmonary dilatation. 89.6% of patients did not need any additional surgery. An average of 4 +/- 2 telemetric adjustments (tightening or opening) per FloWatch-PAB was observed, highlighting all the unnecessary surgeries avoided with the conventional PAB. 80% of patients did not need PA reconstruction. Finally, over 5-years follow up information post-implantation were available for 52 patients. 42 patients are alive and 10 are deceased. Among the 42 patients, 10 had late PA complications either to the main trunk or the peripheral pulmonary branches (such as stenosis, hypoplasia or valvular insufficiency) and only 2 required a corrective angioplasty. 10 patients died, all secondary to the severity of heart disease. There was no direct FloWatch-PAB related death and an unclear link of the FloWatch with the late PA complications.
Conclusion: The FloWatch-PAB was a unique device which had the main advantages of being able to adjust band tightening and opening avoiding multiple surgeries and seem to minimize PA damage avoiding surgical reconstruction and long-term PA complications.

Keywords: Adjustable pulmonary artery banding

Surgery and Intensive Care

O-092
Hospital survival has improved in neonates with congenital heart disease supported on prolonged ECMO – an elso registry based cohort study

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Background and Aim: Increased utilization and longer duration of extra corporeal membrane oxygenation (ECMO) support has been reported in neonates with congenital heart disease (CHD). However, little is known about the risk factors for survival with prolonged ECMO use (> 7 days). We aim to examine risk factors associated with hospital survival in this population from the Extracorporeal Life Support Organization (ELSO) registry.
Methods: Our cohort study included neonates (≤ 28 days age) with CHD supported on veno-arterial ECMO > 7 days (only first run included and congenital diaphragmatic hernia was excluded) at 111 centers in the United States from January 2011- December 2020. Logistic regression was used to determine the odds ratio (OR) of risk factors and 95% confidence interval (CI) to estimate associations with death on ECMO or survival to hospital discharge.
Results: Out of 2155 total ECMO runs, 948 neonates received prolonged ECMO support [Gestational age (mean ±SD) 37.9 weeks; birth weight 3.1 ±0.6 kg; median (interquartile range) ECMO duration 10.7 (8.4 – 14.5) days]. ECMO survival was 51.6 % (489/948) and survival to hospital discharge was 23.9% (226/948) significantly lower than the overall cohort (ECMO survival 69.5% and survival to hospital discharge 46.4% respectively ($p<0.0001$ for both). Body weight at ECMO (OR: 0.74; 95% CI: 0.57-0.96 per kg), female sex (1.38; 1.04-1.84), and pre-ECMO mean arterial pressure (0.93; 0.87-1.00 per 5 mm Hg) were significantly associated with death on ECMO. Body weight at ECMO (0.59; 0.44-0.78 per kg), gestational age (0.89; 0.79-1.00 per week), risk-adjusted congenital heart surgery (RACHS-1) score (1.22; CI: 1.04-1.45), and pump flow at 24 hours (1.11; 1.04-1.18 per 10 ml/kg/minute) were significantly associated with death prior to hospital discharge. In addition, pre-ECMO mechanical ventilation duration, time to extubation after ECMO decannulation, ECMO complications and length of stay were inversely associated with hospital survival.
Conclusion: Neonates receiving ECMO support beyond seven days have reduced survival off ECMO and survival to discharge when compared to the overall neonatal CHD ECMO cohort. Increased gestational age, birth weight and lower RACHS-1 score are associated with better outcomes. Further elucidation of factors associated with reduced survival to discharge in ECMO survivors is needed.

Keywords: Congenital heart disease; Extra corporeal membrane oxygenation; Neonate; Survival

O-093
Personalised external aortic root support (PEARS) applied for dilatation aortopathies in paediatric patients

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Background and Aim: PEARS technique consists on a polyester mesh created using a 3D printer and the patient's aortic root measurements obtained from a CT scan, which stabilises the aorta dimensions and prevents its dilatation. Although initially created for adult Marfan's syndrome patients, its use has been extended to other aortopathies and even into paediatric population
Methods: We present our institutional single surgeon results of the application of this technique in patients aged 18 or under, with underlying diagnosis of dilatation aortopathies, including Marfan's syndrome or bicuspid aortic valve related aorta disease. We analise the preoperative and postoperative aortic root maximum dimensions, as well as MRI and echocardiographic demonstration of stability during follow-up. We have also registered the postoperative complications
Results: 12 patients were included in our study, including a pair of twins operated on the same day. Age ranged from 12 to 18 years old. 9 patients had Marfan's syndrome, while 3 had bicuspid aortic valve dilated aortopathy. Preoperative aortic measurements

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(obtained by MRI or CT scan) ranged from 29 to 59 mm. 2 of the patients with Marfan's had concomitant mitral valve repair, and 1 patient had to go unexpectedly on bypass due to recurrent diathermy induced ventricular fibrillation while performing dissection of the right ventricular outflow tract. The rest of the procedures were performed off by-pass. Immediate postoperative aorta measurements, showed an average reduction of 6.5 mm. Two patients had significant improvement in their preoperative aortic regurgitation after the PEARS graft application, due to reduction and remodelling of the sinus of Valsalva and sinotubular junction. Three patients had a short-lived postoperative inflammatory response, and one had a pericardial effusion requiring drainage. All patients have shown stable aortic dimensions and aortic valve function during follow-up.
Conclusion: PEARS is a safe and effective technique than can be used in paediatric population. This technique, apart from stabilising the aortic root dimensions and prevent any further dilatation, can also improve mild degrees of central aortic regurgitation when applied in selected patients.

Keywords: Marfan's syndrome; aorta dilatation; aortic surgery

O-094

Complete atrio-ventricular septal defect: modified 2-patches technique

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Background and Aim: Complete atrio-ventricular septal defect (CAVSD) concerns 3 % of congenital cardiac malformations. Several repair techniques exist. Reoperations are frequent. Most common causes are residual right or left AV valve regurgitation, conduction abnormalities and left AV valve stenosis.
Methods: Between june 2012 and may 2019, 31 patients (14 female) with CAVSD and non restrictive VSD. The operation was performed through a right oblique atriotomy. Middle of the anterior and posterior bridging leaflets were spotted and split. Left cleft was closed (or partially closed). VSD was closed (half teardrop patch). Anterior and posterior leaflets were reinserted on the patch. ASD was closed with autologous pericardium, leaving the coronary sinus on the right side.
Results: Median age was 5 months (IQR 4–6). Median VSD was 8.5 mm (IQR 5–10.25). Rastelli types were A in 21(67.7%), B in 3(9.7%), C in 6(19.4%) patients. Down syndrome was present in 22(71%) cases. There were 4 premature children (12.9%) and 5 Fallot-type (16.1%).
There were only 2 minimal residuals VSD (6.5%). There was no severe AV valve regurgitations with moderate left and right AV regurgitation in 8 (25.8%) and 5 (16.1%) patients respectively. Transitory AV block happened in 3 patients (9.7%), but none required a pacemaker. Two patients required a reoperation (6.5%) for a subaortic membrane after 2 years and left AV valve repair during a contegra conduit replacement. Mean Left and right AV valve gradient were 2.3 mmHg (IQR 1.5 – 3.0) and 1.4 mmHg (IQR 1.2 – 2.0) respectively. There were no left or right severe stenosis. Moderate left AV valve stenosis (5-10 mmHg) was present in 1 patient (3.2%) and mild stenosis (3-5 mmhg) in 7 (22.6%). Right AV valve stenosis was moderate (3-6 mmHg) in 1(3.2%) patient and mild (2-3 mmHg) in 4 (12.9%) patients.
Conclusion: Short-term results of 2-patches with division of the 2 bridging leaflets showed good correction. Although it implies a slightly longer cross-clamp time, it allows a good optimisation of the left and right new AV valve with minimal residual leak and low gradient. Conduction system is not at increased risk

Keywords: Atrioventricular septal defect; repair; 2 patches; reoperations

O-095

Presence of multiple preoperative risk factors (≥ 3) is associated with poor prognosis in an 11-year national cohort of HLHS patients

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Background and Aim: Several studies have reported mortality risk factors associated with hypoplastic left heart syndrome (HLHS). The most frequently reported preoperative risk factors include small ascending aorta, low birth weight, restrictive atrial septum, presence of a syndrome, tricuspid valve regurgitation, and impaired ventricular function. However, these data are ambiguous and mainly focused on the independent effect of these factors. We examined both the independent and the cumulative effect of preoperative risk factors on one-year mortality in an 11-year national cohort of HLHS patients in Finland.
Methods: We performed a retrospective national 11-year observational study of preoperative risk factors (listed in Figure) for one-year mortality in HLHS-patients (n = 80) born in Finland between January 1, 2004, and December 31, 2014.
Results: Overall, one-year survival was 86.3% (69/80). In a multivariable analysis, total anomalous pulmonary venous drainage (TAPVD, OR 3.017, P = 0.013, 95% CI 1.889-221.153) and aortic atresia (AA, OR = 1.856, P = 0.029, 95% CI 1.204 - 34.003) were significant predictors for one-year mortality. Lower gestational age, restrictive foramen ovale, and extracardiac malformations were not significant independent predictors for one-year mortality in the HLHS cohort.
Survival was analyzed using Cox regression analysis and Kaplan Meier survival analysis for visualization. During the follow- up time of one year, 11 deaths occurred in the 80 patients who underwent the Norwood procedure. Three patients had three or more risk factors. All patients in this category died during the first three months after the Norwood procedure (Figure 1A&B)
Conclusion: HLHS remains a defect with the highest procedural risks for mortality in paediatric cardiac surgery.
From a prognostic point of view, recognition of independent preoperative risk factors for mortality is essential. However, the presence of multiple risk factors (≥ 3) was lethal in infants with HLHS in our 11-year national cohort.

Keywords: Congenital heart disease, hypoplastic left heart syndrome, preoperative risk factors, repair of single ventricle with

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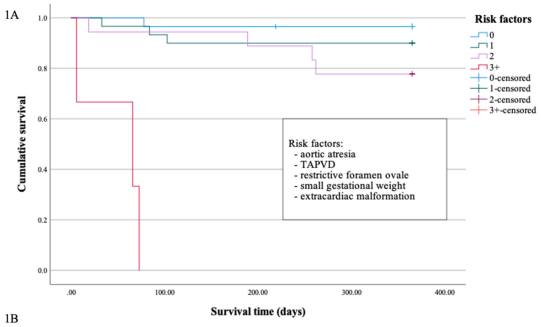
aortic outflow obstruction and aortic arch hypoplasia (hypoplastic left heart syndrome) (eg, norwood procedure), mortality

1A



1B

Risk factors[a]	Number of deaths	Hazard ratio (95% CI) (N=80)
No risk factors	1/29	1
One risk factor	3/30	2.99 (0.311 – 28.72)
Two risk factors	4/18	6.74 (0.75 – 60.29)
Three risk factors	3/3	188.12 (14.23 – 2486.89)

[a]SGA/aortic atresia/TAPVD/restrictive formaen ovale/extracardiac malformation

Figure 1 A&B Survival analysis of multiple preoperative risk factors for mortality in HLHS patients. A. Kaplan Meier curve and B. Cox regression analysis

O-096

Does additional pulmonary anterograde flow at the glenn procedure improve pulmonary artery growth and at what cost?

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Background and Aim: The ultimate objective for patients with a functional single ventricle are well developed pulmonary arteries and low pulmonary vascular resistance (PVR). After the Glenn procedure, the potential for pulmonary artery (PA) growth is limited. To deal with this problem, some institutions leave additional pulmonary anterograde flow (APAF). The aim of this study is to assess the effect of APAF on hemodynamic parameters and PA growth prior to Fontan completion.
Methods: A single institutional retrospective study of patients after the Glenn procedure was conducted for a 14-year period. Patients with missing data and no cardiac catheterization prior to bidirectional Glenn were excluded from the study. The study population (n = 61) was divided in two groups: patients with APAF (Group 1, n = 30) and patients without APAF (Group 2, n = 31). We compared the cardiac catheterization data immediately before the Glenn and Fontan procedure between the two groups. Nakata index (mm2/m2) and pulmonary arteries z-scores, calculated from angiography images, were used to determine PA size. Mann-Whitney U test was used for statistical analysis.
Results: Comparison of cardiac catheterization data between patients with and without additional pulmonary anterograde flow are described in Table 1. Oxygen saturation, mean PA pressure and end-diastolic pressure, PVR, ejection fraction and atrioventricular regurgitation were unaffected by APAF. There was no difference in PA z-scores pre-Glenn between the two groups. Patients with APAF had better left PA dimensions prior to Fontan (z-score 1.57 versus 0.31, p = 0.009), while right PA dimensions didn't differ. The Nakata index was not different between the two groups pre-Glenn and pre-Fontan.
Conclusion: APAF has no adverse effect on hemodynamic parameters before the Fontan procedure. It results in larger left PA dimensions, but doesn't promote PA growth assessed by Nakata index. We may speculate that larger left PA may have positive effect on hemodynamics after Fontan completion, but it needs further evaluation.

Keywords: Glenn procedure, additional pulmonary anterograde flow

Table 1. Comparison of cardiac catheterization data between patients with and without additional pulmonary anterograde flow

	Additional pulmonary anterograde flow (APAF) N = 30	Without additional pulmonary anterograde flow (APAF) N = 31	P
Saturation (%), median (range)	80 (63 - 93)	79 (65 - 90)	0,197
Mean pulmonary artery pressure (mmHg), median (range)	14 (7 - 23)	14 (9 - 17)	0,235
End-diastolic pressure (mmHg), median (range)	11 (4 - 18)	10 (6 - 16)	0,375
Pulmonary vascular ressistance (Wood units), median (range)	1.14 (0.47 - 2.12)	1.34 (0.41 - 3.9)	0,129
Ejection fraction (%), median (range)	60 (47 - 87)	60 (45 - 85)	0,767
More than mild atrioventricular regurgitation (n)	9	9	0,797
Left pulmonary artery pre-Glenn (z-score), median (range)	1.95 (-1.76 - 4.0)	2.39 (-0.11 - 5.22)	0,299
Right pulmonary artery pre-Glenn (z-score), median (range)	1.73 (-1.55 - 4.65)	2.08 (-2.32 - 5.58)	0,681
Left pulmonary artery pre-Fontan (z-score), median (range)	1.43 (-4.7 - 3.45)	0.31 (-3.74 - 2.67)	**0,009**
Right pulmonary artery pre-Fontan (z-score), median (range)	1 (-2.72 - 3.88)	0.87 (-1.94 - 4.62)	0,534
Nakata index pre-Glenn (mm^2/m^2), median (range)	316 (101-617)	351 (133-803)	0,387
Nakata index pre-Fontan (mm^2/m^2), median (range)	281 (70-575)	251 (83-505)	0,109

Imaging/Functional assessment

O-097

Three dimensional echocardiography and global longitudinal strain in the assessment of left ventricular function in children after PIMS-TS

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Background and Aim: Pediatric Inflammatory Multisystem Syndrome Temporally Associated with SARS-CoV-2 (PIMS-TS) is a new disease affecting children, almost always involving cardiovascular system and with potential long-term effects still unknown.
Methods: Prospective study enclosed 80 children aged 1–17 years (mean 8.2 years) diagnosed with PIMS-TS between June 2020 and June 2021 who were controlled 6 weeks and 6 months after the disease. In patients with severe cardiac involvement during acute phase (deteriorated left ventricular ejection fraction (LVEF) <55% and significantly elevated concentration of NT-pro-BNP (>5000 pg/ml) or troponine (>500 ng/ml)) the additional check-up after 3 months was scheduled. In all patients at control points three dimensional echocardiography (3D-ECHO) and average global longitudinal strain (GLS) were used to assess left ventricular function.
Results: In all patients the means of LVEF and average GLS were within normal limits at the time of all check-up points. For the whole group LVEF after 6 weeks was 60.5% (SD: 3.1; 51-69%) and GLS 21.2% (SD: 3.9; 12.4-29.4%). After 6 months LVEF increased to 63% (SD: 2.4; 58-69%) and GLS to 23.6% (SD: 3.2; 17.3-33.3%) – both significantly (p<0.001). In the subgroup of 54 patients with originally mild cardiac involvement LVEF after 6 weeks was 60.7% (SD: 2.6; 57-69%) and GLS 21.8% (SD: 3.4;

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17.3-29.4%). In the subgroup of 26 patients with severe cardiac involvement LVEF after 6 weeks was 59.6% (SD: 3.1; 55-67%) and was not significantly different than in subgroup with mild cardiac involvement ($p = 0.175$) while GLS was significantly lower (19.3%, SD: 3.8; 12.4-24.8%; $p = 0.009$). After 3 months in this group LVEF and GLS did not increase (respectively, 59.9%, 56-67%; $p = 0.794$ and 20.2%, 13.7-26.9%; $p = 0.149$). After 6 months LVEF in this subgroup increased to 62.8% (60-68%) and GLS to 22.6% (17.7-27%) - like in patients with mild cardiac involvement ($p<0.001$).
Conclusion: 3D-ECHO and GLS are highly applicable tools for the assessment of cardiac function in children after PIMS-TS.
Patients with originally severe cardiovascular involvement have lower average GLS after 6 weeks.
6 months after PIMS-TS patients present significant improvement of left ventricular function.
Average GLS seems to be more sensitive test for functional assessment than LVEF.

Keywords: PIMS-TS, three dimensional echocardiography, global longitudinal strain

Other

O-098
Increase in kawasaki disease incidence observed in 30-year population-based study in scandinavia

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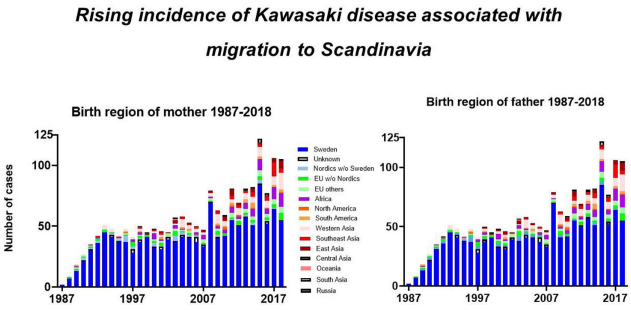
Background and Aim: World-wide, Kawasaki disease (KD) is known to affect predominantly children under the age of 5, mostly boys. An increasing incidence has been reported from select countries, as well as seasonal differences, although with great variation among reports. Sweden has unique population-based health registers which can be linked to population registers via a personal number. In this study we therefore utilized population-based data over a period of more than 30 years to investigate demographics and epidemiology of Kawasaki disease in a Scandinavian country.
Methods: Individuals receiving a diagnosis of Kawasaki disease in Sweden from 1987-2018 (before the occurrence of MIS-C) were identified by ICD9 and ICD10 discharge diagnoses in the Patient register at the National Board of Health and Welfare, and basic demographic information obtained by cross-linking with population registers at Statistics Sweden. Age-stratified population statistics were also retrieved during the corresponding time-period.
Results: A total of 1,785 individuals with a KD diagnosis during the study period were identified, confirming a relatively low incidence in the Scandinavian population. Less than 5% of the cases were born in another country. The majority of cases (78%) occurred before 5 years of age, and there was a male dominance (61%). Sweden has a temperate climate of the northern hemisphere, and analysis of case distribution over the yearly cycle revealed peak incidence during the winter months. Notably, the incidence rose from around 6/100,000 <5-year-olds to 15/100,000 <5-year-olds during the 30-year study period. Two years with prominently higher incidence than prior and following years were observed. A large part of the rise in incidence seems to be associated with immigration and occurred before the occurrence of Multisystem Inflammatory Syndrome in Children related to SARS-CoV-2.
Conclusion: Demographic parameters for Kawasaki disease in Sweden regarding age and sex distribution are similar to previous reports from other countries. Our data from a 30-year study period of population-based observations confirm peak incidence during the cold period, and a rising incidence during recent years, even before the occurrence of MIS-C. Our data also indicate outbursts during two years and immigration-associated patterns in rise in incidence.

Keywords: Kawasaki disease; MIS-C; Multisystem Inflammatory Syndrome in Children, acquired heart diease; epidemiology; SARS-CoV-2

Rising incidence of Kawasaki disease in Sweden before the SARS-CoV-2 pandemic associated with immigration



The rising incidence of KD in Sweden prior to the SARS-CoV-2 is partly explained by immigration to Sweden by the parents of the children who were born in Sweden and who were diagnosed with KD. Other countries have reported a similar rise in incidence. Is it as yet unclear if immigration from regions where KD is reported less has a role to play even in other countries with rising incidence of KD.

O-099
Complete resolution of cardiac sequelae is seen in patients treated for paediatric multisystem inflammatory syndrome (PIMS) by 12 months

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Background and Aim: Paediatric multisystem inflammatory syndrome (PIMS) was a new phenomenon which emerged during the coronavirus pandemic. Mostly affecting children and adolescents, PIMS symptoms were noted 2-4 weeks following the initial covid-19 infection as patients presented with persistent fever, evidence of inflammation and single or multi organ dysfunction. As previously described by our group, over half of patients with PIMS develop significant cardiac involvement including coronary artery dilatation/ aneurysm, impaired cardiac function and pericardial effusion. This study aims to describe the outcomes of patients treated with PIMS over the first 12 months post illness.
Methods: 93 patients have so far presented within the Yorkshire and Humber region of the UK with evidence of PIMS. So far, we have six-month follow up data for 66% of these patients and an ever-increasing proportion of patients reaching 12 months. This study involved a retrospective case notes review to compare the cardiac involvement at presentation; 6 weeks post illness and 12 months.
Results: 52.2% of patients had significant cardiac involvement at presentation. This had decreased to just 14.5% at six weeks and all patients had a normal cardiac echo at 12 months post illness.

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Furthermore, no patients who had a normal echo during their acute admission developed cardiac changes during follow up.
Conclusion: PIMS is associated with a relatively high incidence of significant cardiac involvement. The initial stage of the illness can be a worrying time with cardiac function impairment, valve incompetence and coronary artery changes in patients with no history of congenital heart disease. The unknown prognosis for patients with these changes has provided much concern for patients, their families and clinicians. This study shows that although there may be worrying changes in the initial stages of the illness, there is complete resolution over the first-year post illness. Furthermore, patients with a normal echo during the initial stage of the illness do not develop cardiac involvement at a later stage, indicating that these children can be discharged from cardiology follow up after resolution of the acute illness and followed up by their local paediatric team.

Keywords: Paediatric Multisystem Inflammatory Syndrome PIMS Covid-19

O-100
A comparative study of cardiac involvement of kawasaki disease and multisystem inflammatory syndrome post COVID (MISC) in children

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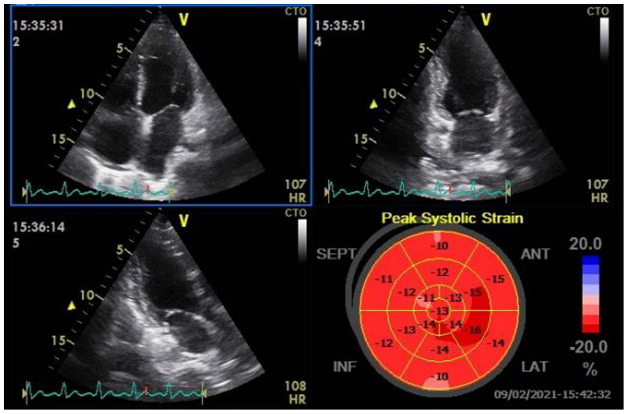
Background and Aim: Kawasaki Disease remains an enigma to the world to this day since first described by Dr. Tomisaku Kawasaki in 1967. In the last half a century there has been widespread global research elaborating the clinical aspects and pathogenesis of this disease entity. Multisystem Inflammatory Syndrome post Covid (MISC) is a relatively new disease which was described in literature in mid 2020. The striking resemblance as well as differences in spectrum of cardiac involvement of both the conditions has been elaborated in this study from a tertiary care centre in Eastern India.
Methods: The study was conducted over a period of 3 years from June 2018 to June 2021. Fiftyone patients with Kawasaki disease (including atypical and incomplete cases) and sixty children diagnosed with MISC were included in the study. Echocardiography details were noted by a single observer. Data regarding the patient particulars, clinical aspects, lab parameters, imaging details and treatment particulars were collected and analysed. Patients were followed up for a minimum period of six months to one year.
Results: In the Kawasaki group(51), infants(20) presented with multiple (and larger) aneurysms. Older children (>5 years) had more of single coronary involvement, (mostly LAD) and also had more atypical presentation(18) associated with infections like Dengue, Staphylococcal infection, Scrub Typhus. There were 4 cases of Kawasaki shock syndrome, all below 5 years. In the MISC group (60), there was also multiple coronary involvement in infants (11). But LV dysfunction was more common in older children and adolesecents (20), of whom 18 (90%) presented with severe dysfunction (LVEF<35%). Those with coronary involvement had normal function and those with dysfunction had no coronary involvement. Mild to moderate aneurysmal dilation of coronaries was found in children one to five years of age. No giant aneurysm was found in MISC. Overall, LMCA with LAD was the commonest pattern of involvement in both the conditions.
Conclusion: KD and MISC had similar pattern of coronary involvement, but absence of giant aneurysm and significantly severe dysfunction in older children in MISC indicates a likely different pathogenesis for myocardial involvement in MISC.

Keywords: MISC, coronaries, dysfunction, Kawasaki disease

Reduced GLS in MISC in a fourteen year old boy



O-101
Kawasaki disease in the time of COVID-19 – the international kawasaki disease registry

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Background and Aim: The considerable overlap in case definition and clinical features between patients with COVID-19 associated Multisystem Inflammatory Syndrome in Children (MIS-C) and Kawasaki disease (KD) suggests shared pathogenesis. We sought to compare demographic, clinical presentation, management and outcomes of patients by COVID-19 status.
Methods: The International KD Registry (IKDR) began enrolling patients with clinical features of either acute MIS-C or KD or fever with hyperinflammation beginning in January 2020. The IKDR is unique regarding broad patient selection and includes sites from North, Central and South America, Europe, Asia and the Middle East. Patient groups stratified by COVID-19 status were compared.
Results: As of October 6, 2021, 1330 patients were registered from 31 sites. COVID status was POSITIVE for 59% (confirmed

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household COVID-19 contact and/or positive SARS-CoV-2 PCR or serology), POSSIBLE for 4% (suggestive clinical features but some negative tests or absent exposure), NEGATIVE for 23%, and UNKNOWN (no known exposure and testing not completed) for 14% (TABLE). Most of the UNKNOWN patients were from early in the COVID-19 pandemic before MIS-C was defined and before COVID-19 serologic testing was widely used. POSITIVE and POSSIBLE patients were older, had fewer KD clinical criteria, greater gastrointestinal symptoms, were more likely to present with shock and require ICU admission and inotropic support. POSSIBLE patients had greater days from symptom onset to first immune modulation treatment, with no differences between groups regarding days from admission to first treatment. Most patients in each group received intravenous immune globulin, with POSITIVE and POSSIBLE patients more likely to have received steroids and anakinra. NEGATIVE and UNKNOWN patients had higher maximal coronary artery Z scores, with a trend to having higher categories of aneurysm involvement.

Conclusion: While there was considerable overlap in presentation, management and outcomes between COVID-19 POSITIVE/ POSSIBLE (presumed MIS-C) and COVID NEGATIVE/ UNKNOWN patients (presumed KD), COVID-19 POSITIVE/ POSSIBLE patients had more severe presentations and required more intensive management, although coronary artery outcomes trended to be less severe. Patient recruitment continues, and in-depth comparison of laboratory features and application of machine learning approaches to patient differentiation and prediction of optimal management pathways are forthcoming.

Keywords: Multisystem Inflammatory Syndrome in Children (MIS-C); Kawasaki disease; 2019 novel coronavirus disease (COVID-19) pandemic; coronary artery abnormality; Pediatric Multisystem Inflammatory Syndrome temporally associated with COVID-19 (PIMS)

TABLE. Comparison Stratified by COVID Status Group

	POSITIVE	POSSIBLE	NEGATIVE	UNKNOWN	
	n=781 (59%)	n=52 (4%)	n=310 (23%)	n=187 (14%)	p value
Demographics and Clinical Presentation					
Males	62%	44%	61%	62%	0.10
Age (years)*	8.7	8.4	2.9	3.4	<0.001
KD criteria					<0.001
4 or 5	12%	25%	32%	53%	
2 or 3	38%	48%	50%	28%	
0 or 1	50%	37%	18%	19%	
Diarrhea	47%	40%	33%	26%	<0.001
Abdominal pain	62%	54%	32%	18%	<0.001
Vomiting	60%	54%	43%	35%	<0.001
Shock	35%	31%	6%	4%	<0.001
Management					
Symptom onset to first immune modulation treatment (days)*	5.3	13.1	8.8	6.9	<0.001
IVIG	92%	89%	89%	86%	0.54
IV steroids	76%	63%	34%	16%	<0.001
Anakinra	21%	10%	6%	3%	<0.001
ICU stay	59%	46%	15%	7%	<0.001
Inotropes	48%	29%	19%	6%	<0.001
Outcomes					
Hospital stay (days)*	6	5	4	4	<0.001
Mean max CA Z score**	1.48	1.26	2.23	1.87	<0.001
Max Z score CA category					0.08
Normal	81.1%	86.5%	76.4%	84.0%	
Dilation	7.8%	5.8%	6.1%	3.7%	
Small CAA	11.4%	7.7%	10.7%	8.5%	
Medium CAA	0.5%	0	3.9%	2.7%	
Large CAA	0.3%	0	2.9%	1.1%	

*Median

**At any timepoint and in any branch

CA, coronary artery; CAA, coronary artery aneurysm; ICU, intensive care unit; IV, intravenous; IVIG intravenous immune globulin; max, maximum

O-102

Anakinra in treatment of pediatric inflammatory multisystem syndrome temporally associated with SARS-COV2 infection (PIMS-TS) with cardiac involvement

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Background and Aim: PIMS-TS often affects the cardiovascular system resulting in myocarditis and coronary artery abnormalities (CAA). Immunosuppressive therapy is the primary treatment of PIMS-TS; however, controversies on the best regimen remain. The use of anakinra as a second-line treatment in children with cardiac involvement is often proposed lately. This prospective observational study aimed to determine the incidence of cardiac involvement in PIMS-TS and to evaluate the effectiveness and safety of anakinra in its treatment

Methods: From July 2020 till December 2021, we have treated 22 children with PIMS-TS (12 boys, 10
girls; aged 0,3-15,75y (median-4y). Echocardiography assessing coronary arteries and systolic function was performed at admission, during the hospitalization, at discharge and
6-8 weeks after the onset.

Results: Fourteen(63%) of the patients had coronary artery dilatation (z-score: 2,1-11,8; median:2,9), 6(27%) deterioration of systolic function, 5(22%) both, 20(90%) had elevated NTproBNP. Only in 2 children (9%) there was no cardiac involvement; in 3(14%) the only cardiac sign was elevated NTproBNP. Two children required admission to ICU. In the first line immunosuppressive treatment, we administered intravenous immunoglobulins (IVIG) in dose 2g/kg in all patients – of which in 16 as a monotherapy, in 6 together with glucocorticosteroids (GCS) because of their severe condition. Seven patients (32%) recovered after a single IVIG infusion, 3(14%) after repeated IVIG infusions. In this subgroup all CAA normalized or got significantly smaller with the change in the median z-score from 2,8 to 1,3. In 10 (45%) patients with cardiac involvment and insufficient response (4 with previous IVIG treatment, 6 with previous IVIG+GCS treatment) we administered anakinra obtaining clinical, laboratory and echocardiographic improvement in all of the patients (good systolic function and normalized or significantly smaller CAA: from z-score median = 2,8 (range: 2,1-11,8) to z-score median = 1,9 (range:0-3,2)), with no side effects. The median time to introduce anakinra (for median 9 days treatment) was 4 days after the first-line treatment. In 2 patients with suboptimal effect of first-line IVIG monotherapy, but no cardiac involvement, GCS as the second-line treatment were used. Median time of hospitalization was 18 days

Conclusion: Cardiac involvement is common in PIMS-TS. Anakinra seems to be effective and safe in its treatment

Keywords: PIMS-TS, cardiovascular involvement, anakinra

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Adult Congenital Heart Disease

P-001
Back to basics - the role of chest X-ray in transposition of the great arteries post atrial switch repair

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Background and Aim: Patients with dextro-transposition of the great arteries after atrial switch procedure (TGA-Mustard/Senning) have increased long-term risk of cardiovascular morbidity and mortality. Despite its potential prognostic information, the role of chest radiography in the follow-up of TGA-Mustard/Senning is currently limited. Therefore, this study evaluates the relationship of cardiothoracic index (CTI) on chest radiography with clinical parameters and outcome in TGA-Mustard/Senning.
Methods: Clinical records of all TGA-Mustard/Senning under active follow-up between January 2000 and December 2018 were reviewed. Frontal chest radiographs at or close to first and last follow-up were collected and CTI was calculated. Descriptive statistics and Kaplan-Meier analysis were performed. The relationship between CTI and clinical parameters was assessed using linear regression models.
Results: One hundred thirteen patients were included in the study, of which 64 patients (44% Mustard, 56% Senning) had at least 1 chest X-ray during follow-up. Median CTI at baseline was 0.52 (IQR 0.49-0.59), which formed the cutoff to classify the cohort in two groups: low CTI (n = 31) versus high CTI (n = 33). At baseline, a significantly higher prevalence of supraventricular tachycardia was seen in the high CTI group (69.7% versus 41.9%, p = 0.043). There was no difference in the prevalence of heart failure, but a linear relationship between log NT-proBNP and CTI was seen ($R^2 = 0.176$, p = 0.024). Furthermore, a linear relationship was observed between peak oxygen consumption during exercise and CTI ($R^2 = 0.179$, p = 0.004). During a median follow-up time of 15.4 years (range 9.7-17.5), 6 patients died. Mortality was only seen in the high CTI group (p = 0.025).
Conclusion: CTI on chest radiography carries prognostic information in the evaluation of TGA-Mustard/Senning. Therefore a classic chest x-ray should be considered intermittently during follow-up of asymptomatic patients and if clinically indicated.

Keywords: Transposition of great arteries; Atrial switch operation; Radiography; Cardiothoracic ratio; Outcome

P-002
Two cases of repaired tetralogy of fallot with coexisting aortic arch abnormalities

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Background and Aim: Tetralogy of Fallot (ToF) is considered to be the most common, complex cyanotic congenital heart disease (CHD), with its operated form (ToFop) being the most common in adulthood. It is not uncommon for other congenital anomalies to coexist in patients with ToF, with more common various variants of the aortic arch [25% right aortic arch (RAA).
Methods: We report two cases of ToF with coexisting aortic arch abnormalities. The diagnostic imaging techniques that were used, included echocardiography and cardiac magnetic resonance (CMR).
Results: The first case concerns a 29-year-old patient with a history of ToFop and a right aortic arch, with an isolated left subclavian artery (LSA). In particular, the left subclavian artery was formed by the left carotid artery by the fusion of smaller vascular branches. In addition, it is worth mentioning the presence of a significant coarctation of the aorta (CoA), also confirmed by CMR. The second case is about a 23-year-old patient with a history of ToFop and a right aortic arch with coexisting abnormalities in the origin of cervical vessels. More specifically, the first branch to arise from the aortic arch was a trunk bifurcating into the two common carotid arteries (common bicarotid trunk), with the right and the left SAs being the second and the third branches, respectively. The LSA also presented a retroesophageal course.
Conclusion: The coexistence of ToF and CoA is a rare combination of obstruction in outflow tracts of both circulations. The presence of an isolated LSA in combination with the other two entities seems to be even rarer, given that we managed to find only one similar case in literature. The cases presented, highlight the importance of using all available imaging modalities, especially when an intervention is considered, to prevent periprocedural complications.

Keywords: Tetralogy of Fallot, Right aortic arch, Aortic arch abnormalities, Coarctation of the aorta

Case (1) & Case (2) images

P-003
Trend in mortality from congenital heart disease according to the area of residence in spain

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Background and Aim: Adults with congenital heart disease (ACHD) are a growing population due to advances in medical and surgical therapies. Therefore, the aim of this study was to estimate the trend in mortality from congenital heart disease according to the area of residence in Spain between the years 2003 and 2018.
Methods: We selected all deaths assigned to the International Classification of Diseases (ICD-10) for total congenital heart diseases. The population, the number of deaths and the population according to the age, sex and population size of the municipality of residence have been obtained from the National Institute of

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Statistics. The size of the municipality of residence has been grouped into three categories: less than 10,000 inhabitants (rural areas), between 10,000 and 100,000 inhabitants (small urban areas) and more than 100,000 inhabitants (large urban areas). In each area we have calculated the average annual percentage change in mortality rate (APCM).
Results: Between 2003 and 2018 the APCM in the mortality rate from congenital heart diseases in large urban, small urban and rural areas was respectively -5.5, -3.6 and -4.6 in men, and -6.0, -4.3, and -5.0 in women.
Conclusion: Large urban areas showed the greatest decrease in mortality from congenital heart diseases. In both rural and urban areas, mortality fell more for women than for men.

Keywords: congenital heart, mortality, rural, Spain, urban

P-004

Health care supply for adults with congenital heart disease associated with aortic involvement

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Background and Aim: All patients with congenital heart disease (CHD) are chronically ill from their cardiac disease. Despite increasing evidence that aortic alterations are becoming relevant, the importance of aortopathy in CHD has long been underestimated. This study was conducted to determine the health-status of patients and/or the provision of health services of adults with CHD (ACHD) with manifest aortopathy or at risk thereof.
Methods: In a questionnaire-based cross-sectional survey, the "real-life"-care of ACHD was analysed, comparing patients with risk of developing aortopathy and/or manifest aortopathy.
Results: Of the 563 enrolled ACHD (49.6% female, mean age 35.8 ± 12.1 years) 56.8% had a risk of developing aortopathy and/or manifest aortopathy. Of the 320 patients at risk, 187 (33.2% of the total number) had a proven aortopathy. Within this subgroup, the basic medical care for CHD-independent medical problems was given by primary medical care providers [family doctors/general practitioners (GP) in 89.4%, internists in 13.4%, physicians of another specialty in 2.5% (n = 8)]. Almost all primary medical care providers knew about the CHD of their patients. Even for CHD-specific health problems, the basic medical care of risk patients was provided by a family doctor or GP in 56.6% and by an internist in 18.4%. 30.0% primarily consulted another specialist, including cardiologists. Only 32.8% of ACHD at risk had ever been referred to a CHD specialist by a GP for cardiac problems related to their CHD. In contrast, the need for advice was high for ACHD with aortopathy and related mainly to physical activity, employment and education, pregnancy, rehabilitation or health and life insurance. Only 35.5% of patients at risk indicated that their information on specific care structures for ACHD was sufficient, and a further 38.1% of patients were aware of patient organizations
Conclusion: As aortopathy gains in importance with increasing age and complexity of CHD, almost all affected ACHD need lifelong medical advice and access to modern, scientifically based care concepts. According to the study-results, primary care providers and also patients are mostly uninformed about the specialized ACHD facilities. The future goal is therefore to create a better awareness of CHD problems among both primary care physicians and the patients concerned.

Keywords: CHD, congenital heart disease, aortopathy, prevention, health care

P-005

Transition of care in congenital heart disease: a single center experience at a large urban tertiary care setting

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Background and Aim: The term "transition of care" (TOC) refers to the continuity of health care during movement from one healthcare setting to another as care needs change during a chronic illness. For young people with chronic illnesses, successful TOC often refers to the process of moving from the pediatric to an adult healthcare setting and is crucial for health outcomes. Large proportions of young pts. with CHD are lost to follow-up (LTF) or experience gaps in care after leaving pediatric cardiology (PC) due to lack of appropriate TOC. We sought to describe key factors related to TOC or LTF in a large tertiary care facility in pts. with CHD.
Methods: Patients were identified using an electronic medical record (EMR) (Epic). Inclusion criteria included: congenital heart defects, age ≥ 15 years seen in the ambulatory PC clinic between 2013 and 2014. Exclusion criteria included: deceased patients. Clinical and demographic variables were collected by retrospective chart review for each pt. meeting inclusion criteria. Patient's charts were reviewed between 2015-2021 to determine if they were a) still in PC care,b) transitioned to adult cardiology (AC)/ACHD or c) LTF. Independent predictors for successful TOC or LTF was performed using Chi-Squared analysis.
Results: Total of N = 322 patients were included. 49% (N = 159) had successful TOC from PC to AC/ACHD, 22% (N = 70) remained with PC and 29% (N = 93) were LTF. More patients with successful TOC to AC/ACHD had complex lesions (21%) and had seen PC more than once a year (28%). The majority of pts. LTF were either uninsured or had public insurance (Medicaid) (62%), had moderately complex CHD (64%) and saw PC less than annually (30%).
Conclusion: In this setting, successful TOC from PC to AC/ACHD happened infrequently in pts. with CHD. Pts. with severely complex CHD lesions and visits more than once a year to the PC had successful TOF. Pts. with public insurance/Medicaid, as well as moderately complex CHD lesions were LTF. Lack of successful TOC begins during early adolescence despite pts. being in contact with healthcare providers. Efforts need to be made to improve TOC to avoid long term care in patients with CHD.

Keywords: Transition to Adult Care, Transition of Care, Adolescent Health Services, Adult Congenital Heart Disease, Chronic Disease

P-006

An unusual cause of pulmonary hypertension, partial anomalous pulmonary venous return in a male with previous percutaneous atrial septal defect closure

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Background and Aim: A 46-year-old male patient admitted with dyspnea who has a history of percutaneous atrial septal

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defect(ASD) closure(10 years ago, 7 mm ASD). He was on bronchodilators for chronic obstructive pulmonary disease(COPD). Transthoracic echocardiography revealed mild tricuspid valve regurgitation, mild right chamber dilation and pulmonary hypertension (Figure 1. A-B) As the COPD was mild, the patient underwent right heart catheterization(RHC) which revealed the right upper pulmonary vein (RUPV) draining into the superior vena cava(SVC) and precapillary pulmonary arterial hypertension(PAH) (Pulmonary arterial pressure: 53/38/22 mmHg, PVR: 4.21 WU, Qp/Qs: 1.66) (Figure 1.C-D).
Methods: To examine all the pulmonary veins and the presence of sinus venosus ASDs transesophageal echocardiogram(TEE) and computed tomografi pulmonary angiography(CTPA) were performed. On TEE interatrial occluder device was seen and IAS was intact. CTPA showed that RUPV was draining to SVC (Figure 1.E). As the pulmonary vascular resistance was high and the patient also did not want to undergo surgery, macitentan 10 mg was initiated for pulmonary hypertension.
Results: We have reported a case of partial anomalous pulmonary venous return (PAPVR) to SVC, leading to PAH. The presented patient had undergone TEE examinations before ASD closure; however, a review of the previous reports indicated no anomalies of the pulmonary veins.
Conclusion: Given the widespread use of TEE, cardiologists should be aware of the possibility of PAPVR, especially in patients with other congenital anomalies. RHC should be performed before the ASD closure and CTPA should be performed in the suspicion of PAPVR.

Keywords: partial anomalous pulmonary venous return, pulmonary hypertension

Transtorasic and angiographic images

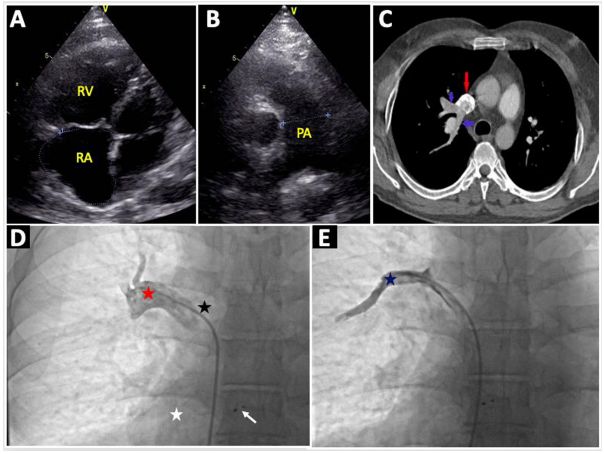

Figure 1: Figure A, Transtorasic echocardiography apical four chamber view (dilated right heart chamber, RA area 21.6 cm2) Figure B, Transtorasic echocardiography short axis view (dilated pulmonary artery with diameter 3.2 cm) Figure C, Axial computed tomography pulmonary angiography section shows that right upper pulmonary veins (blue arrows) are draining to superior vena cava (red arrow) Figure D, Angiographic images of right upper pulmonary vein (red asterics) drain to superior vena cava (blue asterics) with an enlarged root of approximately 2 cm (White asterics: right atrium, White arrow: Atrial septal defect occluder device) Figure E, Angiographic images of right upper pulmonary vein (blue asterics) drain to superior vena cava RA: right atrium, SVC: superior vena cava

P-007

Acquired interruption of the aortic arch; the natural history of unrepaired native coarctation of aorta with successful transcatheter intervention

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Background and Aim: Coarctation of Aorta (COA) and Interrupted Aortic Arch are morphological, pathogenetic and embryogenic distinct phenotypes. There is paucity of literature alluding to the natural history of unrepaired COA. Herein, we describe cases of evolutionary acquired interruption of the Aortic arch in isolated Coarctation and/or Coarctation syndrome (associated functionally bicuspid Aortic valve) with age, that were successfully managed percutaneously.
Methods: Demographics (age, gender), clinical characteristics (symptoms, cormobid factors, examination), pre-procedural imaging data (trans thoracic echocardiography, CT angiography), procedural and outcome (immediate, short- term) data etc. was extracted from the institutional PROTEA filmmaker electronic database, appraised on individual merits and analysed using the latest Stata/SE software analysis package by StataCorp.
Results: A total of three cases who subsequently had percutaneous intervention in November 2021, were reviewed. The youngest and oldest patient was 10 and 36 years old respectively, and all female. They all had systemic hypertension (systolic BP >140 mmHg), significant collateralisation and preserved left ventricle (LV) systolic function at baseline. Pre-procedural pressure differences (peak-peak systolic) ranged from 40- 70 mmHg. Aortography using the kissing technique (simultaneous ascending (radial access) and descending Aorta (femoral)) was performed before an attempt at perforation/ re-canalisation with the stiff end of the Amplatzer and Terumo as well as the Abbott PTCA Crossit guide wires, followed by successful intraluminal stent angioplasty using the covered Andratec stent portfolio. Post-procedural systolic pressure difference ranged between 0-4mmHg. There were no intra- and/or post-procedural complications and the adjacent Subclavian artery could be spared in all cases. Longitudinal assessment will serially focus on blood pressure control, LV reverse remodelling patterns, collaterisation regression and freedom from re-intervention.
Conclusion: Acquired interruption in patients with baseline native COA has not been frequently documented. While limited by the number of observed cases, transcatheter management for this cohort of patients yields good immediate outcomes well within good procedural safety margins, as they may not be favourable surgical substrates owing to their extensive collateralisation contributing to exaggerated intra-operative bleeding risk. This work will contribute to the earlier body of work assessing LV remodelling following transcatheter management, in the absence of a surgical comparative cohort.

Keywords: Natural history, Coarctation of Aorta, Acquired interruption, Transcatheter intervention

P-008

The contemporary outcomes of ebstein's anomaly in south wales: a 30-year review at a single institution

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Background and Aim: Ebstein's anomaly of the tricuspid valve (TV) is increasingly recognised antenatally. Impact of this changing trend has not been evaluated. We aimed to assess the time of

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diagnosis, management, and survival outcomes of Ebstein's anomaly in South Wales.
METHOD:Retrospective review of all children diagnosed with Ebstein's anomaly at the University Hospital of Wales (UHW) over 30 years.
Results: 74 patients were managed at UHW between 1990 and 2020, with 53 cases eligible for inclusion. 13 cases were diagnosed antenatally with a marked increase in detection from 0% to 100% in 2020 following improved antenatal screening across South Wales from 2001. Presenting symptoms varied. 30.00% (n = 3) of neonates and infants had murmurs and 54.55% (n = 6) adults had shortness of breath. 5.66% (n = 3) patients presented in arrhythmia with junctional bradycardia in 1 patient and others in atrial arrhythmia. Review of available echocardiogram images (n = 14) noted GOSE indexes <1 for all. Most common cardiac comorbidities at diagnosis were interatrial communications, 41.51% (n = 22), and tachyarrhythmias, 35.85% (n = 19). However, 45.28% (n = 24) patients had arrhythmias at some point during their clinical course. 45.76% (n = 10) had catheter ablations with 40.00% (n = 4) requiring repeat ablations and half (n = 12) remained on anti-arrhythmic medications at review. Analysis of TV interventions found 8 patients requiring pacemakers post-operatively. Those who had TV replacement were more likely to require pacemakers post-operatively compared to those who had TV repair, OR 3.00 (0.23 - 39.61), p = 0.04. At review, 54.72% (n = 29) were asymptomatic with 41.38% (n = 12) requiring no intervention. In symptomatic patients, majority 41.67% (n = 10) received surgical and medical management, and medical management alone in 33.33% (n = 8). Cumulative survival was 90% at 20 years, 87% at 60 years and overall mortality rate was 13.21% (n = 7). Owing to 13 antenatal diagnoses, meaningful comparison of outcomes was not possible.
Conclusion: Rates of antenatal diagnosis have increased to 100% owing to improved antenatal screening in South Wales and further follow up is required to analyse its influence on outcomes. Survival outcomes are good with over 85% being alive at age 60 and over half remaining asymptomatic with most requiring medical management at most recent follow up.

Keywords: Ebstein's anomaly, outcome, surgery, foetal

P-009/Moderated Poster
Home-based resistance training in fontan patients: it's feasible, safe and efficient!

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Background and Aim: One of the principal limitations observed after a Fontan procedure is an impaired cardiac output adaptation which affects exercise capacity and reduces quality of life. Numerous studies showed that cardiovascular training improves exercise tolerance in Fontan population but less is known about resistance training. Considering that skeletal-muscle pump is crucial to increase venous return and cardiac output, the purpose of this study is to explore feasibility, safety and efficiency of a home-based resistance training on maximal exercise capacity, cardiorespiratory endurance and quality of life in this population.
Methods: 12 young Fontan adults (18 to 32 years old) with a 1-2 NYHA functional class have been recruited. Initial enrolment evaluations at the hospital assessed lean body mass, segmental forces, maximal oxygen consumption (VO2max) during an incremental treadmill test, cardiovascular endurance on treadmill at 85% VO2max, lower limb muscles endurance and quality of life. After 12 weeks of a structured exercise program with 3 one-hour resistance training sessions per week supervised online by a physiotherapist, the same measures have been repeated. All training sessions included 5 exercises performed with elastic band resistance that progressively increased throughout weeks. Progression rate has been individualized to each participant.
Results: 11/12 patients initially recruited completed more than 80% of the training sessions. No adverse event has been reported during the 400 training sessions. No changes were observed regarding the segmental forces, nor in the lean body mass or lower limb muscles endurance. Despite no differences in their VO2max, all participants improved endurance time on the treadmill after intervention (448 ± 240 sec vs 771 ± 446 sec; p = 0,047; mean increase 72%). Quality of life evaluated with SF-36 was similar to the initial evaluation, but all participants reported a significant improvement of their self-perceived exercise capacity (p = 0,002).
Conclusion: Home-based exercise program centered on resistance training is safe and feasible in Fontan population with no more than a slightly impaired functional capacity. It's significantly improving cardiovascular endurance without any changes on muscle mass or VO2max. Further research is needed to evaluate if this improvement is sufficient to increase quality of life in patients with a more blunted functional capacity.

Keywords: Fontan, Exercise, Endurance, VO2max

P-010
Native coarctation in adulthood: modes of referral, diagnosis and treatment in tertiary referral centers for adult congenital heart disease

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Background and Aim: The majority of patients with aortic coarctation (CoA) is diagnosed and treated during childhood while only a minority reaches adulthood with native coarctation. The intention of our study was to describe the symptoms, mode of referral and treatment in patients with adult CoA whose diagnosis was only made in adulthood in 3 tertiary referral centers for Adult Congenital Heart Disease.

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Methods: The patient histories of adolescents and adults treated from 2010 to 2021 with native coarcation were retrieved from the databases of our hospitals.
Results: 14 patients (mean age 36.2 years, range: 17-64) were included. The mean interval between the diagnosis of arterial hypertension and diagnosis of CoA was 10 (0-37) years. In one patient, despite correct diagnosis of CoA, treatment was delayed for 11 years. 10/14 patients received antihypertensive therapy with a mean of 2.8 (1-6) medications. Prior to diagnosis of CoA, 2 patients suffered intracranial hemorrhage due to ruptured aneurysms and 4 patients had claudication or abdominal angina. Other symptoms were hemoptysis, chest pain, headache, and dyspnea. Before referral to a center for Adult Congenital Heart Disease, an average of 2.2 (1-3) consultants from different subspecialties were consulted. All patients had severe CoA with >50% narrowing of the aortic diameter. The mean invasive peak-to-peak gradient was 50.4 (20-80) mmHg. Interventional stent implantation was performed in 9 patients, 4 patients were sent for surgery, the remaining patient is awaiting treatment. Following treatment, the mean residual gradient measured was 2 (0-17) mmHg. Antihypertensive therapy had to be continued in 10/14 patients with an average of 2.4 (1-4) drugs. Blood pressure was within the target range in 10 patients.
Conclusion: Adult patients with native CoA usually presented with refractory arterial hypertension. The diagnosis was established with a mean delay of 10 years although it is integral part of the differential diagnosis of arterial hypertension. Due to the rarity of this condition diagnostics and treatment frequently focused on secondary symptoms which obscured the underlying pathology. Despite the rarity of CoA in adults, it is crucial to highlight the importance of this differential diagnosis in the assessment of arterial hypertension.

Keywords: aortic coarcation, CoA, Adult Congenital Heart Disease, aortopathy, congenital heart disease, stent therapy

P-011
Cardio-thoracic ratio – still an important parameter in patients with ebstein's anomaly?

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Background and Aim: Cardio-thoracic ratio (CTR) was a former indicator for surgical interventions in Ebstein's anomaly (EA). An enlarged heart can affect both, lung function and exercise capacity. Thus, in a prospective study we investigated whether CTR correlates with lung function or exercise capacity in native EA pts.
Methods: Forty-one pts (age range 18-78 years, 27 female) with native EA underwent a cardiovascular magnetic resonance (CMR), spirometry, cardiopulmonary exercise test and their body mass index (BMI) was calculated. We determined forced vital capacity (FVC), forced expiratory volume in 1 second (FEV1), and its ratio as z-scores as well as peak oxygen uptake ($V'O_2$peak) and CTR. For statistical analysis we used Pearson's correlations and multivariate regressions.
Results: Mean CTR was 0.61 ± 0.07 (range 0.44-0.73). Thirty-nine (95%) pts had a CTR >0.5 (0.62 ± 0.06), fifteen of them a CTR greater than 0.65 (0.68 ± 0.02). Lung volumes were mainly normal (FVC z-score: -0.41 ± 1.2 and FEV1 z-score: -0.46 ± 1.2) with only five pts with restrictive (FVC z-score<-1.645) and three with obstructive (FEV1/FVC z-score<-1.645) patterns. $V'O_2$peak was 22.4 ± 8.5ml/min/kg, representing 80.0 ± 22.3 % of predicted. The mean BMI was 26.4 ± 5.4kg/m². Pearson's correlation showed significancy between $V'O_2$peak and CTR ($r = -0.431$, $p = 0.006$), FVC z-score ($r = 0.503$, $p = 0.001$), FEV1 z-score ($r = 0.427$, $p = 0.006$), and BMI ($r = -0.596$, $p<0.001$), respectively. CTR itself is associated with FVC z-score ($r = -0.446$, $p = 0.004$), and BMI ($r = 0.324$, $p = 0.039$). In multivariate regression, the only predictor on $V'O_2$peak in this case remains BMI ($\beta = -0.450$, $p = 0.002$).
Conclusion: These analyses show slight correlations between CTR and lung volumes as well as $V'O_2$peak in native EA. Thus, it seems justified to abandon CTR as a parameter for surgery.

Keywords: Ebstein, CPET

P-012
A case of pulmonary valve reconstruction using ozaki technique

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Background and Aim: Aortic valve neocuspidization with glutaraldehide-treated autologous pericardium according to the Ozaki technique has been proven to be an effective therapy for the treatment of aortic valve diseases in adult and pediatric patients. We report a case where we performed this nouvel technique for reconstruction of pulmonary valve.
Methods: A 33-year-old male patient was admitted to our hospital with shortness of breath. Transthoracic echocardiography showed severely hypertrophic right ventricle and severe pulmonary valvular and sub-valvular stenosis with maximum gradient of 150 mm Hg. Pulmonary annulus was measured 15.6 mm. After the heart valve team meeting, surgery was decided upon.
Results: After median sternotomy pericardium was excised and then was treated with a 0.6% glutaraldehyde solution for 10min and rinsed 3 times using a physiologic saline solution. Aortic-arterial and bicaval-venous cannulation was performed, then cardiopulmonary bypass (CPB) was initiated. When PA was opened, we found that pulmonary valve is monocuspid. Pulmonary valve leaflet was resected, then subvalvular and infundibular hypertrophic tissues were resected. The distance between each commissure was measured following Ozaki's recommendations. The sizes of measured comissures were 25 mm, 27 mm and 29 mm. Appropriate size pericardial cusps were trimmed and sutured to each annulus, making a neotricuspidization of PV (Image 1). Weaning from CPB was performed without complications. Post-operative trans-esophageal echocardiogram showed trivial pulmonary regurgitation and a maximum gradient across the pulmonary valve of 15 mm Hg.
Conclusion: The extubation was performed on the same day and the ICU stay was 2 days. The patient was discharged from hospital at 7-th postoperative day without complications. Post-operative follow-up transthoracic echocardiography showed the reconstructed PV with no residual regurgitation, good coaptation height and maximal pulmonary gradient was measured 25 mm Hg. At 1 year of followup, a favorable evolution was observed.

Keywords: Pulmonary valve reconstruction, pulmonary valve neocuspidization.

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P-013

Feasibility and durability of valve sparing root replacement after previous cardiac surgery for congenital heart disease

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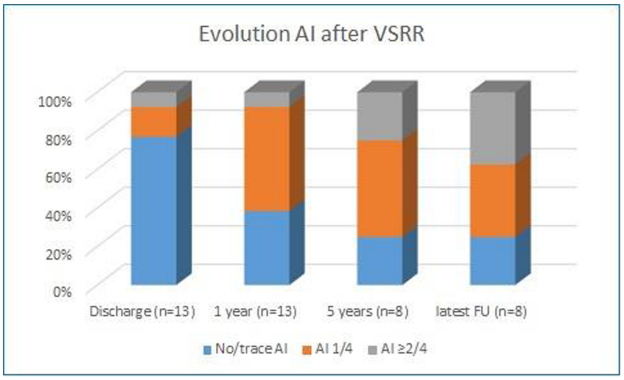
Background and Aim: Patients operated for congenital heart disease (CHD) in childhood may develop aortic valve and root pathology in the native aorta or the neo-aorta, necessitating reoperation in adolescence or adulthood. In selected patients a valve sparing root repair (VSRR) can be considered. Since we started the VSRR program in 2000, over 200 patients have undergone this procedure, of which 13 for CHD (6.5%). We aimed to study the feasibility and durability of the VSRR in CHD, Bentall patients after previous CHD surgery served as controls.
Methods: Over the study period (2000 - 2020), 13 patients underwent VSRR, 10 had a Bentall procedure. Inclusion criteria were need for aortic root and/or aortic valve surgery, and previous surgery for CHD repair by sternotomy. Exclusion criteria were first time sternotomy (e.g. aortic root pathology after coarctation repair), redo on an aortic valve prosthesis, and patients too small to receive an adult sized conduit and having the possibility for a Ross procedure.
Results: VSRR and Bentall were performed at a median age of 29y (range 10-58y) and 45y (7-56y). All patients had undergone at least 1 previous surgery (range 1-3). Interval since last surgery was 14y (7-48y) for VSRR and 23y (3-37y) for Bentall. Both groups demonstrated a large variety of diagnoses (bicuspid valve disease, outlet VSD, subaortic membrane, tetralogy of Fallot, truncus arteriosus, transposition). The maximal root diameter was 53 mm (30-63 mm) in VSRR and 43 mm (22-54 mm) in Bentall. Preoperative peak gradients were higher in the Bentall group (30 mmHg (0-100 mmHg) vs VSRR 6 mmHg (0-34 mmHg), p = 0.023), median aortic incompetence was comparable (grade 2). Median aortic cross clamp time was longer in VSRR (138min (98-183min) than in Bentall (114 min (17-165 min), p = 0.028). No operative nor late mortality occurred at mid-term follow-up (VSRR: 6.7y (1-19.6y), Bentall 6.3y (1.3-11.6y)). No aortic (valve) reoperations were needed, aortic incompetence after VSRR tended to progress slightly during follow-up.
Conclusion: VSRR can be applied in a variety of diagnoses, both in native and in neo-aorta, without mortality nor reoperations at a follow-up of over 10 years. Progressive aortic incompetence after VSRR needs close monitoring. Multicentre efforts from experienced groups are welcomed to combine long-term outcome data.

Keywords: valve sparing root repair, aortic root disease, congenital heart surgery

Evolution of aortic incompetence grade after VSRR



P-014

Properties of plasma clots and cellular liver damage in adult patients following fontan procedure: relation to clot permeability and lysis time

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Background and Aim: Thromboembolic complications are a major cause of morbidity and mortality following Fontan (FO) surgery. It is also well established that altered FO circulation results in systemic complications, including liver damage. We sought to evaluate whether dysfunction of this source of hemostatic factors may result in changes of fibrin clot properties.
Methods: A permeation coefficient (Ks) and clot lysis time (CLT) were assessed in 66 FO patients, aged 23.0 years [IQR 19.3-27.0], and 59 controls, aged 24.0 years [IQR 19.0-29.0]. Ks was determined using a pressure-driven system. CLT value was measured according to assay described by Pieters et al. Liver-derived hemostatic factors along with liver function parameters were evaluated. The median time between FO operation and investigation was 20.5 years [IQR 16.3-22.0].
Results: FO patients had lower Ks (P = 0.005) and prolonged CLT (P<0.001) compared to controls. Ks correlated with CLT (r = -0.28), FVIII (r = -0.30), FIX (r = -0.38), fibrinogen (r = -0.41), ALT (r = -0.25), AST (r = -0.26) and GGTP (r = -0.27), (all P<0.05). CLT correlated with the time between FO operation and investigation (r = 0.29) and FIX (r = 0.25), (all P<0.05). After adjustment for potential cofounders, TAFI antigen and GGTP were independent predictors of reduced Ks (OR 1.041 per 1% increase, 95% CI 1.009-1.081, P = 0.011 and OR 1.025 per 1 U/L increase, 95% CI 1.005-1.053, P = 0.014, respectively). Protein C and LDL cholesterol predicted prolonged CLT (OR 1.078 per 1% increase, 95% CI 1.027-1.153, P = 0.002 and OR 6.360 per 1 µmol/L increase, 95% CI 1.492-39-894, P = 0.011, respectively). GGTP correlated positively with time between FO surgery and investigation (r = 0.25, P = 0.045) and patients with abnormal elevated GGTP activity (n = 28, 42.4%) had decreased Ks, compared to the others (5.9 x 10-9cm2 vs. 6.8 x 10-9cm2, P = 0.042).
Conclusion: Our study shows that cellular liver damage was associated with prothrombotic clot phenotype reflected by Ks and CLT, which increased the risk of complications in adult patients following FO procedure.

Keywords: clot lysis time, permeation coefficient, Fontan procedure, liver dysfunction

P-016

Prevalence of cardiovascular risk factors in adult congenital heart disease

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Background and Aim: Adult patients with congenital heart disease (ACHD) are at an increased risk of accelerated cardiovascular

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disease, such as myocardial infarction, heart failure, or death. There is scarce information on risk factors and acquired heart disease in this ageing population. However, these superimposed cardiovascular risk factors may be amplified in these patients who are often already at risk of cardiovascular complications
We aim to assess the prevalence of cardiovascular risk factors in ACHD patients, and their association with the underlying disease complexity
Methods: We retrospectively collected cardiovascular risk factors from ACHD patients followed at our tertiary center from January 2017 to January 2020. Congenital heart diseases (CHD) were classified in 3 groups according to their complexities based the Bethesda classification: simple, intermediate, or complex.
Results: We included 661 patients (mean age 35.5 +/- 4.9 years, 52% women). Overweight was the most prevalent modifiable cardiovascular risk factor affecting 199 (30%) patients and 8% were obese. A physically active lifestyle was reached by 64%. The prevalence of overweight and sedentary patients was higher in women than in men. ($p = 0.04$ and 0.03 respectively). Tobacco use was reported in 20% of patients, hypertension in 7%, dyslipidemia in 4% and diabetes in 2%.
Dyslipidemia was less common in patients with complex heart disease ($p = 0.03$). Only 97 patients (15%) had no cardiovascular risk factors There was no significant association between the complexity of CHD and the risk factors.
Conclusion: A large majority of ACHD patients have cardiovascular risk factor. A comprehensive cardiovascular prevention including screening, life-style choices, and therapies, with goals to diminish and eliminate modifiable cardiovascular risk factors in primary and secondary cardiovascular prevention is essential regardless of the CHD complexity.

Keywords: Adult Congenital Heart Disease, cardiovascular risk factors

P-017/Moderated Poster
Bioelectrical impedance analysis as a potential risk marker in adults with marfan syndrome

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Background and Aim: : It is clinically widely overlooked that many patients with Marfan syndrome (MFS) are obese, as anthropometric routine parameters are not very suitable for detection of obesity. In contrast, the Bioelectrical Impedance Analysis (BIA) provides reliable noninvasive data about the body composition of patients. The aim of the study was to assess the body composition of patients with MFS/LDS by BIA in order to detect occult obesity, which may be a risk marker for aortic or vascular complications.
Methods: In this exploratory cross-sectional study, 32 patients (56.3% female; mean age: 36.4 ± 10.7 [range: 17 - 56] years) with a molecular genetic ($n = 28$; 87,5%) or clinical ($n = 4$; 12,5%) diagnosis of MFS ($n = 28$) or LDS ($n = 4$) were enrolled between June 2020 an August 2021. All BIA-measurements were performed with the Multifrequence-Impedance-Analyzer Nutriguard-MS (Data Input, Pöcking, Germany).
Results: The MFS collective was significantly different from an age-, sex-, and Body Mass Index (BMI)-adjusted healthy control group in terms of percentage body fat ($p<.001$), percentage cellularity (<.001), ECM/BCM index ($p<.001$), and phase angle ($p<.001$). The mean BIA-measured bodyfat was 29.3 ± 8.2 % [range: 9.5 – 43.3%], while the mean calculated BMI of the included patients was 21.9 ± 3.6 kg/m2 [range: 15.2 – 29.7 kg/m2]. Therefore, using the obesity cut-off values for the body fat percentage of 25% in men and 35% in women, the BIA classifies as many as 14 patients (43.6%) as obese, while only 7 patients (21.9%) were pre-obese by BMI. The significant difference ($p<.001$) had an accordance of 40.2 %. In addition, there is a strong positive correlation ($r = .480$) between body fat determined by BIA and the diastolic diameter of the bulbus aortae ($p = .006$).
Conclusion: The fact that many patients with MFS are obese is widely unknown, although adipositas may be associated with impaired vascular endothelial function and an increased risk of cardiovascular complications. Also in patients with MFS/LDS, BIA allows a reliable assessment of the body composition beyond the normal anthropometric parameters, such as BMI. In the future, BIA-data may be of particular importance for the assessment of the vascular risk of MFS/LDS patients, besides the aortic diameters.

Keywords: ACHD, Marfan, bioelectrical impedance analysis, obesity

P-018
Myocardial fibrosis and its correlation with the predictors of morbidity and mortality in eisenmenger syndrome

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Background and Aim: Myocardial fibrosis which is picked up as late gadolinium enhancement (LGE) on Cardiac magnetic resonance imaging (CMR) is hypothesized to be the underlying mechanism of cardiac dysfunction in Eisenmenger syndrome and our study aims to evaluate the same.
Methods: In this prospective observational study, patients diagnosed with ES were enrolled to undergo CMR imaging after analyzing their demographic details and clinical profile. CINE images were acquired in various planes in Balanced Turbo Field Echo sequences following which LGE images were acquired ~15 minutes after dynamic perfusion sequence. The presence of LGE was analyzed against the clinical parameters predicting the disease severity. The patients were then followed up for next 18 months to look for major adverse cardiovascular events (MACE) if any.
Results: Out of a total of 18 ES patients [median age 20.50yrs (IQR 17-26)], 17 underwent LGE-CMR and 13(76.6%) were noted to have LGE. It was distributed along the RV myocardium (29.4%), RV papillary muscles (23.5%), RV trabeculae (23.5%) and over inter-ventricular septum (17.6%). Ten patients had uni-focal enhancement while 3 patients had multifocal enhancement. All three with multifocal enhancement had h/o hemoptysis($p = 0.045$). The median resting saturation in those without LGE, with uni-focal LGE and with multi-focal LGE was 96.5% (92.75-100), 89.5% (87.5-93.25) and 85% (84.5-87.5) respectively ($p = 0.039$). Median Hb was higher in those with multifocal involvement [20g/dl (19.75-20.3)] than those without LGE [14.4g/dl (13.38-15.43)] ($p = 0.019$). All 4(100%) NYHA class III patients had enhancement on MRI compared to 75% of those of class 1 without LGE($p = 0.026$). No correlation was found between the presence of LGE and severity of ventricular dysfunction (assessed by ejection fraction and cardiac volumes on CMR). Out of 18 patients, 3 succumbed to death over next 18 months of follow-up, secondary to congestive heart failure and sudden cardiac death. The median resting saturation of those who expired

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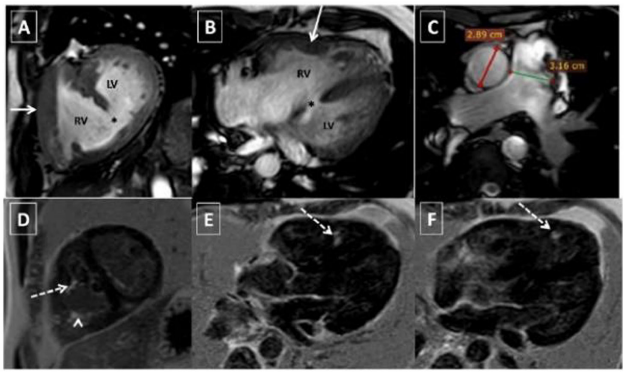
(85%) was lower than those alive [91%(IQR 89-97)] with p = 0.033. However LGE did not correlate with mortality.
Conclusion: Poor Resting Saturation, secondary erythrocytosis, hemoptysis and poor functional class correlate with presence of LGE on CMR and mortality correlates with severity of hypoxia.

Keywords: Eisenmenger syndrome, Pulmonary arterial hypertension, Late gadolinium enhancement, Cardiac magnetic resonance imaging, pro-BNP levels, resting saturation

CMRI



Cardiac MRI images (A-E) in a patient with Eisenmenger syndrome show muscular ventricular septal defect (*) with dilated confluent pulmonary arteries (ratio of main pulmonary artery to ascending aorta >1, as shown in C), dilated right ventricle (RV) with right ventricular hypertrophy (arrow in A, B) and multifocal areas of late gadolinium enhancement along RV papillary muscles (dashed arrow in D-F) and inferior RV wall (arrowhead in C). LV-left ventricle.

P-019
Prevalence and impact of cardiac involvement in turner syndrome

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Background and Aim: Turner syndrome (TS) is a sex chromosome disorder characterized by partial or complete loss of an X chromosome. TS patients have a higher mortality risk than age-matched females which is mainly due to cardiovascular disorders (CVD). Recently, a high prevalence of partial anomalous pulmonary venous connection (PAPVC) was described for TS patients. Data on the clinical impact of PAPVC, e.g. the need for surgical correction, however, is scarce. This study evaluates in a contemporary TS population the prevalence and impact of CVD with a special focus on PAPVC.
Methods: TS patients of all ages followed in a tertiary center were included in this study. Clinical data and reports of supplementary exams like echocardiography, cardiac magnetic resonance imaging (CMR) or computed tomography (CT) were retrospectively evaluated. In addition, CMR and CT images were reviewed for PAPVC.
Results: Seventy-eight Turner were included in this study, 52 adults and 26 children (mean age 25y, range 5 – 67y). Sixty-five underwent echocardiography and 41 CMR. CT was performed in only 3 patients. CVD were present in 45% of patients and more frequently found in karyotype 45X (81%), followed by 45,X/46,XX (75%) vs. 45,X/47,XXX (25%) or 46,Xi (40%, p = 0.002). Congenital heart disease (CHD) lesions were the most frequently encountered pathologies, in decreasing order of prevalence: bicuspid aortic valve (BAV, N = 16, 20.5%), PAPVC (N = 6, 7.7%), aortic coarctation (N = 4, 5.1%). A prolonged QT interval was observed in 4 patients (5.1%).
Only one (1.2% of total population) out of six patients with PAPVC underwent surgical repair. Eight (10.2% of total population) further patients underwent the following procedures: aortic coarctation repair (4, 5.1%), aortic dissection repair, arterial switch procedure for transposition of great arteries, Ross operation and elective ascending aortic replacement (1, 1.2%; respectively).
Conclusion: Almost half of TS patients present CVD which appear to depend on the underlying karyotype. CHD is the most frequent CVD with BAV being the most prevalent followed by PAPVC, aortic coarctation and a prolonged QT interval. The impact of PAPVC seems to be of minor importance, rarely requiring surgical repair unlike e.g aortic coarctation. These data should be confirmed in a larger, multicenter study.

Keywords: Turner Syndrome, cardiovascular disorders, partial anomalous pulmonary venous connection, congenital heart disease

P-021
Outcomes and prognosis of total cavopulmonary connection performed at adult age

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Background and Aim: The aim of this study was to investigate the clinical features, assess the clinical course and outcomes, and identify predictive factors for mortality in patients undergoing total cavopulmonary connection (TCPC) at adulthood.
Methods: Single center retrospective analysis of all patients who underwent TCPC at 18 years of age or older, between 1990 and 2015. Patients were classified according to a previous atriopulmonary connection (TCPC-Conv group), previous palliative surgery (TCPC-Pall group) or absence of previous surgery or bidirectional Glenn anastomosis only (TCPC-Nopall group). Vital status, clinical events during follow up, functional status at last medical visit and quality of life were recorded.
Results: Thirty five adult patients underwent total cavo-pulmonary connection, 19 in TCPC-Pall group, 7 in TCPC-NoPall group, 9 in TCPC-Conv group. Median follow up was 7 ± 6,7 years. Actual survival was 80, 73,2 and 68,3% at respectively 1, 5 and 10 years. There was no significant difference in mortality between surgical groups (22% in TCPC-Conv, 14% in TCPC-NoPall, 42% in TCPC-Pall, p = 0,3). Early mortality was 17%. Mortality was significantly higher in patients in which a fenestration was performed at the time of surgery (100% vs. 22,5%, p = 0,005) and albumin

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blood levels at last follow up was significantly lower among deceased patients (mean 31.3 g/l versus 41.9 g/l, p = 0.04). TCPC resulted in mean NYHA improvement in every groups (TCPC-Conv: 2,5 ± 0,5 vs. 1,5 ± 0,8 p = 0,01; TCPC-NoPall: 2,7 ± 0,7 vs. 1,7 ± 0,9 p = 0,004; TCPC-Pall: 2,6 ± 0,6 vs. 1,9 ± 0,8 p = 0,02). The frequency of arrhythmia episodes decreased from 60.6% to 22.2% at latest follow-up (p = 0.5), but preoperative PLE in conversion group did not resolved.
Conclusion: Total cavo-pulmonary connection in adults can provide a benefit in long-term survival and symptoms improvement in selected patients. However high postoperative mortality may lead to renunciation or foreseeing postoperative care in high-risk patients as fenestration did not improve outcome

Keywords: Adult Congenital Heart Disease, total cavopulmonary anastomosis, outcomes

P-022
Nutritional assessment and associated factors in children with congenital heart disease - ethiopia

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Background and Aim: Worldwide, congenital heart disease is the principal heart diseases in children and constitutes one of the major causes of infant mortality, particularly in developing countries. Infants and children with congenital heart disease exhibit a range of delays in weight gain and growth. In some instances, the delay can be relatively mild, whereas in other cases, cause the failure to thrive.
The objectives of this study are to determine the nutritional status and associated factors of pediatric patients with congenital heart disease.
Methods: A cross sectional analytical study was done over a period of 6months. A total of 228 subjects with congenital heart disease that come to the cardiac center during the study period where included until the calculated sample size was attained. Data was collected from patient card and care givers of the children included in the study after obtaining their informed consent using data inquiry sheet. Data was then analyzed using Statistical Package for Social Sciences (SPSS) for windows version 25.0. Data is presented using tables and text form. Odds Ratio with 95% Confidence Interval (CI) was used to determine the effect of the independent variables on the outcome variable and P-value less than 0.05 was considered statistically significant.
Results: A total of 228 children from age 3month to 17yrs with mean age of 4.7 years (SD = 3.8 years) were included in the study. Most of the subjects had acyanotic heart disease accounting for 87.7%. The overall prevalence of wasting, underweight and stunting were 41.3%, 49.1% and 43% respectively. Among these children those with pulmonary arterial hypertension were found more likely have wasting compared to those without; an odds of 1.9 (95% CI: 1.0-3.4) and also greater chance of being stunted with an odds of 1.9 (95% CI: 1.0-3.4). children above 5years of age were 2.3 times more likely to be underweight.
Conclusion: Malnutrition is a major problem of patients with CHD. Pulmonary hypertension and older age are associated with increased risk of undernutrition.

Keywords: congenital heart disease, Pulmonary hypertension, underweight, wasting and stunting.

Fetal Cardiology

P-024
Prediction of postnatal mortality in prenatal echocardiography in fetuses with ebstein anomaly

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Background and Aim: We reviewed our institution's experience with prenatal diagnosis of Ebstein anomaly to identify prenatal predictors of postnatal mortality.
Methods: We analyzed fetal echocardiograms and clinical histories of 40 patients with a prenatal diagnosis of Ebstein anomaly at our institution from 2012 to 2020. We compared prenatal echocardiographic parameters in patients of two groups – survivors (including follow-up) and nonsurvivors.
Results: Of the 40 pregnancies, 8 (20%) were terminated, 3 (7,5%) ended with intrauterine fetal demise, 29 (72,5%) resulted in a live birth. The median of birth term was 38 weeks (range 35–40). Vaginal delivery occured in 4 (31%) and caesarean section in 9 (69%) cases. The mean birth weight was 3,1 ± 0,5 kg, height 51 ± 2. Seven (24,1%) of the 29 patients received surgical treatment. Mortality among live births was 34,5 percent (n = 10): 60% (n = 6) of children died before 1 month, 20% (n = 2) before 6 months and 20% (n = 2) after one year of life. Mortality in operated patients was 42,9% (3 out of 7 patients), in non-operated patients 31,8% (7 out of 22 patients). In both groups (deceased and survivors), a retrospective analysis of prenatal echocardiographic parameters was carried out. In all fetuses main dimensions of heart structures have been measured and Z-score was calculated for each of them. In our study there were no statistically significant difference of left and right ventricles end-diastolic diameters, the mitral valve, the aortic valve diameters, the ascending aortic diameter, diameters of the valve and the main of pulmonary artery size between the groups. The groups were also compared for the degree of tricuspid insufficiency, the degree of cardiomegaly, the degree of fetal heart failure, the presence of fetal hydrops, hemodynamic atresia of the pulmonary artery, circulatory shunt. There was a statistically significant difference in the group of nonsurvivors in the severe tricuspid regurgitation (P = 0,006) and dilated (Z-score >2) of the diameter of the anatomical ring of the tricuspid valve (P = 0,004).
Conclusion: Severe tricuspid valve regurgitation and dilated anatomic tricuspid valve ring (Z-score >2) in the fetus associated with mortality after birth.

Keywords: Congenital heart defects, prenatal diagnosis, echocardiography, Ebstein anomaly, tricuspid regurgitation.

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P-025

The role of genetics in fetal bradycardia

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Background and Aim: Fetal bradycardia can have different acquired causes in obstetrical care. However, there are also congenital diseases in which fetal sinus bradycardia can be the first and only prenatal manifestation. We present an overview on different genetic disorders that should be taken into consideration in case of diagnosed fetal sinus bradycardia.
Methods: Medical records of cases with fetal sinus bradycardia and positive genetic results referred to our institutions were evaluated. A literature review was done using a Pubmed- and OMIM-based search for monogenetic disorders causing fetal sinus bradycardia.
Results: We identified four cases of fetal sinus bradycardia in which a genetic cause was uncovered. Underlying monogenetic disorders were associated with variants in HCN4 (two cases) and TBX5 (one case). In one case, the prenatal phenotype might be explained by two polymorphisms in SCN5A. Literature review identified nine monogenic diseases that could lead to fetal sinus bradycardia. Four of these disorders can be associated with extracardiac findings.
Conclusion: Genetic testing should be considered in cases with fetal sinus bradycardia, especially in cases of additional extracardiac findings. Broad sequencing techniques as well as improved prenatal phenotyping could help to establish a diagnosis in an increasing number of cases.

Keywords: fetal, bradycardia, genetics

P-026

Abnormal ratio of three-vessel view in fetuses with right aortic arch

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Background and Aim: The prenatal detection rate of a right aortic arch (RAA) has increased after the introduction of the three-vessel view (3VV) formed by the vena cava superior (VCS), the aorta (Ao) and the pulmonary artery (PA), and three-vessel trachea view (3VTV). However, the 3VTV is not mandatory in all prenatal screening programs and imaging can be difficult for unexperienced sonographers. Therefore, this study aims to examine the value of an additional marker in the 3VV plane, namely the "interspace" between the Ao and PA in cases with RAA.
Methods: This is a case-control study performed in the fetal medicine unit of the Amsterdam UMC. Fetuses with RAA without major intra-cardiac anomalies or associated extra-cardiac anomalies were included and 3 controls per case were matched for maternal BMI, gestational age and year of diagnosis. The 3VV-images were collected, distances between the VCS, Ao and PA were measured and ratios were calculated between the distances. Absolute measurements and ratios were compared between cases and controls. Subsequently, a cut-off point of the ratios were calculated by a ROC-curve.
Results: We included 37 cases with RAA and 111 healthy controls. The mean absolute distance between PA and Ao was 3.2mm in cases and 1.9mm in controls (p<.001). The mean ratio between PA and Ao distance and Ao and VCS distance (PAAo/AoVCS) was 3.1 in cases and 1.4 in controls (p<.001). The ROC-curve of the ratio PAAo/AoVCS showed a cut-off point of 1.8 at the upper left corner with a sensitivity of 92% and specificity of 85%.
Conclusion: The distance between PA and Ao – also "the interspace of the 3VV" – and the ratio between PA and Ao distance and Ao and VCS distance (PAAo/AoVCS ratio) were significantly bigger for cases with RAA as compared to controls. This study concludes that the distances between vessels in 3VV could be added as an additional marker to prenatal screening. If an interspace between PA and Ao is noticed by eyeballing, it is recommended to use the PAAo/AoVCS ratio with a cut-off point of 1.8 to assist in the diagnosis of RAA.

Keywords: prenatal screening, fetal echocardiography, three-vessel view, right aortic arch

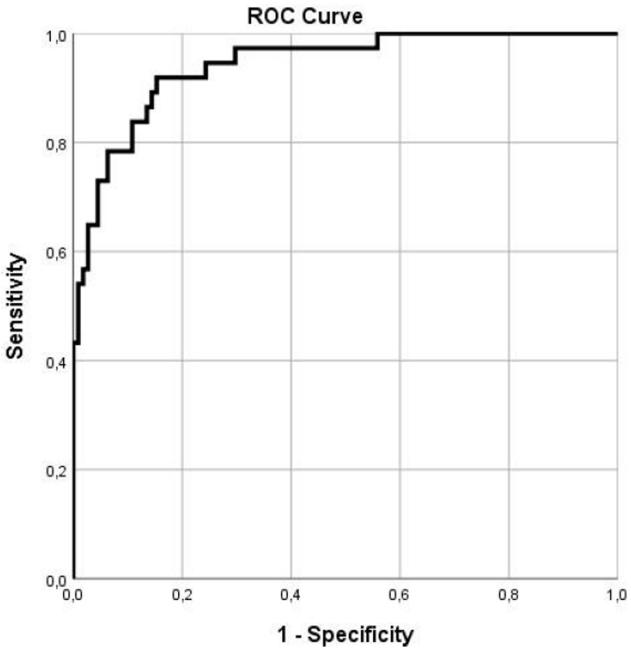

Figure 1: ROC-curve of the ratio PAAo/AoVCS

P-027

Tetralogy of fallot in premature neonates: risks and outcomes

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Background and Aim: Premature infants with tetralogy of Fallot (pToF) have up to 13-fold increase in mortality, but the evolution and outcome of multi-level right ventricular outflow tract obstruction (RVOT) in pToF is unclear. We sought to evaluate clinical

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and imaging features and correlate with mortality and type of intervention [valve-sparing (VS) vs transannular patch (TAP)]
Methods: A retrospective review of pTOF, born at gestational age < 37 weeks between Jan 2010 and Dec 2020 and admitted to our tertiary hospital identified from institutional databases was completed. Infants with pulmonary atresia or nonconfluent pulmonary arteries were excluded. Demographic, echocardiographic, catheterization and surgical data were compiled and reviewed.
Results: 24 pTOF met inclusion criteria ranging from 24 - 36 6/7 weeks gestation and birthweight (BW) ranging from 0.35 – 2.8 kg. Additional extracardiac concerns were present in 62%. Intervention was performed at an average of 3.3 months. Cyanotic spells were observed in 10 pTOF, all were started on β-blocker therapy and all had significant increase in RVOT obstruction leading to repair at a mean postnatal age of 3.5 months, including 1 who underwent emergent catheterization. TAP was needed in 60% of this group. 14 pTOF did not have any documented cyanotic spells and TAP was need in 54% of those patients.
In pTOF with BW <1.5 kg, only 5/9 survived to complete repair and only 1 was VS. Mortality was 17% (n = 4); all in pTOF with BW < 1 kg (average 585 g) and all had intra-uterine growth restriction (IUGR), were delivered by urgent C-Section and died before surgical intervention, although 1 received an RVOT stent. Of the surviving cohort, 20% suffered adverse events (2 pericardial effusion, 1 pleural effusion, 1 esophageal injury).
Conclusion: In our cohort, no IUGR pTOF with BW < 1kg survived to surgery. Despite rapid increase in dynamic RVOT obstruction in pTOF with cyanotic spells, their age at repair and feasibility for VS surgery was equivalent to the group that did not have any reported cyanotic spells. pTOF without cyanotic spells were equally likely to need TAP even though they had no dramatic increase in RVOT obstruction.

Keywords: Tetralogy of Fallot, Premature infants, congenital heart disease, intra-uterine growth restriction (IUGR)

P-028
Prenatal and postnatal management of foetal supraventricular tachycardia over two decades at a tertiary care centre

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Background and Aim: Foetal arrhythmia is detected in approximately 1% of all pregnancies. Foetal supraventricular tachycardia (SVT) can be associated with significant morbidity and mortality. Despite this, significant variation exists in treatment of SVT between some centres. This study aims to describe the current approach to management of foetal SVT in a tertiary referral centre, and postnatal follow up including requirement for antiarrhythmic treatment and occurrence of postnatal SVT.
METHOD:A retrospective cohort study was carried out on foetuses diagnosed with SVT at a single centre, between January 2010 and December 2020. Patients were identified from a database of foetal arrhythmia and included those referred from peripheral centres. Clinical review of both maternal and foetal medical notes, and echocardiograms, was performed.
Results: Thirty one foetuses were identified for inclusion, 19 (61%) of whom were male. Mean gestational age at diagnosis was 28.5 weeks (SD 5 weeks). Hydrops foetalis occurred in 8 cases and single compartment fluid accumulation occurred in 7 cases. All received antenatal treatment with antiarrhythmic medication. Flecainide was the most common first-line antiarrhythmic medication with 13 cases (42%) requiring another antiarrhythmic therapy (digoxin, sotalol or amiodarone). Outcomes included two intrauterine deaths related to SVT and one case of maternal mirror syndrome. Overall survival was 90%. Twenty three of 28 infants were in normal sinus rhythm at birth, and 12 were transferred directly to a paediatric cardiology centre. Eighteen infants were treated postnatally with antiarrhythmics for a median of 12 months (IQR 5 months). Postnatal SVT recurred in 10 cases, 70% of which occurred within the first three months of life. The median duration of follow up was 19 months (IQR 41 months).
Conclusion: Foetal supraventricular tachycardia can be successfully managed with antiarrhythmic medication. However different drug therapies are frequently required, necessitating an individualised approach to treatment. Multidisciplinary input from foetal medicine specialists as well as paediatric cardiologists optimises management of foetal arrhythmia. Further multicentre studies may optimise medical management of foetal supraventricular tachycardia.

Keywords: Foetal supraventrcular tachycardia, anti-arrhythmic treatment, hydrops

P-029
Speckle-tracking analysis of fetal left ventricular function in shone syndrome

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Background and Aim: Shone Syndrome is a congenital heart disease characterized by sequential obstructions of left ventricular (LV) inflow and outflow. It can require multiple interventions with a poor long-term prognosis. Prenatal diagnosis is challenging and diagnostic suspicion is raised for the presence of borderline left ventricle with small aortic annulus, similarly to aortic coarctation (CoA). Recent studies have demonstrated LV contractility depression in fetuses with CoA, but assessment of LV function in those with Shone Syndrome has not yet been conducted.
The aim of this study is to assess LV function in fetuses with postnatally confirmed diagnosis of Shone Syndrome using 2D speckle-tracking echocardiography (2D-STE) comparing with fetuses with postnatally confirmed simple aortic coarctation (S-CoA) and those with false-positive aortic coarctation (FP-CoA).
Methods: From January 2010 to December 2019 we retrospectively analyzed the four-chamber view (4CV) of 73 consecutive fetuses referred for suspected CoA and 30 control cases. 2D-STE was used to calculate LV global longitudinal strain (GLS), global circumferential strain (GCS), strain rate (SR) and ejection fraction (EF). Those data were correlated to mitral valve diameter/tricuspid valve diameter ratio (MV/TV).
Results: Postnatally Shone Syndrome was diagnosed in 15 fetuses (20.5%), CoA in 24 fetuses (33%) and FP-CoA in 34 fetuses (46.5%). In fetuses with Shone Syndrome GLS (mean -13.2%, SD 2.2, $p < 0.001$), SR (mean -1.1 s^{-1}, SD 0.15, $p < 0.001$) and EF (mean 47.1%, SD 5.1, $p = 0.03$) were significantly reduced compared to fetuses with S-CoA. FP-CoA showed normal LV function parameters but GLS that was significantly reduced compared to controls ($p = 0.01$). Univariate linear regression showed positive correlation between MV/TV and GLS ($p = 0.02$, R2 0.3) in Shone Syndrome fetuses.
Conclusion: Fetuses with post-natal diagnosis of Shone Syndrome showed markedly depressed LV function compared to S-CoA fetuses. We speculated that this could be related to the same genetic messages causing the anatomical abnormalities. To our knowledge this is the first study describing significant LV

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dysfunction in fetuses with post-natal diagnosis of Shone Syndrome. These data may improve the parental counselling, being the post-operative outcome in patients with Shone Syndrome significantly more severe than in those with S-CoA.

Keywords: strain, aortic coarctation, fetal ventricular size discrepancy, shone, GLS

P-030

Prenatal detection rate of major congenital heart disease in switzerland

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Background and Aim: Detection of major congenital heart disease (MCHD) is an important aim of second trimester screening for improving parental counseling and perinatal treatment. Prenatal detection rate of MCHD in Switzerland is unknown. We sought to analyse the prenatal detection rate and outcome of MHCD of the referral region of the largest pediatric heart centre in Switzerland.
Methods: Definition of MCHD was defined as any structural cardiac malformation diagnosed in the first 6 months of life and requiring invasive therapy, i.e. cardiac surgery and/or catheter-guided intervention. The cases were retrospectively identified by reviewing all echocardiographic reports performed at the age <6 months between 2012 and2017 in our institution. All cases were cross-checked with the fetal echocardiographic database, if diagnosis was already done prenatally. In addition, all fetal cases expected to be delivered in the same period were matched to the identified cases.
Results: A total of 643 patients with MCHD were identified. Overall prenatal detection rate was 26.5% (n = 170). Distribution of cases (n, % of total; % of prenatal detection) was large ventricular septal defect (VSD) (n = 108, 17%; 10%), coarctation (n = 86, 13%; 21%), single ventricle (n = 62, 10%; 60%), atrioventricular septal defect (n = 58, 9%; 33%), d-transposition of the great arteries (TGA) (n = 48, 7%; 23%); tetralogy of Fallot (n = 47, 7%; 19%), pulmonary arterial stenosis (n = 43, 7%; 9%), double outlet right ventricle (n = 35, 5%; 31%), aortic stenosis (n = 27, 4%; 19%), pulmonary atresia (PA) with VSD (n = 25, 4%; 36%), total pulmonary venous return (n = 15, 2%; 13%), PA/intact ventricular septum (n = 9, 1%; 56%), common arterial trunk (n = 6, 1%; 50%), Ebstein's anomaly (n = 6, 1%; 83%), congenitally corrected TGA (n = 6, 1%; 50%) and others (n = 62, 10%; 26%). In summary, detection rate in single ventricle was relatively high. Detection rate in critical heart lesions such as coarctation and d-TGA remains below 25%.
Conclusion: Prenatal diagnosis of MCHD for distinct cardiac lesions remains low. Efforts to improve detection rate for critical lesions such as d-TGA and aortic coarctation are crucially required.

Keywords: prenatal detection rate

P-031

Amniotic fluid levels of nt-probnp are a novel biomarker of heart failure in neonates and infants

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Background and Aim: Recent advances in fetal echocardiography technology and its widespread use have made it possible to evaluate fetal cardiac function and heart failure more accurately, although there is a paucity of clinically useful biomarkers for the diagnosis of heart failure in neonates. N-terminal prohormone of brain natriuretic peptide (NT-proBNP) in blood is one of the established biomarkers for heart failure and is known to be reduced exponentially after birth in children born full-term but not in premature infants. Nearly all (95%) NT-proBNP in the blood is excreted in urine. All amniotic fluid is replaced by fetal urine after the second trimester. The aim of this study was to investigate whether amniotic fluid NT-proBNP could be a potentially useful biomarker to assess severity of heart failure in neonates.
Methods: We conducted a retrospective cohort study of 84 neonates and infants, including 11 congenital heart disease patients not affected by chromosomal anomalies. We collected peripheral blood, umbilical vein blood, urine, and amniotic fluid to assess: 1) baseline levels of serum NT-proBNP (sNT/cre) in premature infants and concentration changes overtime; 2) urine NT-proBNP levels (uNT/cre) and the correlation between uNT/cre and sNT/cre; 3) NT-proBNP in amniotic fluid (aNT/cre) and umbilical vein blood (uvNT/cre) and examined possible correlations among sNT/cre, uvNT/cre, aNT/cre and uNT/cre.
Results: Average gestational age for the subjects was 28.1 ± 3.8 weeks and average birth weight were 1096 ± 539.6g Our data revealed that sNT/cre levels in neonates decrease exponentially over time after birth similar to those of full-term infants. We also found a weak ($r = 0.453$) but statistically significant positive correlation ($p = 0.003$, after the data were logarithmically transformed) between sNT/cre and uNT/cre. Importantly, our results showed that there was a very strong correlation between sNT/cre and uvNT/cre ($r = 0.992$, $p<0.001$) as well as between uvNT/cre and aNT ($r = 0.903$, $p<0.005$), respectively, in neonates.
Conclusion: Our data demonstrated that aNT/cre could be a useful biomarker for diagnosing heart failure in neonates/fetuses after the third trimester.

Keywords: NT-proBNP, heart failure, biomarker, infant, amniotic fluid, fetal echocardiology

P-032

A case series of persistent left and absent right superior vena cava with prenatal diagnosis

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Background and Aim: Persistent left superior vena cava (PLSVC) is the most common systemic venous variant. However, isolated PLSVC with absent right superior vena cava (ARSVC) only occurs in 0.09 - 0.13% of patients, with 46% associated congenital heart defects and 36% rhythm events. Spread of protocolized fetal echocardiography and three vessel-trachea view allows prenatal diagnosis of these venous asymptomatic anomalies. We therefore present our case series of PLSVC and ARSVC with prenatal diagnosis, the largest to our knowledge, wishing to describe main demographic and clinical characteristics and postnatal course.
Methods: Retrospective case series including all reports of PLSVC and ARSVC with prenatal diagnosis. Patients and data were collected from one tertiary centre computerized database. This tertiary centre receives patients from the whole country. Diagnosis was made by a Pediatric Cardiologist, by an extensive cardiovascular examination including three vessel-trachea view

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in an independent consultation. Follow-up was made in a Pediatric Cardiology consultation in our centre or in those centres where patients were referred from.
Results: Twelve fetuses were prenatally diagnosed with PLSVC and ARSVC, 33.33% were male with gestational age at diagnosis 29.36 (22.29 – 33.43) weeks. Fetal echocardiography showed 5 ventricular or great vessels discrepancy, 3 ventricular septal defects and 1 mesocardia associated, and 7/12 (58.3%) isolated PLSVC and ARSVC. No complex cardiopathies were found. In the series, 3/12 cases underwent karyotype with 1/3 inherited partial trisomy of 9th chromosome (dysmorphic phenotype, epileptic encephalopathy, failure to thrive), other 2 were normal. Delivery and perinatal care were influenced by the cardiopathy in none of the cases. Patients were followed-up during 23.5 (8.5 - 67) months. All of them were asymptomatic, presenting 1/12 dysplastic aortic valve with mild regurgitation, 1/12 minimal patent ductus arteriosus, 1/12 patent foramen ovale and 1/12 mesocardia (cavities discrepancy and septal defects described were spontaneously resolved during follow-up). All of them had normal electrocardiograms, with a Holter monitoring included in one patient.
Conclusion: Spread and standardisation of fetal echocardiographic studies have allowed the diagnosis of anomalies otherwise asymptomatic and missed. Therefore, PLSVC and ARSVC could be a less rare anomaly and in more cases isolated than previously described.

Keywords: persistent left superior vena cava, absent right superior vena cava, fetal echocardiography

P-033
Diagnosis of severe congenital heart defects in 2 consecutive periods: increased prenatal detection rate but similar mortality rate

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Background and Aim: To determine the actual prenatal detection and mortality rate in patients with severe congenital heart diseases (CHD) and to compare these data with an earlier study period (2006-2014).
Methods: Single center retrospective study in patients with severe CHD diagnosed prenatally or postnatally between 2015 and 2020.
Results: A total of 379 patients were included. Univentricular heart (20%), coarctation of the aorta (18%), transposition of the great arteries (17%) and tetralogy of Fallot (15%) were the most prevalent. 194 diagnoses (52%) were made prenatally, with large differences among CHD types. Prenatal detection rate was the highest for univentriculair heart defects (85%) and the lowest for coarctation (15%).
Overall mortality rate was 30% mostly due to prenatal mortality including termination of pregnancy (TOP; 48%) and postnatal compassionate care (17%). In the group who underwent surgery, mortality rate was 8% (<30d postop: 5%). Postnatal mortality was higher in patients born after prenatal diagnosis than in those with a postnatal diagnosis (32% vs 6%; $p<0.001$).
Compared to the earlier study period, the prenatal detection ratio increased from 30% to 52% ($p<0.001$) with a marked improvement in the prenatal detection of tetralogy of Fallot (23% vs 40%) and transposition of the great arteries (24% vs 45%). Mortality rate remained similar both for overall and postnatal mortality. In the prenatal diagnosis group, TOP rate dropped from 41 to 29% ($p = 0.001$).
A molecular diagnosis was found in 18% of the patients that were genetically tested ($N = 296$). In patients born alive, additional abnormalities were found in the gastro-intestinal tract (4%), the airway tract (2%), the renal system (6%) or the nervous system with or without neurodevelopmental problems (11%).
Conclusion: Prenatal diagnosis of CHD has improved significantly in the last years. Despite this improvement, overall mortality remained similar with major contributors still being TOP and compassionate care. Genetic abnormalities and extra-cardiac pathology were present in an important amount of patients indicating that prenatal and postnatal counseling should include the possibility of having an underlying genetic disorder or associated extra-cardiac defects which could impact the overall outcome

Keywords: prenatal detection rate; mortality rate; prenatal diagnosis; outcome

P-034
Congenitally corrected transposition of the great arteries: a single centre experience of clinical findings and long-term outcomes

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Background and Aim: There are scarce data on the antenatal detection rates, foetal and postnatal outcomes of congenitally corrected transposition of the great arteries (CCGTA). This paper aims to provide data on the survival and long-term outcomes of patients with CCTGA in both antenatal and postnatal periods.
Methods: A retrospective cohort study of 59 patients diagnosed with CCTGA between 1966 and 2021. Information was collected and analysed on diagnosis, management, and prognosis of patients diagnosed with CCTGA at the University Hospital of Wales (UHW)in South Wales.
Results: 20 cases (36%) were diagnosed antenatally, with an increase from 6% pre-2000 to 81% post-2000 and 100% since 2020. 50 patients (91%) showed other cardiac lesions, with no non-cardiac or genetic associations. Q wave abnormalities on ECG, tricuspid regurgitation and reduced ejection fraction on echocardiography were the most common findings. However, exercise tests, Holter and additional imaging modalities were underutilised. There was a survival probability of 0.83 at 50 years, but freedom from heart block or surgery were 0.5 and 0.38 respectively. Pulmonary artery banding surpassed Blalock-Taussig shunt and double switch as preferred surgery, and pacemaker insertion decreased post 2000. ACE inhibitors were utilised in 60% of the cases. At latest follow up, 50 patients (91%) are in NYHA stage 1 or 2, albeit only 31 (56%) remained free from heart block.
Conclusion: CCTGA has high association with additional cardiac lesions and complications, but no connection with genetic abnormalities. Improved diagnostic process post 2000 has led to

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increased antenatal detection and improved postnatal status. Pulmonary artery banding is the most common intervention in infancy. The survival rate in this cohort is high exceeding the previous literature.

Keywords: congenitally corrected transposition, surgery, outcome, fetus

P-035

Congenital heart defects in the fetus, classifications and embryology: cladistics or phenetics?

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Background and Aim: Congenital heart defects (CHD) are a common cause of fetal death and termination of pregnancy for fetal anomaly (TPFA). Large series describing detailed anatomy of fetal cardiac specimen are lacking.

The aim was to phenotype all fetal cardiac specimens of our institution's collection, and to create a user-friendly database including photographs.

Methods: A complete morphological examination of each specimen according to segmental analysis was performed by two observers (MH, LH). The main CHD was determined according to the 11 categories and 23 subcategories of the clinical and anatomical classification of CHD, and the associated lesions were coded using IPCCC ICD-11 Nomenclature. All codes and photographs of each heart specimen were linked into the database.

Results: Among 1236 cardiac specimens, 120 were normal hearts (10%). The three main groups of CHD were: anomalies of the ventricular outflow tracts (VOT) (354, 32%), functionally univentricular hearts (258, 23%), and anomalies of atrioventricular junctions and valves (153, 14%). The most frequent segmental anatomical lesions were: valvar anomalies (all valve types, 677, 55%), ventricular septal defects (VSD, 488, 40%), ventricular hypoplasia (433, 35%), interatrial communications (428, 35%). Results of different analyses of segmental cardiac characteristics will be presented. The cladistic grouping can be superimposed on the anatomical classification to identify similar characteristics and derive embryological hypotheses. The most common example is the term "conotruncal" anomaly grouping defects supposed to have the same embryological origin/"common ancestor". The phenetic approach can explore how a segmental character is distributed in association with other segmental defects. For example, common arterial trunk can be observed in association with outlet VSD in the majority of cases but this segmental character can also be associated with intact interventricular septum, hypoplastic ventricle, or with right aortic arch. Clustering segmental characters of the heart without a priori hypotheses on development may help identifying new mechanisms of altered cardiac development.

Conclusion: This anatomical phenotyping of the fetal collection should allow us to identify rare associations of malformations. Having all details of cardiac anatomy can help elaborating different approaches of describing and grouping CHDs.

Keywords: foetal congenital heart defects, embryological anatomical classification, phenetic, cladistic

P-036

Ventricular ectopics in the context of fetal heart block needing salbutamol treatment

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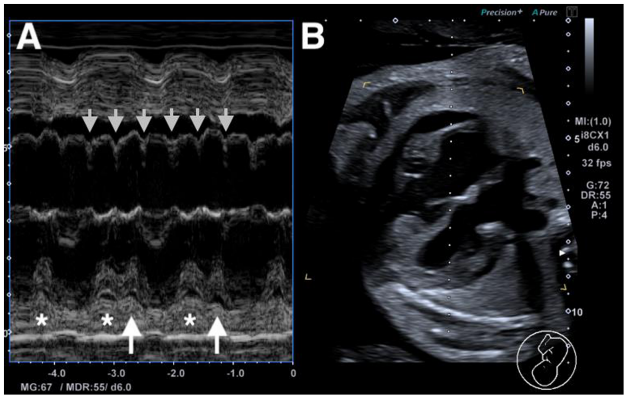
Background and Aim: Congenital complete heart block (CCHB) is a rare entity associated with maternal autoantibodies. Salbutamol has previously been used to improve the ventricular rate and cardiac function. Ventricular ectopics (VEs) are occasionally present during exercise in affected patients but have not been described in fetuses. We present a fetal case with CCHB treated with salbutamol that developed VEs late in pregnancy.

Methods: 29-year-old woman with Sjogren syndrome and rheumatoid arthritis, positive anti-Ro/La antibodies, on hydroxychloroquine and aspirin. At 18 weeks gestational age, the fetal heart rate (FHR) and AV interval were 146 beats per minute (bpm) and 114 ms. At 20+4, isolated supraventricular ectopics were seen. At 21+4, CHB was diagnosed, with a ventricular scape of 57 bpm. A small pericardial effusion and echobright myocardium were noted, with no fetal hydrops. In following appointments, the FHR continued to reduce and the pericardial effusion increased. At 31+5, FHR decreased to 51bpm and salbutamol was started at 5 mg three times daily (TDS), FHR improved to 55 bpm. The dose was further increased to 8 mg TDS, FHR improved up to 60. The pericardial effusion remained stable with no hydrops. Maternal side effects (sinus tachycardia, tremors and dizziness) led to a reduction to 6 mg TDS. At 32+5, VEs were noted in the fetal echo, salbutamol was decreased to 4 mg TDS. At 35 weeks, VEs were seen again, salbutamol was reduced to 2 mg TDS.FHR dropped to 52bpm in subsequent appointments, so the dose was adjusted to two doses of 2 mg and one of 4 mg per day.

Results: He was delivered at 36+5 weeks via caesarean section in view of a large pericardial effusion and reduction in FHR to 47bpm. He was born in good condition and was discharged home two weeks later. He is currently 5 months old, clinically stable, with no pacemaker.

Conclusion: Salbutamol can increase the heart rate in the fetus affected with CHB, decreasing the pericardial effusion size and and allowing delivery at a more mature gestation. Its association with VEs in the fetus and the impact of their presence is uncertain but should be a call for caution.

Keywords: Fetal complete heart block, anti-Ro/La antibodies, ventricular ectopics



MMode across fetal RA and LV free wall (A) and still image showing cut plane (B). Note regular atrial beats (grey arrows) at the top of the image and ventricular contractions (*) with ectopics (white arrows). A small pericardial effusion is present.

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P-037/Moderated Poster

Foetal tricuspid valve agenesis/atresia: testing predictions of the embryonic aetiology

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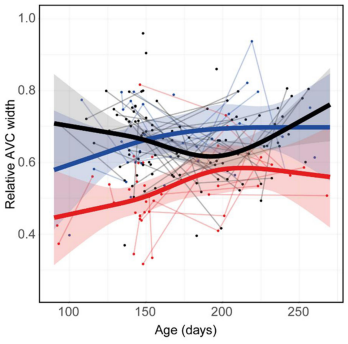
Background and Aim: Tricuspid valve agenesis/atresia (TVA) is a congenital cardiac malformation where the tricuspid valve is not formed. It is hypothesised that TVA results from a failure of the normal right-ward expansion of the atrioventricular canal (AVC). We tested predictions of this hypothesis by morphometric analyses of the AVC in foetal hearts.
Methods: We used high-resolution MRI and ultrasonography on a post mortem foetal heart with TVA and with tricuspid valve stenosis (TVS) to validate the position of measurement-landmarks that were to be applied to clinical echocardiograms. This revealed a much deeper right atrioventricular sulcus in TVA than in TVS. Subsequently, serial echocardiograms of in utero foetuses between 12 and 38 weeks of gestation were included (n = 23 TVA, n = 16 TVS and n = 74 controls) to establish changes in AVC width and ventricular dimensions over time.
Results: Ventricular length and width and estimated foetal weight all increased significantly with age, irrespective of diagnosis. Heart rate did not differ between groups. However, in the second trimester, in TVA, the ratio of AVC-to ventricular width was significantly lower compared to TVS and controls.
Conclusion: This finding supports the hypothesis that TVA is due to a failed right-ward expansion of the AVC. Notably, we found in the third trimester that the AVC-to-ventricular width normalised in TVA foetuses as their mitral valve area was greater than in controls. Hence, TVA associates with a quantifiable underdevelopment of the AVC. This underdevelopment is obscured in the third trimester, likely because of adaptational growth that allows for increased stroke volume of the left ventricle.

Keywords: Morphometry, embryonic development, congenital malformations, fetal ultrasound, fetal echocardiography

Atrioventricular canal and ventricular width in TVA, TVS and controls



Gestational age-related changes in AVC width over total ventricular width. TVA cases are indicated in red, TVS in blue, and controls in black. Individual measurement trends are shown in the background of the average trend. The shaded areas are 95% confidence intervals.

P-038

Fetal echocardiography-based three-dimensional heart models: creating a web-based application as a new learning tool

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Background and Aim: Conventional two-dimensional (2D) ultrasound imaging is the primary screening modality used to diagnose congenital heart defects (CHD) before birth. However, the real-life heart is a complex three-dimensional (3D) structure, making prenatal assessment based on conventional ultrasound imaging extremely challenging. Currently, educational resources that directly link conventional prenatal ultrasound imaging to the real-life 3D heart are lacking. Here, we present a pilot study in which we used spatiotemporal image correlation (STIC) technology to create digital 3D fetal heart models. The aim of this pilot study is to determine the feasibility of generating ultrasound-based 3D fetal heart models for the development of web-based learning tool, which could provide this missing link.
Methods: The hospital database was retrospectively screened for high-quality STIC volumes, obtained as part of routine clinical management of fetuses with a prenatal CHD diagnosis, as well as healthy reference cases. Prenatal findings, pregnancy outcome, postnatal discourse, including postnatal cardiac imaging when available, were retrieved from patient records. Image segmentation and 3D reconstruction of the cardiac ultrasound volumes was performed blinded. Completed models were validated by a senior paediatric cardiologist, who had access to all clinical information.
Results: A total of 16 models have been generated, including four cases of normal cardiac anatomy, and 12 cases with various CHDs; atrioventricular septal defect (3), transposition of the great arteries (3), venous return anomalies (2), double outlet right ventricle (1), tetralogy of Fallot (1), critical pulmonary stenosis (1), Ebstein's anomaly (1). In all cases, the morphology of the heart and the arterial and venous connections were successfully reconstructed and validated.
Conclusion: Our results show that reconstructing the heart and major vessels for the generation of educational 3D fetal heart models is feasible, both in normal cardiac anatomy and various types of CHD. The next step will be expanding our database, and initiating the development of the learning tool that combines the generated 3D models with an interactive ultrasound interface. By developing this tool, we aim at providing the missing link between conventional fetal cardiac ultrasound imaging and the real-life 3D heart.

Keywords: prenatal diagnosis, congenital heart disease, fetal ultrasound, fetal echocardiography, education, 3D visualisation

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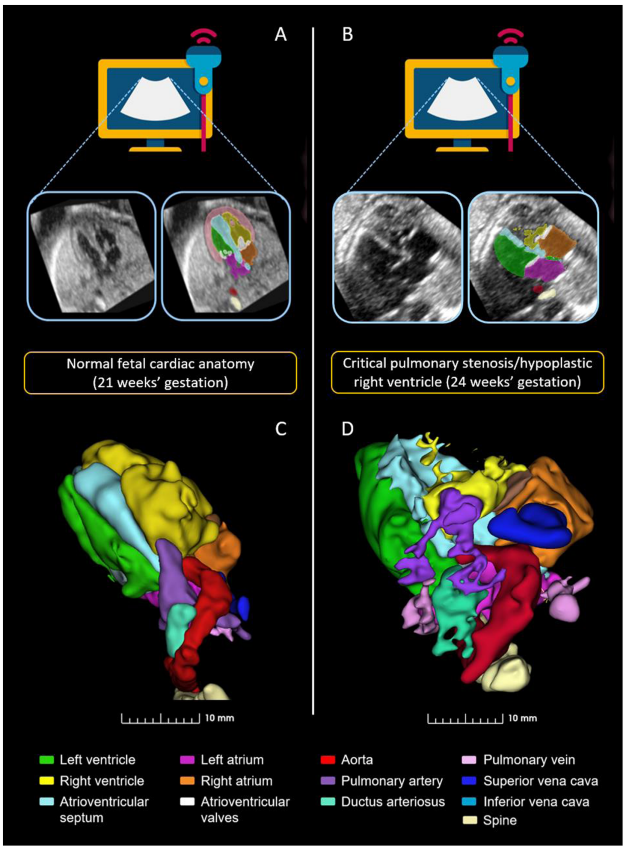
Fetal echocardiography-based three-dimensional heart models



A and B: Reconstructed four-chamber view, selected from a spatiotemporal image correlation (STIC) ultrasound volume of a 21-week old fetus with a structurally normal heart (A) and a 24-week old fetus with critical pulmonary stenosis and hypoplastic right ventricle (B). Cardiac structures and the arterial and venous connections have been segmented, as indicated by the coloured areas. C and D: Fetal echocardiography-based 3D digital heart models of a healthy 21-week old fetus (C – corresponding to panel A), and a 24-week old fetus with critical pulmonary stenosis and hypoplastic right ventricle (D – corresponding to panel B). Both models are seen from above, with the spine located posteriorly and the cardiac apex directed left-anteriorly.

P-039

ALVT diagnosis: a case report

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Background and Aim: The ALVT It is defined as a rare CHD in which where is a paravalvular communication between the aorta and the left ventricle.

Methods: CASE:

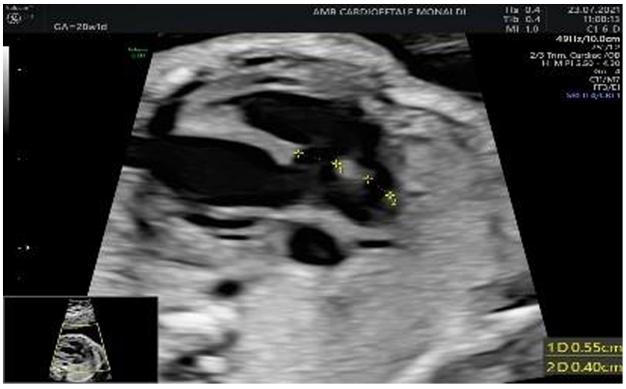
A pregnant woman of 33 year's old was invited to perform a fetal echocardiographic examination in our fetal lab, for suspected aortic stenosis at ultrasound screening scan performed at 21 weeks of gestation. Extracardiac fetal anatomy appeared normal. The fetal echocardiographic was performed at 22 weeks of gestation. The four-chamber scan appeared abnormal. The LV was dilated. In LVOT were seen subaortic and supravalvular defects. The aortic valve appeared dysplasic, Doppler color flow demostrating regurgitation around LVOT and consequent rapid run-off of blood. In right ventricular outflow tract (RVOT) the pulmonary artery appeared normal. At Doppler color flow in three vessels view (3V) the ascending aorta appeared dilated. In three vessels and trachea view (3VT) and in sagitall view of aortic arch was found regurgitation. With these elements ALVT was suspected. Follow-up every 4 weeks and there were no signs of heart failure but at 37 weeks were also clearly visualized apical trabeculae in the myocardium of the left ventricle

Results: At 39 weeks of gestation birth a male infant of 3112 g by CS. The infant was immediately transferred to our PCU and the prenatal suspect of ALVT was confirmed with a bicuspid and dysplastic aortic valve without signs of stenosis. The LV appeared spongy with presence of middle apical trabeculae and systolic function was slight reduction.Tunnel entry orifice misured 4 mm, the tunnel exit orifice misured 12 mm. Cardiac surgery was performed on day 3 of life. In median sternotomy under cardiopulmonarybypass, the ALVT was closed through its longitudinal incision, direct closure of direct communication on the ventricular side and by patching of heterologous pericardium on the aortic side, DIA closure os with patch. The post-operative course was uneventful and the infant was discharged at one month and one week.Duration of ICU and hospital stay were 6 and 36 days, respectively.

Conclusion: Fetal diagnosis of ALVT is possible but is challenging.and should be considered in any fetus with aortic regurgitation.

Keywords: Fetal diagnosis

Prenatal



P-040

First 3D acquisition of fetal cardiovascular magnetic resonance cine images: simplifying diagnosis of fetal cardiac malformation

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Background and Aim: Fetal cardiovascular magnetic resonance (CMR) imaging is a valuable tool for assessing fetal cardiovascular malformations. As fetal CMR is often used in suspected cardiac malformation with complex anatomy and physiology as a complement to ultrasound, planning of 2D image stacks of interest in the fetal heart is challenging.
Therefore, the aim of this study was to overcome these challenges by showing the feasibility of acquiring fetal 3D cine CMR with maternal free breathing for the evaluation of cardiac anatomy and function.
Methods: The Regional ethics board approved the study and the participant signed informed consent before participating. Examinations were performed using a 1.5 T MAGNETOM Aera scanner (Siemens Healthcare, Erlangen, Germany).
A 3D fetal cine CMR data set was acquired in one fetus using a radial slab-selective balanced steady-state free precession (bSSFP) cine sequence (TE/TR = 1.58/3.54ms, flip angle = 46°, voxel 1.9mm^3) with 65,500 spokes. For comparison, the standard clinical 2D Cartesian bSSFP cine sequence (TE/TR = 1.38/37.92ms, flip angle = 45°, voxel 1.4x1.4x4.0mm^3) was acquired. A Doppler Ultrasound transducer (northh medical GmbH, Hamburg, Germany) provided the fetal heart rate allowing for cardiac gating during reconstruction.
Image reconstruction of the 3D cine volume was performed in Matlab (The MathWorks, Natick, MA, USA) using the Berkeley Advanced Reconstruction Toolbox. Reconstruction as performed using 16 cores on an Intel Xeon Gold 6154 running at 3.0 GHz and took 60 minutes for eight cardiac phases. The 3D cine image was interpolated using multi-planar reformatting to obtain standardized views for evaluation of cardiovascular anatomy and function.

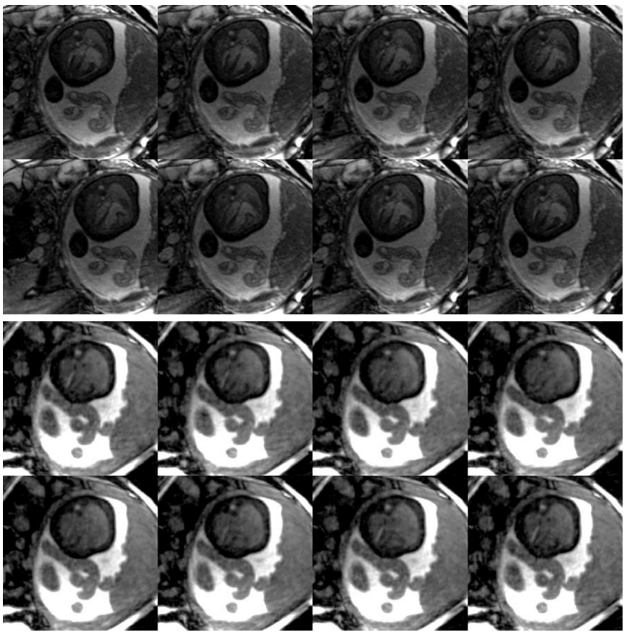

Figure 1 A fetus at 32 gestational weeks with slightly small left ventricle and aortic arch hypoplasia, but good biventricular function. Four-chamber view for the 3D cine (bottom) compared to standard clinic al 2D cine (top).

Results: Fetal images from the fetus with cardiac malformation are presented in Figure 1, which show the reconstructed eight cardiac phases for the 3D cine compared to the clinical standard 2D cine. Contrast was lower in the 3D cine images but playing the 3D in cine mode revealed similar ventricular function as 2D cine.
Conclusion: This is the first study to provide truly acquired 3D fetal cine CMR images. For clinical application, image contrast and perceived resolution need to be increased while remaining at low SAR levels.

Keywords: 3D, Fetal, Cine Imaging

P-041

Truncus arteriosus from prenatal diagnosis to clinical outcome: a single center experience

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Background and Aim: The aim of this study was to review our institution's experience with truncus arteriosus (TA) from prenatal diagnosis to clinical outcome, in order to provide more accurate prenatal counselling and improve clinical and surgical decision making in the current era.
METHOD:We conducted a single-center retrospective cohort study for the years 2005-2020. Women that visited our foetal cardiology department between January 2005 and January 2020 who were carrying a child with TA were identified through our antenatal records. Patients with TA referred by other centers were also identified from our surgical database. Demographic, echocardiographic and procedural data were collected. The perioperative course and need for reinterventions were reviewed, and clinical follow-up analysed.
Results: Overall diagnostic accuracy within our institution was 92.3%. After antenatal diagnosis, five parents (31%) decided to terminate the pregnancy. After inclusion of additional patients from referring hospitals, postnatal follow-up was available for 16 patients (median follow-up 5.4 years). All patients underwent a primary surgical correction. At our center, right ventricle to pulmonary artery (RV-PA) continuity was preferably reconstructed through direct connection (without the use of a valved conduit), which was possible in 14 patients (88%). There was no mortality during follow-up. Half of the patients were free of any reinterventions at latest follow-up, the longest freedom from reintervention was 12.5 years. At a median age of 5.5 years (range 0.2 – 12.5), 13 out of 14 patients were free from RV-PA valve implantation (figure 1). On echocardiography, the dimensions and function of the left ventricle were normal at latest follow-up (ejection fraction 66.3 ±

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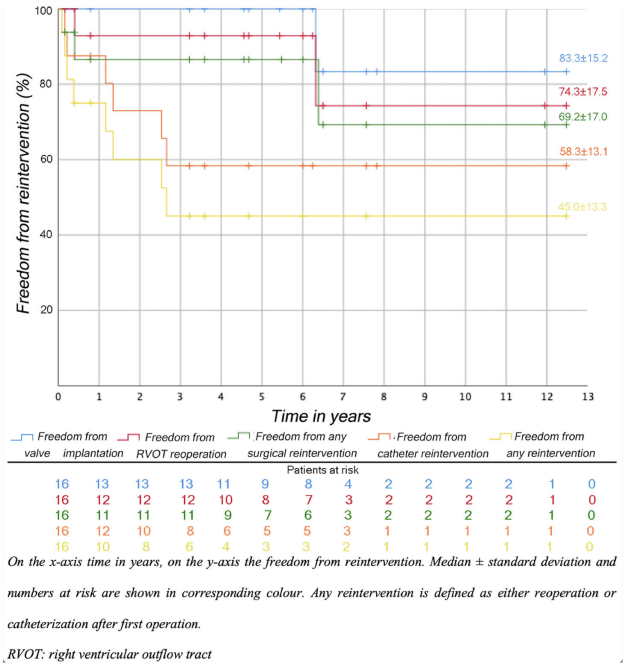
6.2%). The right ventricle demonstrated overall preserved systolic function as expressed by normal TAPSE (17.7 +/- 4.3 mm) and normal FAC (44+/-12%), except for two patients (FAC 24.2% with TAPSE 24mm; FAC 26.6% with TAPSE 13.6mm, respectively). There was no truncal valve insufficiency in 9, mild in 5, and moderate in 2 patients.
Conclusion: This study demonstrates good prenatal diagnostic accuracy of TA, no mortality in the group of operated patients, and favorable clinical outcomes at mid-term follow-up. This supports the notion that current perspectives of patients with TA are good, in contrast to the poor historic outcome series.

Keywords: Truncus Arteriosus, Echocardiography, Prenatal Detection, Direct Connection, Single Center Experience, Clinical Outcome



On the x-axis time in years, on the y-axis the freedom from reintervention. Median ± standard deviation and numbers at risk are shown in corresponding colour. Any reintervention is defined as either reoperation or catheterization after first operation.

RVOT: right ventricular outflow tract

Figure 1: Freedom from reintervention following initial truncus arteriosus repair

P-042
The fetal ductus arteriosus and its abnormalities: a single-center study

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Background and Aim: The ductus arteriosus (DA) has a vital role in the integrity of fetal circulation. Here, we present our experience in detecting DA anomalies by fetal echocardiography, the outcomes of affected patients and its association with cardiac and genetic anomalies.
Methods: The records of two thousand three hundred and sixty-six fetuses January, 2007 to January, 2021 at a single center were evaluated retrospectively.
Results: Thirty seven patients who had been diagnosed with DA anomaly prenatally and evaluated after delivery were enrolled in the study. Bilateral DA in one patient, constriction of DA in seven patients and aneurysm of DA in ten patients and right aortic arch and right DA in nine-teen patients were detected in our study. The patient with bilateral ductus had complete atrioventricular septal defect (AVSD) and right aortic arc and also mutliple phenotypic anomalies. In our constriction of DA group, three of them were resulted as intrauterin exitus (one of them was Charge syndrome) whose gestation age was younger then 30 weeks. In our aneurysm of DA group, ductus closure completed spontaneously in eight patients during first month of age without any symptoms. Constriction of DA patients' referring reasons to our clinic are mostly heart failure symptoms. According to referrals of aneurysm of DA group, only two of them referred as tortuous shaped DA and the other one referred as aneurysm of DA. Right-sided DA and right aortic arch was diagnosed in nine-teen patients. Tetralogy of Fallot were diagnosed in fifteen of them and four of those patients' intracardiac structure evaluated normal. DiGeorge Syndrome (22q11 microdeletion) was confirmed with genetic test in two cases (one for each group).
Conclusion: The ductus arteriosus holds major functional importance within the fetal circulation, and anomalies within the ductus arteriosus may interfere with the integrity of the fetal circulation and can be associated with genetic anomalies even when they are found without other congenital heart disease. We would like to highlight wide spectrum of it's anomalies due to location, shape and maintanence with our long experience.

Keywords: Fetal Ductus Arteriosus, Bilateral Ductus Arteriosus, Fetal Ductal Constriction, Fetal Ductal Closure, Fetal Ductal Aneurysm, Right Ductus Arteriosus

P-043
A rare congenital anomaly: atrial appendage aneurysm in intrauterine life and childhood

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Background and Aim: To analyze the echocardiographic and clinical features of atrial appendage aneurysm (AAA) in fetuses and children to point the importance of early diagnosis according to various potential symptoms besides asymptomatically well-prognosed patients.
Methods: The echocardiographic and clinical features of nine fetuses and three children whom diagnosed AAA in our center were analyzed including follow-up data.
Results: We report nine cases of fetal AAA diagnosed in second trimester and three incidentally found AAA at the age of six months, seven-years and eight-years. A total of twelve patients' aneurysms originated from the right, left or both atrial appendages were diagnosed in our center for a six years period. The incidence of right AAA was 73.7% (8/11) and left AAA was 27.2% (3/11)), and one biatrial AAA patient diagnosed in our center. Seven patients diagnosed with isolated AAA (58.3%). Besides, three patients diagnosed pericardial effusion (25%) and four patients combined with other intracardiac malformations (33.3%) in fetal echocardiography. In our study group, there is no evidence of cardiorespiratory system, connective tissue, and genetic diseases in family history. Fetal karyotype analysis was normal in four fetuses with late maternal age. In addition, one of three children with a complain of palpitation was decided in favor of surgery because a huge atrial appendage dilatation appeared via echocardiography Excision of approximately 70x40mm left AAA with some areas

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as thin as 2-3mm in surgery. Early diagnosis before arhythmia or thromboembolic event can lead in well-prognosed results even with medication or surgery like our patient's post-op good progress.
Conclusion: Our hypothesis is fetal echocardiography may be better in early diagnosis of cardiac aneurysms. We would like to demonstrate importance of fetal echocardiography and to contribute natural history of congenital atrial appendage aneurysms with our results.

Keywords: Atrial Appendage Aneurysm, Fetal Echocardiography, Fetus, Children

P-044

Benefit of prenatal diagnosis of vascular ring: case series and postnatal outcomes

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Background and Aim: Vascular rings are rare cardiovascular malformations which may present with respiratory symptoms or/and feeding difficulties in early childhood. Over the last twenty years, prenatal diagnosis of vascular rings has increased and can positively impact on the postnatal management and outcomes
Methods: We present four different subtypes of vascular ring detected at the mid trimester scan. Case 1: 33-year-old woman, gravida 2, para 0, referred at 29 weeks gestation, because of an abnormal 3 vessel-trachea (3VT) view and a suspected tracheoesophageal fistula. Fetal echocardiography confirmed normal intra-cardiac anatomy with a dominant right aortic arch (RAA) and antegrade flow into a smaller left aortic arch (LAA) composing a double aortic arch (DAA). Case 2: 26-year-old woman, gravida 1, para 0, referred at mid trimester screening scan due to suspected intra-cardiac abnormality, fetal diagnosis of tetralogy of Fallot and DAA with dominant RAA was made. Case 3: 32-year-old woman, gravida 2, para 1, referred at 22+5 weeks' gestation. Left circumflex aortic arch (LCAA) was identified, aorta ascended to the left of the trachea, crossed posterior to the oesophagus and descended to the right of the spine, along with right ductus arteriosus (RDA). Case 4: 31-year-old woman, gravida 1, para 0, referred at 21+1 weeks gestation with an abnormal 3VT, and a RAA with left ductus arteriosus and possible aberrant vessel was diagnosed.
Results: All the cases were confirmed postnatally by echocardiography, CT and ENT assessment were required. All four cases had symptoms of airway compression postnatally. Case 1 & 2 were born prematurely and required ventilator support. Case 3 had stridor within a few weeks of life and associated with 22q11deletion syndrome. Due to the presence of a prenatal LAA and RDA the diagnosis of a vascular ring was suspected early and may have otherwise been delayed if only a LAA was noted postnatally. Case 4 had symptoms of airway compression and was found to have a DAA postnatally and underwent surgical resection. No patient had residual airway symptoms or feeding difficulties within the two-year-follow-up.
Conclusion: Prenatal detection of vascular ring continues to improve, ensuring prompt postnatal investigation and facilitating surgical correction.

Keywords: vascular ring, antenatal diagnosis

P-047

Prenatal diagnosis of vascular rings: clues to diagnosis and postnatal correlation

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Background and Aim: Diagnosis of vascular rings has improved lately because of the widespread use of the 3-vessel and trachea view (3VV) in fetal echo. Although most patients with vascular rings are asymptomatic, some may have tracheal or esophageal compression needing surgery. Therefore, accurate prenatal diagnosis should be warranted.
Methods: We present a series of 16 fetuses diagnosed in the last 6 years with vascular rings and give some clues for an easier diagnosis, such as the "Z sign" at the 3VV for double aortic arch (DAA) (figure 1A). We included 8 patients with DAA and 8 with right aortic arch (RAA), aberrant left subclavian artery (ALSA) and left arterial duct.
Results: The main indication for echocardiography was suspected abnormality on obstetric ultrasound and the mean gestational age at the time of diagnosis was 25,3 weeks (20 to 34 weeks). Two families elected to terminate the pregnancy: one with an associated left congenital diaphragmatic hernia and one with a 22q11 deletion. All other patients were born at term. All patients with DAA underwent a Computed Tomography Angiography (CTA) before neonatal discharge, only one with tracheal compression who had surgery before discharge. During follow-up, 5 patients with DAA required surgery at a mean age of 6 months. Patients with RAA and ALSA underwent a CTA at a mean age of 18 months (1 week to 6 years), 5 of them had elective surgery at 21 months of age (5 months to 6 years). Indications for surgery were tracheal or esophageal compression more than 50% (figure 1B) and/or respiratory or gastrointestinal symptoms.
Conclusion: Diagnosis of vascular rings is feasible in prenatal life with fetal echo. The "Z sign" is useful for diagnosing DAA. The grade of compression of encircled structures and the definitive diagnosis may be confirmed by CTA.

Keywords: vascular ring, double-aortic arch

1A "Z sign". 1B Tracheal compression

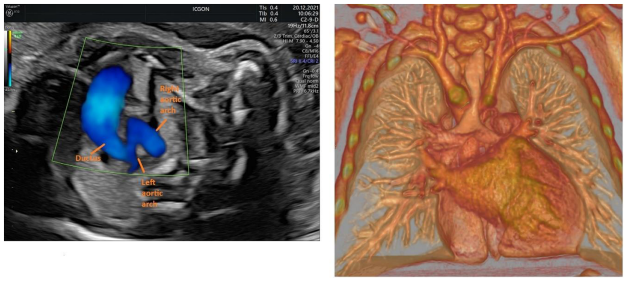

Figure 1A "Z sign" in fetal echo in a patient with DAA Figure 1B Computed Tomography Angiography showing tracheal compression in a patient with DAA

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P-048

Fetal cardiac arrhythmias: from prenatal evaluation and treatment to postnatal long-term follow-up

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Background and Aim: Fetal tachyarrhythmias(FT) may cause low cardiac output, hydrops, preterm delivery and perinatal morbi-mortality. An accurate diagnosis and treatment is crucial to improve prognosis.
Our aim is to describe the prenatal and postnatal evolution of FT, treatment effectiveness, morbi-mortality and neurological complications detected during follow-up.
Methods: FT in a single center were prospectively included for 18 years (2004-2021). To determine the type of FT, atrial and ventricular rates, ventriculo-atrial (VA) and atrial-ventricular intervals and relationships were assessed by fetal echocardiography. Hydrops and cardiac dysfunction were registered. Postnatal data was collected at neonatal age, first year and at the end of study.
Results: FT were diagnosed in 59 pregnancies at a median of 28 gestation weeks (interquartile range (IQR) 7). 59.3% had short VA intervals, 11.9% long VA intervals and 13.6% were flutter; until 2007 VA intervals were not routinely analyzed (n = 9, 15.2%). At diagnosis, 14 had hydrops and 11 heart dysfunction; severe presentation was not associated with time of diagnosis.
43 fetuses received transplacental antiarrhythmics, digoxin as first-line agent (95.2%), requiring 2 drugs in 24/43(55.8%), mainly flecainide; 88.4% converted to sinus rhythm. 7(12.3%) fetuses were delivered preterm, most of them due to uncontrolled arrhythmia and worsening hydrops (median 33 weeks-of-gestation, IQR 3.5). 2 fetal deaths were reported, one with hydrops, the other with extrasystoles. Flutter had more severe affectation at diagnosis ($p = 0.006$) and required early delivery (Relative Risk 4.78). Treatment success (born on term and with sinus rhythm) was 74.6% and overall survival 96.6%. After delivery, 40.7% relapsed (72.4% in long VA intervals, 48.5% in short VA, 37.5% in flutters), a newborn with refractory Permanent Junctional Reciprocating Tachycardia died after attempting an ablation. Only 4 patients had tachycardia at one year-of-life (23 under treatment) and 2 at the end of the study (3 under treatment). Within the patients with long follow-up (≥5 years), 17.1% had neurological sequelae. Bad prognosis (death, prematurity, neurological sequelae) was associated with hydrops ($p<0.001$) or flutter ($p = 0.015$).
Conclusion: Identifying high risk FT (flutter, hydrops) is useful to stratify postnatal prognosis. Postnatally, few persist with arrhythmias at follow-up, but it's worth noting the proportion of neurological sequelae, many related to hydrops.

Keywords: Arrhythmia, supraventricular tachycardia, atrial flutter, prenatal diagnosis, fetus.

P-049

Impact of antenatal echocardiography and pronostic factors for postoperative outcome in infants with aortic coarctation of the aorta

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Background and Aim: The aims of this study were to: 1)analyse impact of antenatal echocardiography, 2) assess pronostic factors of outcome, in neonates and infants with coarctation of aorta (COA).
Methods: This is a single-center analysis of clinical and echocardiographic data of fœtus with suspicion of, and neonates and infants <1 year of age with COA. Groups DAN (antenatal diagnosis) and DPN (postnatal diagnosis) were compared.
Results: Overall 286 cases were included, 106 in DAN group and 180 in DPN. Among 106 antenatal suspected COA, 11 were not confirmed after birth (sensitivity = 34.5%). Antenatal ascending aorta diameter (AAO) and AO/PA ratio were predictors of postnatal COA. Heart failure (HF) was the main symptom in DPN group, absent in DAN ($p< 0.0001$). Antenatal echocardiography was associated with lower diameter of isthmus (AOI), transverse arch (TrAO) and AAO. DPN had lower LV systolic fraction (LVSF) than DAN. Preoperative HF was associated with narrowest AOI, AAO, TrAO and decreased LVSF. PGE was administred in 53% preoperatively and mechanical ventilation in 39%. Among 275 confirmed COA (95 DAN and 180 DPN), 272 underwent Crafoord operation, at mean age 29 days (median 12 days) and mean time after admission 3.4 days. Age at surgery was lower and time to surgery longer in DAN than DPN group. Residual postoperative aortic gradient was observed in 18 cases (6.7%), and spontaneously disappeared in the majority of them within 2 postoperative days. Median hospital stay was 11 days. Survival was 98.5%. Restenosis occurred in 20 cases (7%), was related to postoperative AOI diameter and gradient, and AAO, and was more frequent in patients <1 month at surgery (8.5%). Mortality was associated with Shone complex and parachute mitral valve ($p<0.0001$), LV diameters ($p = 0.02$), ICU stay ($p<0.0001$), hospital stay ($p = 0.02$).
Conclusion: Antenatal diagnosis impacts on early outcomes of neonates with COA. Postoperative isthmus diameter and gradient, and <1month age at surgery impact on restenosis.

Keywords: antenatal diagnosis, coarctation of the aorta, pronostic factors

P-050

Accuracy of fetal echocardiography diagnosis and anticipated perinatal and early postnatal care in congenital heart disease

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Background and Aim: Our aim was to determine discrepancies in fetal congenital heart disease (CHD) diagnoses and anticipated early postnatal care and outcomes.

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Methods: Retrospective review of 462 randomly selected cases (23% of all cases) referred to a fetal cardiac assessment during second trimester (mean 26 weeks) at the Children's Hospital in Helsinki between 10/2010 and 12/2020. Discrepancy between prenatal and postnatal CHD case evaluations was assessed with independently provided cardiac severity and surgical complexity scores.
Results: 250 cases, 181 CHD and 69 normal, with complete prenatal and postnatal live birth data as well as seven terminations with fetal autopsy reports available were included in the analysis. There were 11 false normal and 7 false abnormal prenatal assessments, the accuracy of the normal/abnormal assessment was 97%. The prenatally anticipated level of early neonatal care was actualized in 61.6% and prostaglandin infusion in 95 %. In total 32.7% (84/257) cardiac severity scores were discrepant and in 13.2% (34/257) cases the discrepancies were considered significant (≥+/- 2 scores). Among significant discrepancies, CHD severity score was overestimated in 12 and underestimated in 22 in fetal assessment. The most common diagnostic categories included VSDs (N = 7), borderline ventricles (N = 7; five left heart, one right heart, and one DORV/TGA), arch anomalies including coarctations (N = 5), and tricuspid valve dysplasias (N = 4) with a change in postnatal diagnoses, treatment and outcomes.
Conclusion: Although fetal CHD diagnosis and counselling is accurate and reliable in general, the study highlights specific areas of uncertainty that may be important to consider in fetal CHD evaluation and counselling provided in mid-gestation.

Keywords: Diagnostic Accuracy, Congenital Heart Disease, Fetal, Postnatal Care

P-051
Congenital heart defects and increased nuchal translucency - associations with mortality

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Background and Aim: Fetuses with increased nuchal translucency (NT) measured in the first trimester of pregnancy are at risk of chromosomal abnormalities and congenital malformations including congenital heart defects (CHD).The risk for CHD is doubled if the NT is increased above the 95th centile, even if the karyotype is normal. It is unknown if an increased NT in first trimester increases the risk of mortality. Our aim was to investigate if increased NT was associated with increased mortality in children with CHD.
Methods: We performed a national cohort study based on the Danish health registries and identified 4469 children with CHD from 2008-2018 (0.7%); 3715 (83%) with simple CHD and 754 (17%) with complex. Increased NT was defined as having a NT >1.5 Multiples of Median (MoM). NT measurements, at corresponding crown-rump lengths, from the background population was used as reference. Children with chromosomal abnormalities or no first trimester screening were excluded. Proportions were compared using chi-squared test. Mortality rates were analyzed with Cox proportional hazards regression. A significance level of 0.05 was used in all analysis.
Results: Mortality was more frequent in children with any type CHD and NT >1.5 MoM than <1.5 MoM (3.3 vs. 1.5%, p = 0.03), HR 2.2 (95% CI 1.1, 4.8). Adjusting for complex CHD decreased the HR and significance; HR 1.6 (95% CI 0.8, 3.5). There were significantly more mortalities in the group of simple CHD with increased NT > 1.5 MoM (2.3% vs. 0.6%, p = 0.01) and the Hazard Ratio for mortality was 3.7 (95% CI 1.3, 10.7), compared to those with NT <1.5 MoM.
Complex CHD was more frequent in the group with NT>1.5 MoM (27% vs. 16%, p<0.01), but was neither associated with higher mortality (6.1% vs. 5.8%, p = 0.93) nor higher HR (HR = 1.0 (95% CI 0.4, 2.9)
Adjusting for confounding variables; extracardiac anomalies, birthweight, small for gestational age, preterm birth and preeclampsia did not alter the results.
Conclusion: Increased NT in the first trimester of pregnancy was associated with increased mortality in children with simple CHD. Increased NT may be a marker to identify children at extra risk besides their CHD.

Keywords: Nuchal Translucency Congenital Heart Defects Mortality Prenatal Screening

P-052
Long term follow-up of fetal cases with tricuspid valve anomalies

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Background and Aim: Retrospective-prospective study of the characteristics and long-term outcomes of fetal cases with tricuspid valve anomalies, aiming to assess negative prognostic factors.
Methods: The echocardiographic data and long term outcome (9m-30yrs) of 76 fetuses diagnosed between Dec.1986 – Sept. 2021 as Ebstein (Ebst) or non-Ebstein anomaly (NE), at 13-34 week's gestation (wg), median 28, were analysed. Twentytwo cases had Ebst, 1 with mild coarctation (CoA), 2 with ventricular septal defect (VSD) and 34 had NE, 1 with mitral dysplasia, 1 CoA with multiple VSDs. One Ebst had labiopalatoschisis, 1 NE spina bifida, 3 Ebst had familial history of congenital heart disease, 1 mother was taking lithium and 1 gardenal. One NE was twin, and another one was a pregnancy after ICSI. Following variables were compared by means of Fisher's exact test in cases continuing pregnancy (died versus survivors): grade of tricuspid regurgitation (TR), Celermajer index (CInd), fetal hydrops (FH), pulmonary stenosis/atresia (PS, PAtr).
Results: Echocardiographic features: 16/22 Ebst had a moderate-severe displacement of the TV and severe TR at presentation, 3 had PS, 7 PAtr, 5 had FH; 7/34 NE cases had severe TR and 7 moderate one, 5 had PS, 8 PAtr, 4 FH. Outcome: 2 Ebst and 4 NE cases opted for the termination of pregnancy, 4 Ebst and 1 NE died in utero at 13-32 wg (4 FH, 1 supraventricular tachycardia), the remaining cases were delivered at 31-39 wg. Spontaneous neonatal death occurred in 3 Ebst and 4 NE, at 1-17days in FH; 5/6 Ebst and 2 NE died postoperatively. One Ebst died at 3 yrs for resistant complex arrhythmias and severe TR. Total mortality in cases continuing pregnancy was 14/20 Ebst (60%) and 7/30 NE (23.3%). Risk factors for death were

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TR and CInd in Ebst (p = 0.03 and 0.003) and PAtr and CInd in NE (p<0.001 and p = 0.001). Survivors had minor TR and CInd and no FH.
Conclusion: Our data confirm a relevant mortality of severe tricuspid valve anomalies diagnosed in utero, main negative prognostic factors being the grade of TR, CInd and PAtr. Milder forms of both variants stabilized after birth.

Keywords: fetus, Fetal Cardiology, Ebstein, Non-Ebstein dysplasia, Fetal hydrops

Imaging/Functional assessment

P-055
Magnetic resonance imaging measurement of brain tissue motion in humans: a systematic review

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Background and Aim: Recent advances in magnetic resonance image (MRI) acquisition and analysis have allowed study of the biomechanics of the brain in unprecedented detail. As neurovascular pathology have potential to affect tissue biomechanical properties, the measurement of brain tissue pulsations (BTPs) may be used in the future for diagnosing pathology and determining prognosis. This review summarises existing MR techniques for exploring cardiac-induced brain movement to identify future directions for research into brain tissue motion in health and disease.
METHOD:Two databases were searched on the 10th of May 2021 using MEDLINE and SCOPUS, with no date limit. The search strategy included a prospectively registered protocol, with predefined study selection criteria, quality evaluation, and data extraction by two reviewers. The findings of the review were reported according to the PRISMA scoping review reporting guidelines.
Results: Our systematic search retrieved 18 eligible articles investigating cardiac-induced brain tissue motion using MRI. Four main MRI techniques were identified for visualising and/or quantifying BTP in humans; most studies are feasibility studies involving small numbers of participants. MR techniques agree that brain tissue motion varies regionally, with larger displacements closest to the centre of the brain compared to peripheral brain regions. Some studies have found that cardiac-induced pulsations are affected by the presence of a Chiari-I vascular malformation (CM-I). A pooled analysis suggests that there was a statistically significant difference in pulsation amplitude [ΔP] for the cerebellar tonsillar region of 0.31 mm [95% CI: 0.23, 0.38, p<0.0001] between CM-I patients and healthy controls.
Conclusion: This review establishes that measurements of brain pulsations using MR techniques gives valuable regional information of brain motion. However, studies of brain pathology are extremity limited to CM-I but suggest BTP can significantly modified in the presence of pathology. Further larger studies are required to obtain normative values for comparison with data from patients with conditions such as stroke.

Keywords: Brain tissue displacement. Brain tissue pulsation. Magnetic resonance imaging. MR

P-056
Associations between new cardiac blood biomarkers, magnetic resonance advanced imaging and exercise capacity in fontan patients

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Background and Aim: Early diagnosis and staging of circulatory failure in single ventricle (SV) patients remain challenging. We sought to test the diagnostic value of new cardiac blood biomarkers in relation to advanced measures of cardiac function and exercise capacity.
Methods: Cardiovascular magnetic resonance (CMR), exercise testing and blood samples were prospectively performed in 49 SV patients, of which 30 were males (61%) and 24 (49%) had a single left ventricle (LV). Time interval from Fontan operation was 11 (8-16) years. In addition to standard volumetric measurements, CMR provided measures of T1 mapping (pre- and post-contrast), circumferential (GCS) and longitudinal strain (GLS) of the SV. New cardiac biomarkers included growth differentiation factor 15 (GDF-15), C-telopeptide for type I collagen (CITP), type III procollagen aminoterminal propeptide (PIIINP), soluble suppression of tumorigenicity 2 (sST2), protein delta homolog 1 (DLK1), insulin-like growth factor-binding protein 7 (IGFBP-7), tissue inhibitor of metalloproteinases (TIMP1, TIMP4), matrix metalloproteinase 2 (MMP2), and fatty acid binding protein 4 (FABP-4).
Results: A significant correlation was found between GDF-15 and EDV (ρ = 0.31, p = 0.04), ESV (ρ = 0.3; p = 0.05), mass (ρ = 0.32, p = 0.04), T1 (post) (ρ = 0.42, p = 0.01), and GLS (ρ = -0.31, p = 0.05). CITP correlated with reduced GCS (ρ = -0.28, p = 0.05), PIIINP with T1 (post) (ρ = 0.28, p = 0.04), sST2 with EDV (ρ = 0.28, p = 0.05) and ESV (ρ = 0.31, p = 0.04), DLK1 with T1 (post) (ρ = 0.3, p = 0.07). IGFBP-7 correlated with EDV (ρ = 0.27, p = 0.05), mass (ρ = 0.26, p = 0.04), and T1 (post) (ρ = 0.51, p<0.01). No correlation existed between TIMP1, TIMP4, MMP2, and FABP-4 and any other CMR parameters. Exercise capacity was not correlated to any biomarker levels.
Conclusion: In SV patients, different biomarkers correlate with different measures of ventricular volume, mass, fibrosis, and deformation. In contrast, decreasing exercise capacity is a late clinical sign and may be influenced by other factors. The role of novel biomarkers requires further investigation in future longitudinal studies analyzing changes, similarly as for advanced imaging parameters. Some of these biomarkers may help for early detection of deteriorating cardiac function.

Keywords: Single ventricle, Fontan, biomarkers, magnetic resonance advanced imaging

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P-057/Moderated Poster

Reduced neonatal fronto-limbic connectivity is associated with higher externalizing symptoms in toddlers with congenital heart disease

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Background and Aim: Infants with Congenital Heart Disease (CHD) are at risk of impaired neurodevelopment. Our aim was to characterise the relationship between neonatal brain connectivity and behavioural outcomes at 22 months.

Methods: Forty-three infants [21 male; gestational age at birth median (interquartile range, IQR) = 38.57 (38.29-38.86) weeks; postmenstrual age at scan median (IQR) = 39.29 (38.71-39.71) weeks] with serious or critical CHD underwent presurgical brain diffusion MRI on a 3T scanner situated on the neonatal unit at St Thomas' Hospital, London. Infants underwent a neurodevelopmental assessment at median (IQR) 22.1 (22.0-22.7) months corrected age. Cognitive abilities were assessed with the Bayley Scales-3rd Edition. Parents completed the Child Behaviour Checklist questionnaire, and internalizing (emotionally reactive, anxious/depressed, withdrawn behaviours, somatic complaints) and externalizing (attention problems and aggressive behaviours) scores were calculated (higher scores indicate increased symptomatology). Index of multiple deprivation (IMD) was calculated from postcode at birth as a measure of socioeconomic status. Cumulative days in intensive care post-surgery, minutes on cardiopulmonary bypass and days to final corrective or palliative surgery were also calculated. Partial Spearman's rank correlations were used to assess the association between days in intensive care, time on bypass and days to surgery and internalizing and externalizing scores covarying for type of CHD, gestational age at birth, sex, brain injury severity, cognitive composite score and IMD. p-values underwent false discovery rate correction (pFDR). Structural connectivity was investigated using networks constructed with brain regions as nodes and quantitative streamline densities connecting each region as edges. Edge-wise associations between connectivity and internalizing and externalizing scores was assessed using network-based statistics controlling for sex, gestational age at birth, postmenstrual age at scan, brain injury severity, cognitive composite score and IMD.

Results: Reduced connectivity in a fronto-limbic network was associated with higher externalizing scores (Figure 1). Externalizing scores were not associated with days in intensive care ($\rho = -0.368$ pFDR = 0.901), days to surgery ($\rho = -0.502$, pFDR = 0.590) or bypass time ($\rho = -0.130$, pFDR = 0.590). Higher internalizing scores were associated with fewer days to surgery ($\rho = -0.680$, pFDR = 0.034) but not bypass time ($\rho = -0.480$, pFDR = 0.590) or days in intensive care ($\rho = -0.453$, pFDR = 0.629).

Conclusion: Reduced structural connectivity in a fronto-limbic network at birth may underlie externalizing symptoms in children with CHD.

Keywords: brain imaging; MRI; neonatal; behavioural outcome

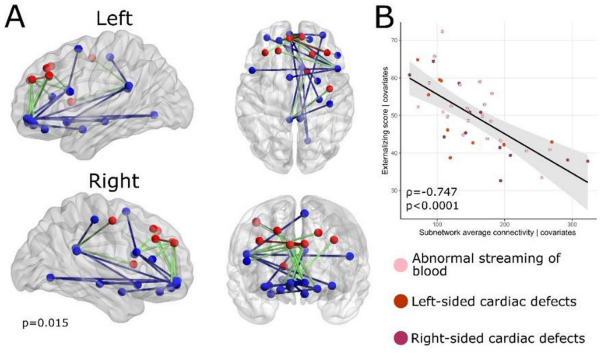

Figure 1. A) Network of reduced connectivty associated with increased externalizing behaviours, red nodes are core, blue nodes are peripheral; red edges are core connections, blue edges are peripheral connections and greed edges are peripheral to core connections according to Ni Bhroin et al 2020. B) Association between externalizing scores and average connectivity from subnetwork corrected for covariates

P-058

The role of 18F-FDG PET/CT in the diagnosis of pediatric infective endocarditis

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Background and Aim: Infective endocarditis is a severe disease diagnosed using a combination of clinical, microbiologic, and imaging criteria. Diagnosis is challenging, especially in children with right-sided endocarditis and patients with prosthetic materials. In this study, we evaluated the role of fluoro-18 fluorodeoxyglucose positron emission tomography/ computed tomography (18F-FDG PET/CT) for infective endocarditis diagnosis in children.

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Methods: Infective endocarditis was diagnosed using the modified Duke criteria. The studies were acquired in the supine position on a GE Discovery-710 PET-CT. No intravenous contrast was administered other than the 18F-FDG injection (5.5 MBq/kg). The PET studies were acquired in a three-dimensional mode for a total acquisition time of 2 minutes per bed position adjusted according to the patient weight, from vertex to toes and 5 minutes a single bed additional cardiac scan at about 1-hour post-injection.
Results: A total of 27 pediatric patients diagnosed with infective endocarditis at our institution between December 2018 and December 2021 were recruited to the study. The median age and weight of the patients were 13,4 years (range 1,2 – 25 years) and 34 kg (range 7–65kg), respectively. Eighteen of the patients were boys, and 9 were girls. The most common presenting symptom was fever (19 patients) followed by cough (4 patients) and chest pain (3 patients). The mean duration of symptoms before the diagnosis was 7 days. Except the patient diagnosed with acute lymphoblastic leukemia, all patients had congenital or acquired heart disease. Twenty-two patients were operated previously. Nineteen of them had prosthetic material. Infective endocarditis involved left-sided structures in 8 patients and right-sided structures in 19 patients. Nine of patients had extracardiac involvement. In one patient, the FDG-PET-CT was performed twice to evaluate the treatment of infective endocarditis, and the regression of infection was detected.
Conclusion: The diagnosis of endocarditis might be challenging due to the limited visualization of the conduits and peripheral pulmonary artery stents by transthoracic or transesophageal echocardiography. We suggest that PET/CT could be an excellent option to make a confident diagnosis in pediatric infective endocarditis.

Keywords: infective endocarditis, positron emission tomography, 18F-FDG, PET/CT

P-059

The results of pulmonary hypertension treatment in single ventricle patients

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Background and Aim: Anti-Pulmonary Hypertension (antiPH) treatment in single ventricle patients is still a controversial issue, and different experiences and recommendations are presented in the literature. This study was planned to evaluate the antiPH treatments and prognosis of single ventricle patients followedup in our clinic.
Methods: The records of 33patients with single ventricle who received antiPHtherapy were included in the study. Patients ages, genders, cardiac diagnoses, antiPH treatments and follow-up periods, course during treatment, examinations, findings of poor prognosis were recorded.
Results: Of the 33patients, 15(45.4%) were male and 18(54.5%) were female. The median age of the patients was 10 years (1-41 years), median follow-up period was 60 months (9-113 months) under antiPH treatment. 26 patients were under the age of 18, 4 patients had Down syndrome.
3 patients died at the median follow up of 4.2 years (2-6 years). 10 patients were unbalanced CAVSD, 9 patients were DILV, 4 patients were Tricuspid atresia, 4 patients were correctedTGA with systemic ventricle hypoplasia, 3 patients were mitral atresia, 3 patients were under follow up with other diagnoses. Monotherapy was initiated in 29 patients (ERA in 22, Sildenafil in7), multidrug therapy was initiated in 4 patients (ERA+/- PDE5 inhibitors+/- prostocyline analogues).
After the initiation of antipH treatment 4 patients were operated following echocardiographic and catheter evaluations and antiPH treatment was ceased at follow ups. 6 patients were operated but antiPH treatment was continued. A patient could not tolerate the Glenn shunt performed after 2.5years of antiPH treatment and Glenn takedown was performed. 7 more patients became suitable for palliation after antiPH treatment and waiting for the surgery. Of the 17 (51%) patients who became suitable for palliation after antiPH treatment initiation, 9 (90%) out of 10 operations were successful.
Conclusion: AntiPH therapy in single ventricle patients is a strategy that is still being questioned and boundaries havenot been fully drawn. When the fact that approximately half of our patients became suitable for palliation with antiPH treatment and the successful outcome of the operation in 90% of these patients is taken into consideration, we think that it is a treatment option for symptomatic patients who do not have a chance of biventricular repair but also not suitable for single ventricle palliation

Keywords: pulmonary hypertension, single ventricle

P-060

The natural history, management, and outcomes of pediatric subaortic stenosis in a tertiary referral center

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Background and Aim: Subaortic stenosis is remarkable for its unpredictable hemodynamic progression in childhood and high reoperation rate. This study presents the natural history, surgical outcomes, and follow-up of pediatric patients diagnosed with subaortic stenosis.
Methods: We retrospectively reviewed 66 pediatric patients who operated for subaortic ridge from 2015 to 2021 in a tertiary referral center. Patients with atrioventricular septal defects, accessory mitral valves, hypertrophic cardiomyopathies were excluded.
Results: The mean age of 66 patients was 5.1 ± 4.7years, and 47 (71.2%) patients were male.
Ridge resection was performed in 30 patients during VSD closure(Group I), while 36 patients required surgery due to significant subaortic stenosis (Group II; ridge was present initially in 30 of them). Four of the patients in the first Group had moderate subaortic stenosis. In Group I, there was Aortic insufficiency (AI) in 20 of the patient (with AVP in 14), on follow-up, residual ridge in 14 patients and residual AI in 19 patients (AI degree decreased or remained the same in 18 patients) were determined. In the second group peak gradient at diagnosis was 79 mmHg with AI in 80.5%. Preoperatively their peak gradient progressed to 89.5 mmHg. In 8 patients, additional Aortic valvular pathologies were diagnosed, and in all of those, valvuloplasty was performed. In these cases with valvular pathologies, the mean gradient was 91 mmHg. After resection of subaortic stenosis in 36 patients, there was no residual stenosis in 21 patients; further more, the residual Doppler peak systolic instantaneous gradients were under 40 mmHg in 14 patients 55 mmHg in 1 patient. The patients were followed with echocardiography for a median of 2 years (range:1 months-6.5 years) after surgery. The degree of AI progressed in 9(25%) patients (trivial to

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mild: 1, trivial to moderate: 2, mild to moderate: 6). Six of 36 surgical patients (16.7%) required reoperation due to recurrent stenosis in 5 and AI in 1 of the patients after a median time of 3.2 years(range:1.4-8 years).
Conclusion: Left ventricular outflow tract gradient and flow turbulence occurring in discrete subaortic stenosis causes valvular distortion over time. Therefore, frequent echocardiographic evaluations are crucial for these patients to detect subaortic stenosis progression and early complications.

Keywords: Aortic regurgitation, children, subaortic stenosis, subaortic membrane.

P-061
The evaluation of the patients with posterior transposition of great arteries

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Background and Aim: Transposition of great arteries (TGA) with posterior aorta (p-TGA) is a rare form of transposition characterized by the right and posterior course of aorta after arising from right ventricle. The first case was reported by Quero-Jimenez et al in 1969. We want to present here in detail the demographic, echocardiographic and surgical features of our 9 patients with this rare diagnosis in detail.
Methods: The records of TGA patients who were admitted to our clinic between 2015-2021 were reviewed retrospectively and 9 patients with the diagnosis of pTGA were included in the study. The demographic, echocardiographic and surgical reports were examined
Results: 8 of the patients were male and 1 patient was female. The median age at admittance was 3 months (23 days-14 months). Patients usually admitted to the clinic with the complaints of cyanosis, rapid breathing or murmur at pediatric examination. Echocardiography revealed a concordant atrioventricular and a discordant ventriculoarterial relation. All patients have a near normal relation of great arteries with aorta localized on the right and posterior to pulmonary artery. A ventricular septal defect (VSD), mostly doubly committed was present in all of the patients. Patent ductus arteriosus, atrial septal defect, pulmonary hypertension, additional VSDs, pulmonary stenosis and arch hypoplasia were other pathologies demonstrated on echocardiographic evaluations. Pulmonary banding was performed in 2 patients before admitting to our clinic. All of the patients were operated and arteryel switch was performed without Lecompte maneuver in 7 patients.
Conclusion: Transposition of great arteries accounts for 5-7 % of the all congenital cardiac anomalies and pTGA is a rare kind of this pathology without a reported incidence. We want to emphasize that this rare pathology with spesific echocardiographic features might require changes in operative techniques. The anterior location and shortness of pulmonary trunk in comparison with aorta and also the posterior location and length of left coronary artery might be obstacles for correction although a lecompte maneuvre is not usually needed

Keywords: posterior transposition of great arteries, echocardiography, pediatric

P-062
The juxtaposition of the atrial appandages and its effect on surgical approach

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Background and Aim: The juxtaposition of the atrial appandages (AAJ) was first reported in1893. Both of appendages or a part of an appandage and whole of the other appendage localized on the same side of the great artery. It is almost always associated with complex congenital heart diseases. Left-sided juxtaposition is more common. Transposed tricuspid atresia or complete transposition of the great arteries are the two pathologies most frequently associated with juxtaposition.
Methods: The records of patients who had the diagnosis of AAJ in years between 2012-2021 was reviewed retrospectively and the demographic data, cardiac pathologies, the Echocardiography (ECHO), Computed tomography (CT) and surgical notes of the patients were recorded. Our aim is to define the characteristics of the patients with AAJ and to investigate the effect of this pathology on the operation.
Results: 19 of the patients were male (%76). Left AAJ was present in 20 of 25 patients (80%). 92% of the patients were levocardic and ventricular looping was D-loop. 92% of the patients with AAJ had complex congenital heart disease and great vessel malposition or transposition. In all 5 patients with right AAJ, atrial situs were solitus and AV connections were concordant. 2 patients had DORV, 1 patient had TGA, 1 patient had TOF and 1 patient had VSD. Thirteen of 20 patients with left AAJ were single ventricular pathology. VA relation was discordant in 12 patients and AV valve atresia accompanied VA discordance in 5 of the patients. Total correction could not be performed in 1 out of 25 patients due to right AAJ and coronary anomaly, intraoperative central shunt decision was made. In another patient with left AAJ, the location of the atrial appendage had to be corrected to complete the operation.
Conclusion: In our study, we found that this rare anomaly is seen with complex congenital heart diseases where great vessel malposition almost always accompanies. We found that AAJ on the left is more common than on the right, in line with the literature. In addition, we wanted to emphasize that although this anomaly is rare, it can change the type and course of the operation and that is why it is important to diagnose with noninvasive imaging methods preoperatively.

Keywords: juxtaposition of atrial appendages, surgery, echocardiography

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P-063/Moderated Poster
The role of cardiac magnetic resonance imaging in pediatric myocarditis

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Background and Aim: A teenager presenting with chest pain and an elevated troponin level is a very common phenomenon in pediatric cardiology daily routine. CMRI is the gold standard method for diagnosis of acute myocarditis in these patients. However, there is insufficient data on the use of cMRI in the follow-up of patients with acute myocarditis. The aim of this study was to describe the follow-up results of patients with clinically diagnosed myocarditis.
Methods: Patients below 19 years of age, were included if they were admitted for suspected myocarditis and diagnosis confirmed by cMRI. Presenting signs and symptoms, diagnostic tests, medical treatment and patient outcomes were recorded. Acute myocarditis was diagnosed according to Lake Louse criteria. After six mounths later, the control cMRI evaluation was performed in patients.
Results: A hundred four patients were included in the study. The most common presenting symptoms were chest pain (88%, n = 91). Median troponin I (TnI) was 314 ng/L. The most common finding on electrocardiogram (ECG) was ST segment elevation, which occurred in 22% (n = 23). Of the 104 patients identified with myocarditis, 85 had normal LV systolic function on admission. Antienflamatuar treatment were provided to 63% (n = 66) of patients. Troponin levels were decreased within a median of 4 days. Extracorporeal Membrane Oxygenation (ECMO) was used to 2 (n = 2) of patients. Abnormal cardiac function at the time of discharge was noted in 4% (n = 4). One patient was referred to another hospital for heart transplantation due to poor left heart systolic functions and dilated cardiomyopathy. According to initial cMRI, 40% (n = 42) had MRI evidence of infamation on T1-weighted and T2-weighted images. The late gadelinium enhacement (LGE) was detected in 62% patients with myocarditis. In the outpatient examination after 6 months, normal LV systolic function was detected in 101 Patients. The late gadelinium enhacement was detected in 53% (14/26) in follow-up control MRI.
Conclusion: Myocarditis is a benign disease. LGE could be a valuable parameter in predicting the prognosis in patients with acute myocarditis. There is a need to develop a consensus regarding the timing for follow-up CMR study in patients with acute myocarditis if we are to use LGE as a marker for the prognostic significance and risk stratification.

Keywords: Cardiac magnetic resonance, Myocarditis, Troponin, Pediatric

P-064
Clinical and laboratory characteristics of children with infective endocarditis: a single-center experience

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Background and Aim: This study aimed to evaluate the clinical presentations and outcomes of infective endocarditis (IE) in a tertiary hospital in Turkey over three years period.
Methods: A retrospective study was conducted examining pediatric IE cases diagnosed and treated between January 2018 and October 2021 at the Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Center. Clinical presen,tation, treatment, complications, outcomes of IE, underlying pathogens, and congenital heart defects were reviewed. Two pediatric cardiologists assessed the clinical records.
Results: A total of fifty-one children were identified retrospectively. Twenty-four (47%) of patients were male. All patients had underlying cardiac conditions, and forty-two (82.3%) of them underwent cardiac surgery. A causative pathogen for IE was identified in 28 (54.9%) cases; Methicillin-resistant coagulase-negative staphylococci (CNS) in 13 (46%) and S. aureus in 5 (17%) patients. Twenty-two (43%) of the patients met the criteria for "Definitive IE" as per modified Duke criteria, with the remainder categorized as "Possible IE." Transesophageal echocardiography contributed to the diagnosis in 15 (29%) patients. Surgery was required in 28 (54.9%) of the patients. The median duration of antibiotic treatment was 44 (interquartile range = 34–52) days, and days of hospitalization was 51 (interquartile range = 32–75) days. One patient died secondary to IE related complications.
Conclusion: Congenital heart disease was the most common risk factor for IE, and S. aureus was the most common responsible organism. The incidence of pediatric IE in our center was higher than the reported incidence in the literature. However, our mortality rates were lower than in the literature. Awareness of IE and its prevention is crucial in patients with congenital heart defects.

Keywords: infective endocarditis, children, congenital heart disease, congenital heart surgery

P-065
Tetrallogy of fallot and hypertrophic cardiomyopathy - a rare association. clinical case

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Background and Aim: We present a rare clinical case of Tetralogy of Fallot associated with hypertrophic cardiomyopathy in a 9-month-old girl, without previous history of heart defect, who was admitted to intensive care unit with a clinical presentation of a hypoxic spell.
Methods: Child presented with severe general condition, with increased work of breathing, coughing and fussiness. Her saturation level was - SatO2- 44%, heart rate - 202 pbm., respiration - 40 breaths\m, BP – 95\50 mm/Hg. Physical development was below 10-th percentile. At chest examination she had severe subcostal retractions, and coarse breath sounds without wheezing or crackles. During physical examination skin appeared pale with prominent cyanosis of the lips. Cardiac exam showed increased precordial activity with 5\6 systolic ejection murmur that was heard along the left sternal border with radiation to the back. The liver edge was 2 cm below the right costal margin at abdominal examination.
Results: Electrocardiogram demonstrated sinus rhythm, right-axis deviation, incomplete right bundle branch block and right ventricular hypertrophy. Posteroanterior chest radiograph showed abnormal cardiac contour with upturned cardiac apex, diminished pulmonary vascular markings and mild cardiomegaly. During 2D echocardiographic exam: from parasternal long axis view hypertrophy of the left ventricle posterior wall, interventricular septum and right ventricle wall were visualized. Subaortic ventricular septal defect and overriding aorta were evident from long parasternal view. From short parasternal view the pulmonary stenosis with

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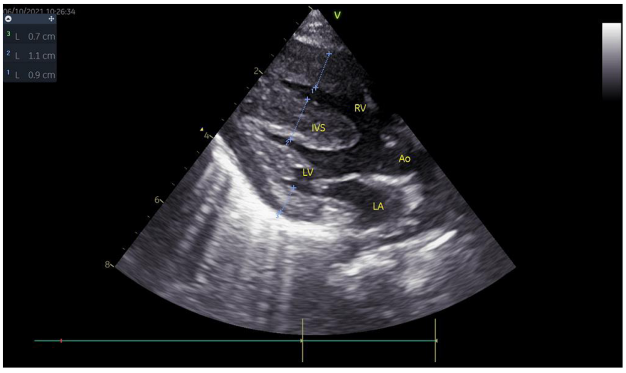
maximum pressure gradient of 104 mmHg was identified. The diagnosis of hypertrophic cardiomyopathy and tetralogy of Fallot was made and confirmed on computed tomography.
Conclusion: Tetralogy of Fallot is the most common cyanotic congenital heart disease with a prevalence of 10% of all congenital heart defects. Tetralogy of Fallot is often associated with chromosomal aberration, while hypertrophic cardiomyopathy associates with certain gene loci. Although many other heart anomalies may coexist, the association of tetralogy of Fallot and hypertrophic cardiomyopathy is extremely rare. Management of patients with tetralogy of Fallot associated with hypertrophic cardiomyopathy differs greatly from that of patients with tetralogy of Fallot alone, whose clinical and surgical management in most cases assures good long-term results.

Keywords: Tetrallogy of Fallot, hypertrophic cardiomyopathy, congenital heart defect, infant

parasternal long axis view.



Hypertrophy of the LV wall, IVS and right ventricular free wall. Overriding aorta and ventricular septal defect.

P-067
Automatic signal thresholding of ventricular volumes in young patients with various heart diseases – can we trust the numbers?

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Background and Aim: Systolic and diastolic volumes of the right (RV) and left (LV) ventricles are important for diagnosis and prognosis in many cardiac diseases. They are usually measured by segmentation of the myocardial borders on cardiac magnetic resonance (CMR) images. Signal thresholding techniques allow automatic myocardial contour detection but need to be validated before clinical use. We aimed to determine the accuracy and precision of these measurements in our paediatric center's referral population.
Methods: Consecutive CMRs of subjects with biventricular hearts performed between January 2018 and March 2019, were retrieved. Inclusion required steady-state free precession short-axis stacks and phantom-corrected, two-dimensional phase contrast flow sequences perpendicular to the ascending aorta and the main pulmonary artery. Patients with more than mild stenosis or regurgitation of one or more cardiac valves were excluded.
RV and LV volumes were measured in end-diastole (EDV) and in end-systole (ESV) manually and using the MassK application (QMass by Medis, Leiden, The Netherlands), with signal thresholds at 30% (k30), 50% (k50), and 70% (k70) and indexed to body surface area. Stroke volumes (SV) and cardiac indices (CI) were calculated and compared to aortic and pulmonary flow volumes and CI. Contouring was repeated by a second operator in 20 patients.
Results: Data were gathered from 94 patients (22 female) aged 15 +/- 9 years referred for evaluation of cardiomyopathy (32%), congenital heart disease (28%), aortic pathology (24%), healthy volunteers (14%) and others (2%). EDVi and ESVi were largest with manual segmentation of both ventricles and decreased with higher thresholds (manual versus k70: LV EDVi 82.3 ± 16.1 versus 59.6 ± 13; RV EDVi 86.6 ± 22.2 versus 62.7 ± 18.8; LV ESVi 35.1 ± 11 versus 20.7 ± 7.9; RV ESVi 41 ± 13.9 versus 25.3 ± 9.7 ml/m2; all $p<0.0001$). Manual and k30 SV and CI agreed best with flow measurements (Table). Interobserver bias was small for all volumes by manual and MassK (-0.44 to -4.79 ml/m2).
Conclusion: Manual and 30% threshold based segmentation of biventricular volumes agree best with phantom-corrected two-dimensional phase contrast flow measurements in a young referral population and are well reproducible.

Keywords: Cardiac magnetic resonance imaging, ventricular volume, segmentation, automation, flow

Table: Bland-Altman analysis (bias +/- 1.96 standard deviations)

	Manual vs. flow	k30 vs. flow	k50 vs. flow	k70 vs. flow
LV SV [ml]	2.06 ± 9.35	1.06 ± 9.70	-3.95 ± 8.79	-10.23 ± 8.26
RV SV [ml]	-3.691 ± 12.53	-4.50 ± 12.43	-8.25 ± 11.68	-15.40 ± 11.02
LV CI [l/min/m²]	0.05 ± 0.54	0.01 ± 0.56	-0.25 ± 0.51	-0.57 ± 0.46
RV CI [l/min/m²]	-0.27 ± 0.70	-0.31 ± 0.71	-0.51 ± 0.66	-0.90 ± 0.63

CI – cardiac index; k30 – MassK with 30% signal threshold; k50 – MassK with 50% signal threshold; k70 – MassK with 70% signal threshold; LV – left ventricle; RV – right ventricle; SV – stroke volume.

P-068
Review of the applicability of a semiautomatic device for the determination of flow-mediated vasodilation

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Background and Aim: Flow mediated dilation (FMD) is the standard diagnostic method to assess vascular function in greater vessels. New semi-automatic ultrasound devices are available with the advantage of simplifying the examination and reducing dependence on the user. The objective of this study is to investigate the reliability of the automated analysis and the dependence of the user on the results.
Methods: For FMD we used the UNEX EF 38 G ultrasound device (UNEX Corporation, Japan). 40 subjects were under standardized conditions examined and automatically analyzed. Then two raters processed the ultrasound images manually and independently of each other. The quality of the images was also assessed. After a one-week break, the data were reevaluated by each rater. We evaluated the agreement of the automatized whit the manually analysis and the inter- and intrarater reliability.
Results: The results from the automated Evaluation to the initial post-processing correlated with 0.32 ($p<0.001$). The overall ICC of the manually analysis was 0.82 ($p<0.001$). In detail: The interrater reliability showed a correlation of 0.75 ($p<0.001$).

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The intrarater reliability of the first rater was 0.88 ($p<0.001$) and the second 0.86 ($p<0.001$). The results are independent of the data quality of the images.
Conclusion: The results show that there is only poor agreement between automated and manual evaluation. This shows that for better data quality, the ultrasound images must be post-processed manually, but this does not necessarily have to be done by the same rater to ensure good quality.

Keywords: Vascular diagnostic, vascular function

P-069

A case series of prospective usage of the 2V-score in neonatal patients with borderline left ventricle and aortic arch anomaly

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Background and Aim: Newborns with aortic arch anomaly and hypoplastic left-sided structures constitute a broad spectrum of pathology ranging from patients eligible for two-ventricle repair to those who require one-ventricle palliation. For patients at either extreme, the treatment decision is relatively straightforward, but the management strategy of children that fall within the grey zone of the spectrum continues to be controversial. To date, the most promising scoring system for these patients is the 2V-score developed by Mart C.R. et all. It was determined from retrospectively obtained data, and, to our knowledge, has not been used yet prospectively to make a clinical decision. We aim to describe the results of the prospective usage of this method in five consecutive patients with borderline left ventricle (LV) and aortic arch pathology.
Methods: From May to October 2021, five consecutive patients who fulfill the next criteria were included: aortic and mitral valves that are hypoplastic but not atretic or stenotic; LV hypoplasia without endocardial fibroelastosis; hypoplasia of the LV outflow tract, ascending aorta, and aortic arch with or without coarctation. We validated patients for one- or two-ventricle repair based on the 2V-score. The cutoff value for two-ventricle repair of ≤16.2 was selected. Gender, anthropometric and echocardiographic data, postoperative complications, and outcomes were analyzed.
Results: The median weight was 3.16 (2.3–4) kg. The median 2V-score was 13.29 (10.51 - 16.01). All patients underwent two-ventricle repair. There was no hospital mortality. 3 of 5 had an uneventful clinical course with ICU stays 7, 14, and 19 days. One patient with 2V-score - 11.9 required chest tube insertion due to recurrent pleural effusion not related to heart failure. The last patient with the 2V-score 16.01 had the most severe postoperative period, complicated by heart failure with low cardiac output, supraventricular arrhythmia, and acute kidney injury with 59 days of ICU stay. To date, all patients are alive.
Conclusion: To conclude, the present study suggests that 2V-Score may be a useful tool in decision-making process in neonatal patients with the borderline left ventricle and aortic arch anomaly. Moreover, the absolute value of the score may predict the severity of the postoperative course.

Keywords: Newborns, Borderline Left Ventricle, 2V-Score

P-070

Exploring the benefits of 3D modeling in the pre-procedure evaluation of atrial septal defects

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Background and Aim: Computed tomography (CT) with the additional technique of 3D modeling and printing provide accurate information regarding the anatomy of various congenital heart malformations and allow virtual planning of procedures. Our aim is to explore the benefits of contrast CT derived 3D models in the pre-procedure evaluation of Atrial Septal Defects (ASDs). Comparative analysis focuses on the subgroup of patients who underwent transcatheter closure of the ASD
Methods: Within the past 2 years 3D modeling was performed on CT data of 40 patients with ASD. 47.5% (n = 19) patients underwent transcatheter closure. Of which 42.1% (n = 8) were male and 57.9% (n = 11) female with mean age - 21.96 years (6.19 to 60.11) at the time of procedure. Closure was performed in 89.5% (n = 17) with an Amplatzer ADO device, 5.3% (n = 1) with an Amplatzer PFO device and 5.3% (n = 1) PFM device.
3D modeling was performed via 3D Slicer software. Dimensions and anatomical characteristics of the ASD were compared to the transthoracic echocardiography (TTE), transesophageal echocardiography (TEE) and the performed balloon sizing during procedure. For the comparative analysis we chose the balloon sizing for a reference value to compare against ASD sizing by the imaging methods. For precise comparison derivative parameters were defined as the difference between ballon sizing and the analyzed method – i.e "Balloon-TTE"; "Balloon-TEE" and "Balloon-3D model". Parameters were analyzed via IBM SPSS statistics software.
Results: Mean values for the derived parameters are as follows: "Balloon–TTE" – 1.522 ± 2.78; "Balloon-TEE" – 1.42 ± 2.81; "Balloon-3D model" – 0.633 ± 2.68. The described parameters were tested against an expected mean value of 0. Only "Balloon – 3D model" showed no significant difference from 0 ($p = 0.33$ – retained null hypothesis).
Conclusion: Our data suggests that 3D modeling of an ASD provides accurate representation of size and anatomical characteristics – comparable to other imaging methods. In our experience this modality is useful in cases with difficult echographic windows. Additional benefits along with 3D printing include the option for virtual simulations and device planing, improved patient communication and trainee education with hands-on training. The process provides an invaluable learning curve in 3D modeling.

Keywords: 3D modeling, ASD, CT, Transcatheter closure, Virtual planning

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P-071

Left ventricular mechanics after arterial switch operation: just ejection fraction or something more?

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Background and Aim: Arterial switch operation (ASO) is considered the treatment of choice for infants with complete transposition of the great arteries (D-TGA). Despite its low operative mortality and high long-term survival, reduced left ventricular function is a reported complication. Speckle-Tracking Echocardiography (STE) is able to recognize subclinical myocardial abnormalities despite a normal ejection fraction.

The aim was to evaluate the impact of several perinatal clinical variables on left ventricular function of D-TGA patients, by using STE.

Methods: We enrolled 53 TGA-ASO patients born between 2013 and 2021 and followed in our outpatients clinic. They performed a standard echocardiography and a STE to assess global longitudinal strain (GLS).

Several perinatal clinical variables impacting left ventricular function were identified from the collected data: birth weight <2.5 kg; coronary anomalies; complex TGA; preoperative inotropic support; preoperative ECMO support; age at surgery >14 days; duration of surgery >5 hours; major arrhythmias; acute kidney injury requiring peritoneal dialysis; postoperative ICU length of stay >4 days; right ventricular outflow tract obstruction with systemic pressure; moderate or severe left ventricular outflow tract obstruction.

Accordingly, all the patients were divided in three groups: group 1 (up to two positive variables); group 2 (3 or 4 risk factors) and group 3 (5 or more variables fulfilled).

Results: Surgery was performed at a median age of 10 [7-12] days. Mean a follow-up was 51 ± 33 months. Mean left ventricular ejection fraction and GLS were 67.73 ± 9.09% and -19.07 ± 2.29%, respectively.

A significant reduction of longitudinal myocardial deformation parameters was observed when groups 1 and 2 were compared to group 3 (mean GLS difference: -2.64%, p = 0.038 and -2.07%, p = 0.030, respectively)(figure 1), while no significant difference was observed when evaluating ejection fraction (65 ± 5% vs 70 ± 6% vs 69 ± 7%) or TAPSE.

Conclusion: Patients who underwent ASO showed a good mid-term outcome. On the other hand, despite normal EF values, patients with three or more perinatal risk factors showed significantly lower longitudinal strain. Thus, GLS may be used to detect patients at risk. In case of low GLS values or worsening GLS values during follow-up, further exams might be indicated to exclude hidden residual defects, like coronary lesions.

Keywords: arterial switch operation, transposition of great arteries, speckle tracking, longitudinal strain, echocardiography

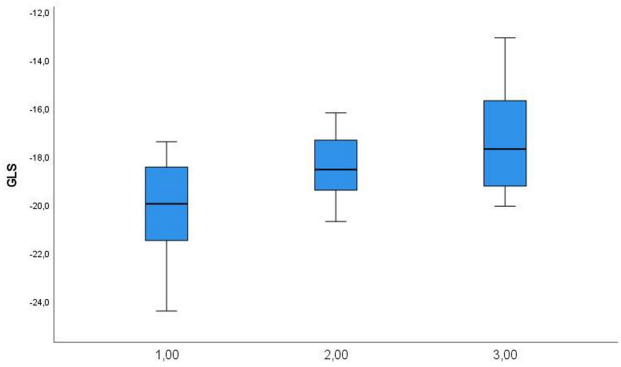

Figure 1. Box and plot of GLS values in the three groups.

P-072

Impact of gore cardioform asd occluder on atrial and ventricular mechanics in a pediatric population

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Background and Aim: Trans-catheter closure is nowadays considered as the first-line treatment of ostium secundum atrial septal defect (ASD). The GORE® CARDIOFORM ASD occluder (GCA) recently received CE mark. The device design is potentially innovative compared to the other self-centering devices. It puts together high softness and anatomic compliance with the potential to close defects as large as 30 mm.

The aim of this study was to compare the mechanical changes in atrial and ventricular properties before and after GCA implantation.

Methods: Study design: single arm prospective study. Enrollment: all the patients aged <18y performing isolated ASD closure with a single GCA device. Echocardiogram analysis was performed the day before, 24 h and 6 months after ASD closure. Atrial and ventricular speckle tracking parameters were recorded to study the heart mechanical remodeling after ASD closure with GCA device. Longitudinal strain evaluation was performed from bidimensional analysis, with the gray scale images analyzed offline with the Echopac v.12 workstation.

Results: Between January 2020 and February 2021, 70 pediatric patients with hemodynamically significant ASD were enrolled to undergo transcatheter closure with the GCA device. Mean age was 7.9 ± 3.9 years, mean defect diameter was 17.1 ± 4.5 mm. A 27 mm device was used in 15 patients, a 32 mm GCA in 31, a 37 mm in 15 patients, a 44 mm GCA device in 10 patients.

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Longitudinal strain analysis showed no change in global longitudinal function after ASD closure (Figure 1). A transient reduction in longitudinal strain was detected in basal septal segments and on atrial and right ventricular longitudinal function 24 after closure, secondary to changes in flow distribution after ASD closure. Six months after the procedure only left atrium showed a mild global longitudinal strain reduction, due to the presence of the device on the septum.

Conclusion: This study demonstrated that GCA had no impact on global and regional right and left ventricular longitudinal function. Atrial mechanics was preserved, excepting of the segments covered by the device. Based on the data available in Literature, this was the first device demonstrating no impact on the left and right ventricular mechanics, irrespectively from the device size used.

Keywords: atrial septal defect, Interventional Cardiology, device, Gore Cardioform Device, speckle tracking, echocardiography

	Time 0	24 h FUP	6 months FUP
LV A4C LS	-23.3±3.7	-22.9±3.0	-23.2±3.3
LV A3C LS	-23.3±3.3	-22.5±3.2	-23.0±3.0
LV A2CLS	-22.9±3.0	-23.2±3.5	-23.7±3.0
BS LS	-19.7±3.3	-18.3±3.4*	-19.5±2.7
BAS septum LS	-22.5±5.2	-20.6±5.0*	-22.6±5.0
LV GLS	-23.2±2.7	-23.0±2.7	-23.3±2.7
LA LS	41.5±15.4	29.4±12.2*	39.0±12.0*
RA LS	53.0±29.0	46.4±24.9*	56.5±26.0
RV LS	-27.4±5.5	-23.9±4.8*	-28.0±4.4

Figure 1. Longitudinal strain data before closure, 24h after closure and 6 months after closure. A4C: apical 4 chambers view, A3C: apical 3 chambers view, A2C: apical 2 chambers view; BS: basal septum, BAS: basal anterior septum, FUP: follow-up, GLS: global longitudinal strain, LA: left atrium, LS: longitudinal strain, LV: left venricle, RA: right atrium, RV: right ventricle. *: $p<0.05$ vs Time 0.

P-073

Evolution of echocardiographic and cardiac MRI abnormalities during follow-up in patients with previous MIS-C diagnosis

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Background and Aim: Growing evidence has documented a severe systemic hyperinflammation syndrome affecting children previously exposed to SARS-CoV-2, known as Multisystem Inflammatory Syndrome in Children (MIS-C). Cardiovascular manifestations in MIS-C are frequent (34%–82%). The aim of our study was to describe the early and late cardiac abnormalities in patients with MIS-C, assessed by standard echocardiography, speckle tracking echocardiography (STE), and cardiac MRI (CMR).

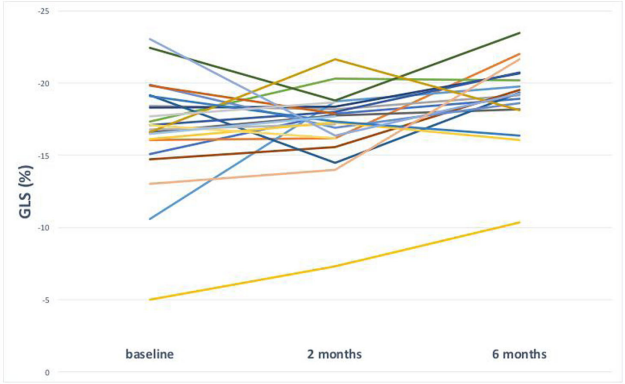
Methods: 32 consecutive patients (21M, 11F), mean age 8.25 ± 4years (range 1.3-17.7), with confirmed MIS-C diagnosis were enrolled in this study. Clinical, laboratory and microbiological data were collected. At disease onset, all children underwent standard transthoracic echocardiography, STE with analysis of left ventricle global longitudinal strain (GLS) and 23 (75%) of them performed CMR. Patients underwent complete cardiological evaluation, including echocardiography and STE at two months (T1) and six months (T2) after diagnosis. CMR was repeated at six months after diagnosis.

Results: Cardiovascular symptoms were present in 45.8% of cases. Thirteen children (40.6%) shared Kawasaki Disease-like symptoms, and 5 (15.6%) needed ICU admission. Early survival was 100%. All patients showed an hyperinflammatory state. Tn-I was elevated in 20 (62.5%) and BNP in 28 (87.5%) patients. Mean LVEF at baseline was 58.8 ± 10% with 10 patients (31%) below 55%. STE showed reduced mean LV GLS (-17.4 ± 4%). On CMR, LGE with nonischemic pattern was evident in 8/23 patients (35%). Follow-up data showed rapid improvement of LVEF at T1 (62.5 ± 7.5 vs. 58.8 ± 10.6%, p value 0.044) with only three patients (10%) below ≤ 55% at T1 and one (4%) at T2. LV GLS remained impaired at T1 (-17.2 ± 2.7 vs.-17.4 ± 4, p value 0.71), and significantly improved at T2 (-19 ± 2.6% vs. -17.4 ± 4%, p value 0.009). LV GLS was impaired (>-18%) in 53% of patients at baseline and T1, while only 13% showed persistent LV GLS reduction at T2. Follow-up CMR showed LGE persistence in 33.4% of cases.

Conclusion: Even though, early cardiac involvement significantly improves during follow-up, subclinical myocardial damage seems to be still detectable 6 months follow up in one third of MIS-C patients.

Keywords: multisystem inflammatory syndrome in children; COVID 19; speckle tracking echo-cardiography; longitudinal strain; cardiac magnetic resonance; myocardial injury

LV GLS improvement during Follow Up



Distribution of GLS values during follow-up

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P-074

Pulmonary perfusion asymmetry in patients after repair of tetralogy of fallot: a 4D flow mri based study

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Background and Aim: Repaired Tetralogy of Fallot (rTOF) patients may have residual lesions such as main (MPA) and branch pulmonary artery stenosis (BPAS). While MPA stenosis is well studied, few data are available on BPAS in rTOF. We aimed to describe pulmonary perfusion in a large paediatric cohort of rTOF and its impact on right ventricular and outflow-tract hemodynamics using 4D flow CMR

Methods: 130 consecutive patients (mean age at CMR 14.3 ± 4.6 years) were retrospectively reviewed.

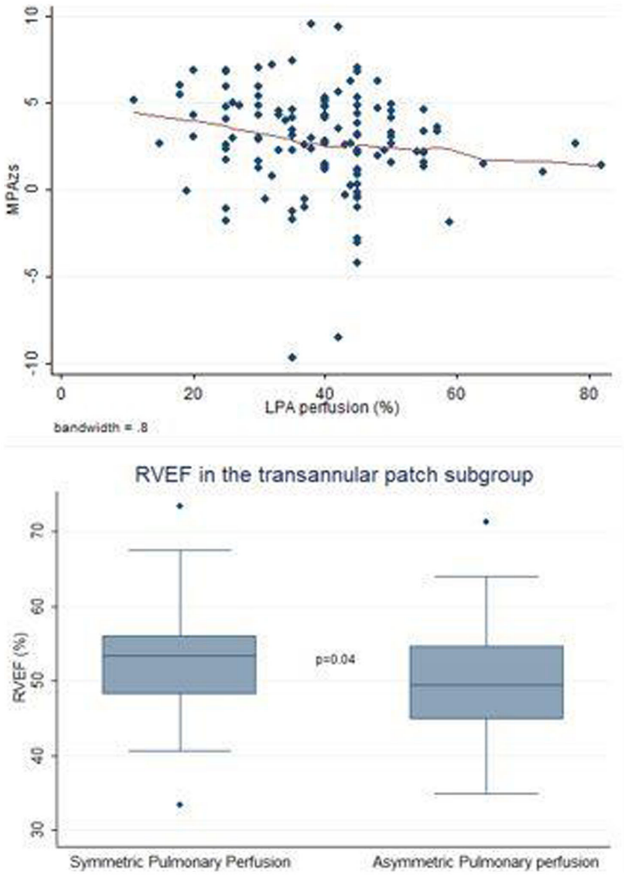
96 patients had transannular patch without valve preservation while 34 patients had conserved annulus or valved conduit. A pulmonary blood flow ratio (right pulmonary artery (RPA)/left pulmonary artery (LPA)) between 0,75 and 1,56 was considered normal.

Results: Asymmetric pulmonary perfusion was present in 59/130 patients (45%), with 54/59 (91%) having left lung hypoperfusion (blood flow ratio >1,56). RPA/LPA perfusion ratio in the whole cohort was independently associated with the LPA Z-score (-0.053, p = 0.007), the RPA regurgitant fraction (RF) (0.013, p = 0.011) and previous LPA stenting (0.648, p = 0,004). Decreasing LPA % perfusion (and conversely increasing RPA % perfusion) was significantly associated with higher MPA diameter Z-score (-0.06, p 0.007). On multivariate analysis, MPA Z-score was independently associated with pulmonary RF (0.48, p<0,001) and with right ventricular indexed volumes (coefficient 3.6, p = 0.023). In patients with transannular patch repair, asymmetric pulmonary flow was an independent predictor of right ventricular ejection fraction (RVEF) (-3.66, p = 0.04).

Conclusion: Pulmonary perfusion asymmetry is frequent in rTOF and is associated with abnormal right ventricular and outflow-tract hemodynamics, including MPA dilatation and decreased RVEF in patients after transannular patch.

Keywords: Tetralogy of Fallot, Cardiac MRI, 4D Flow MRI, pulmonary perfusion

Main RESULTS: correlation between left pulmonary perfusion and MPA Z-score, correlation between right ejection fraction and asymmetric pulmonary perfusion



Decreasing LPA % perfusion (and conversely increasing RPA % perfusion) is associated with a higher MPA Z-score, an independent predictor of both PRF and RV dilatation. Asymmetric pulmonary flow is the only independent predictor of lower RV ejection fraction that we found in transanular patient group

P-077

Semi-automated measurement of the common carotid artery intima media thickness in children: inter- and intra-operator validation study

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Background and Aim: Although atherosclerotic plaques appear in adulthood, atherogenesis starts during childhood. Intima media thickness (IMT) is proven to be a subclinical radiological marker for early atherosclerosis in adults and children, but validation studies for the semi-automated IMT measurement technique lack in children. Specific aim is to study the inter-operator and

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intra-operator correlation of the semi-automated technique of the common carotid IMT on B-mode ultrasound in children. The study will set the grounds for the development of a subclinical radiological marker for atherosclerosis in children.

Methods: We calculated a sample size of 30 patients who were recruited in a prospective study among healthy children presenting to the Radiology Department between November 2020 and March 2021. Semi-automated Math'SR B-mode ultrasound technique was used to measure carotid IMT according to the Manheimm recommendations. Edge detection and quality index were verified for all the 696 measurements. Measurements were done twice by six different operators on both sides. Anthropometric indicators, including blood pressure and carotid artery diameter, were measured. Student t-test and intra-class correlation coefficient (ICC) were performed using SPSS for the inter- and intra-operator variability.

Results: The average age was 12.5 years (range 11-14.9), 46.4% were girls. Twenty-one patients had normal Body Mass Index (BMI) and seven increased BMI (>85th percentile). Two of the obese patients had elevated blood pressure (>95th percentile). Fifty percent of the patients had Tanner stage 4. Intra-operator ICCs for IMT were: 0.87, 0.90, 0.89, 0.90, 0.84 and 0.84 for each of the six operators. Intra-operator ICCs for the ratio IMT/diameter were: 0.68, 0.583 0.81, 0.71, 0.52 and 0.65. Inter-operator ICC was 0.97. The mean IMT calculated for healthy children was 0.51 mm (+/-0.004) and 0.55 mm (+/-0.1) for overweight/obese children.

Conclusion: There is excellent inter- and intra-operator agreement using the semi-automated IMT B-mode technique in children provided operators pay attention to edge-detection. There is no gold standard technique for IMT measurement in children. Further studies are needed to compare the reproductibility with the radiofrequency technique and generate normative charts for both techniques for the same cohort.

This test could guide treatment to prevent disease progression in at risk children.

Keywords: Intima media thickness, ultrasound, pediatrics, cardiovascular imaging

Inter-operator agreement table of the mean IMT measurements according to the 6 operators

Inter-operator Agreement for the Semi-automated Mode B IMT Measurement Technique in Children

ICC	Operator1	Operator 2	Operator 3	Operator 4	Operator 5	Operator 6
Operator 1	1	-	-	-	-	-
Operator 2	0,92	1	-	-	-	-
Operator 3	0,95	0,95	1	-	-	-
Operator 4	0,91	0,95	0,96	1	-	-
Operator 5	0,91	0,91	0,85	0,83	1	-
Operator 6	0,97	0,96	0,96	0,94	0,93	1

P-078

Pulse wave velocity measurements by magnetic resonance imaging in neonates and adolescents; methodological aspects and their clinical implications

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Background and Aim: Pulse wave velocity (PWV) by cardiovascular magnetic resonance (CMR) imaging is a surrogate marker for aortic stiffness. Clinical use has been hampered by lack of standardization. The overall aim of this study was to investigate methodological aspects of PWV measurements by CMR, as a means towards standardization and increased reproducibility in neonates and adolescents.

Methods: A computer phantom was created to validate the temporal resolution required for accurate PWV. Fifteen neonates and 71 adolescents underwent CMR with non-contrast enhanced 3D angiography for aortic length measurements and phase-contrast flow acquisitions in the ascending and descending aorta (Figure 1). A subset of participants had coronal overview images assessed for aortic length. Velocity and flow curves, transit time methods (time-to-foot (TTF), maximum upslope, and time-to-peak (TTP)), and baseline correction methods (no correction, automatic and manual) were investigated.

Results: For neonates, required timeframes per cardiac cycle for accurate PWV was 42 for the aortic arch and 41 for the thoracic aorta. For adolescents, corresponding values were 39 and 32. Variation in aortic lengths by overview images and reference standard 3D angiography in adolescents were -16–18mm (aortic arch) and -25–30mm (thoracic aorta). Agreement in PWV between automatic and manual baseline correction was -0.2 ± 0.3m/s for neonates and 0.0 ± 0.1m/s for adolescents. Velocity and flow-derived PWV measurements did not differ in neonates or adolescents (all $p>0.08$). For neonates, transit time methods did not differ (all $p>0.19$) but in adolescents PWV was higher for TTF (3.8 ± 0.5m/s) and maximum upslope (3.7 ± 0.6m/s) compared to TTP (2.7 ± 1.0m/s; $p<0.0001$). Interobserver variability for PWV was 0.0 ± 0.1m/s, corresponding to a PWV error of $-1 \pm 2\%$.

Conclusion: This study provided required temporal resolution for phase-contrast flow acquisitions in both neonates and adolescents for their typical heartrates. This study supports the use of 3D angiography for length measurements and time-to-foot with automatic baseline correction for accurate PWV measurements to

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increase comparability and standardization both for clinical use and research.

Keywords: Neonates, Adolescents, Aorta, Pulse wave velocity, Cardiovascular magnetic resonance imaging.

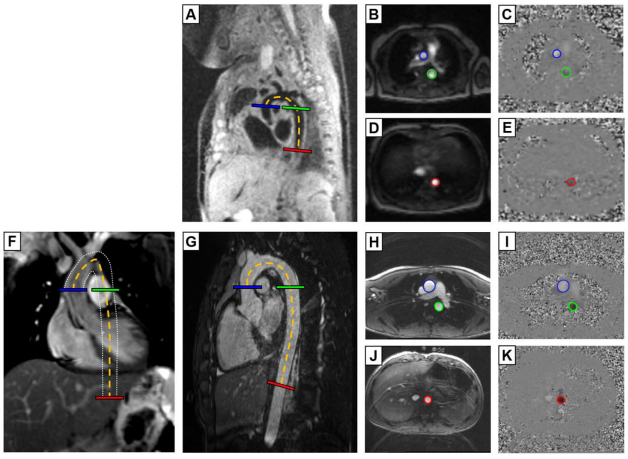

Figure 1. Image delineations for aortic length and flow measurements. Neonatal black-blood (A) and adolescent white-blood (G) 3D angiography, and adolescent thoracic coronal overview image with white dotted lines outlining the aortic walls throughout the 2D image stack (F), all with length delineation (Δd) (dashed orange) and perpendicular flow imaging planes in the ascending aorta (blue), descending proximal aorta (green) and descending aorta at the level of the diaphragm (red). Magnitude and quantitative phase-contrast images at ascending and proximal descending aorta (B-C, H-I) and descending aorta at the level of the diaphragm (D-E, J-K), corresponding to the flow imaging planes localizations. Worth noting is that both the ascending and descending flow plane in the aortic arch (B-C, H-I) is collected in a single flow plane as in clinical routine which may result in angular error in the descending aorta, whereas the flow plane at the level of the diaphragma is perpendicular.

P-079

Left ventricular strain and strain rate during submaximal semisupine bicycle exercise stress echocardiography in childhood cancer survivors

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Background and Aim: Childhood cancer survivors (CCSs) show relevant cardiac morbidity and mortality throughout life. Early detection is key for optimal support of patients at risk. 2D-Speckle-tracking strain echocardiography (STE) and stress echocardiography may help identifying subclinical cardiac dysfunction. Aim of this study was to evaluate 2D-STE strain analysis during semisupine exercise stress in CCSs for detection of subclinical left ventricular (LV)-dysfunction after cancer treatment.
Methods: 77 CCSs ≥ 1-year post chemotherapy were prospectively examined according to a standardized stress protocol at rest, low and submaximal stress level and compared to a previously established cohort of healthy adolescents and young adults (n = 50). Image acquisition was optimized for strain analysis. Global longitudinal strain (GLS), short axis circumferential strain (CS) and corresponding strain rates (SR) were analyzed using vendor-independent software.
Results: CCSs at median 7.7 years post chemotherapy showed comparable LV GLS, CS and SR values during rest, low and submaximal exercise stress level to healthy controls. Yet, prevalence of abnormal GLS (defined as <2 SD of controls reference) in CCSs was 1.3 % at rest, 2.7 % at low and 8.6 % at submaximal stress. In CCSs relative change of CS from rest to submaximal stress was lower than in healthy controls, median 16.9 (IQR 3.4;28.8) % vs. 23.3 (IQR 11.3;33.3) %, p = 0.03, most apparent in the subgroups of CCSs after high-dose anthracycline treatment and cancer diagnosis before age of 5 years.
Conclusion: In this prospective 2D-STE strain study prevalence of abnormal LV-GLS increased with stress level reflecting impaired cardiac adaption to exercise stress in some CCSs. However, relatively early after last chemotherapy this did not result in significant differences of mean GLS-, CS- and SR-values between CCSs and controls at any stress level.

Keywords: Cardio-Oncology, 2D-Speckle-Tracking Strain Echocardiography, Stress Echocardiography

P-080

Paediatric brain mri findings following congenital heart surgery: a systematic review

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Background and Aim: This systematic review aimed to establish the relative incidence of new post-operative brain MRI findings following paediatric congenital cardiac surgery.
Methods: To distinguish perioperative changes from pre-existing MR findings, our systematic search strategy focused on identifying original research studies reporting both pre- and post-surgery brain MRI scans. Patient demographics, study methods, and brain MR findings were extracted.
Results: Twenty-one eligible publications, including two case-control, and one randomised controlled trial were identified. Pre-existing brain MRI findings were noted in 43% (513/1205) of neonates prior to surgery; mainly white matter injuries (WMI). Surgery was performed at a median age of 8 days with comparison of pre- and post-operative MR scans revealing additional new post-operative findings in 51% (550/1075) of patients; mainly WMI. Four studies adopted a brain injury scoring system, but the majority did not indicate the severity or time course of findings. In a sub-group analysis approximately 32% of patients with pre-existing lesions went on to develop additional new lesions post-surgery. Pre-existing findings was not found to confer

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a higher risk of acquiring brain lesions post-operatively. No evidence was identified linking new MR findings with neurodevelopmental delay.
Conclusion: This review and meta-analysis suggests that surgery approximately doubles the number of patients with new brain lesions.

Keywords: Paediatric, congenital heart surgery, CHD, brain lesion, brain MRI

P-081
Early signs of myocardial remodeling in children with multisystem inflammatory syndrome associated with COVID-19 and cardiac involvement

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Background and Aim: Multisystem Inflammatory Syndrome in Children (MIS-C) associate with Coronavirus disease – 19 is a life-threatening clinical condition in which cardiovascular system is frequently affected. Shock, cardiac arrhythmias, myocarditis, reduced left ventricular ejection fraction (LVEF), pericardial effusion, and coronary artery dilatation are amongst the most common cardiac complications. In this study, we aim to assess myocardial status in patient with cardiac involvement in MIS-C.
Methods: Over a 14-month period, we retrospectively collected clinical, biological, echocardiographic data in children who were admitted to our hospital with a diagnosis of MIS-C and cardiac involvement. WHO criteria for clinical case definition of MIS-C were adopted. Elevation in brain-natriuretic-peptide and troponin-I, electrocardiographic abnormalities, echocardiographic evidence of pericarditis, myocarditis, reduced LVEF, valvular disease, and coronary artery dilatation were including criteria. LV indexed end-diastolic (EDVi), end-systolic (ESVi), stroke volumes were measured with Cardiac Magnetic Resonance (CMR). T2 mapping, Cine-RM and late gadolinium enhancement studies were performed.
Results: 14 children were identified and included in the study, 71% of which were male. Median age at disease onset was 7 years old (IQR 5 to 9 years). All patients underwent cardiological evaluation in the first 48 hours of hospital staying. LVEF was <45% in 28.6% and <35% in 14.3% of patients. Myocarditis was detected in 78.6%, pericarditis in 28.6%, valvular damage in 35.7%, coronary abnormalities in 42.9%. All patients underwent CMR after on average 4 months (median: 3.87, IQR 2 to 4) from disease onset, after full clinical and biological recovery. ESVi and stroke volumes resulted within normal range in 100%. CMR abnormalities were observed in 21%. Particularly, left ventricular EDVi resulted elevated in 7%, delayed washout in T2 was described in 7%, and increased T2 mapping in 7%.
Conclusion: Despite complete clinical and biological resolution, increased EDVi, delayed washout in T2 and increased T2 mapping at follow-up CMR in patient with cardiac involvement due to MIS-C may be signs of myocardial remodeling.

Keywords: MIS-C, COVID-19, Cardiac Magnetic Resonance, myocardial remodeling

P-082/Moderated Poster
Neonatal cardiac, feed-and-wrap, free-breathing 4D flow MRI is comparable to standard free-breathing 2D flow assessment

Hannah Panayiotou[1], Lily Mills[1], David Broadbent[2], David Shelley[1], Jutta Scheffczik[3], Alexandra M Olaru[4], Ning Jin[5], John P Greenwood[1], Helen Michael[6], Sven Plein[1], Malenka M Bissell[7]
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Fondazione Policlinico Universitario A.Gemelli IRCCS
Background and Aim: Traditionally, neonatal cardiovascular magnetic imaging (MRI) requires a general anaesthetic for breath-holding. However, free-breathing 4DFlowMRI now allows targeted neonatal feed-and-wrap scans for comprehensive haemodynamic and anatomical assessment in under 10 minutes. This validation study examines the practicability of neonatal free-breathing feed-and-wrap MRI without using contrast agent, in terms of comparability to standard 2D imaging, and clinical reproducibility.
Methods: 13 neonates underwent non-contrast feed-and-wrap MRI scan on a 3T clinical scanner (MAGNETOM Prisma 3T, Siemens Healthcare, Erlangen, Germany) using a dedicated 18 channel variety coil (NORAS MRI products GmbH, Hoechberg, Germany). 5 were scanned in an open-top bassinet, and 8 in the MRI-compatible incubator (LMT Medical Systems GmbH, Luebeck, Germany) facilitating seamless transfer from the neonatal unit into the scanner; this allows a completely sedation-free process. The imaging protocol included compressed sensing accelerated 4DFlowMRI* and additional free-breathing binning cine imaging if the neonate stayed asleep.
Data was analysed using PIE medical imaging software (CASS, The Netherlands). Inter-and intra-observer variability of 4DFlowMRI measurements was assessed.

* prototype sequence
Results: All neonates scanned in the incubator completed the protocol, whilst only 2/5 (40%) completed the protocol in the bassinet. Forward flow measurements were comparable between standard 2D phase contrast vs 4DFlowMRI with a bias of 0.15 and 95% limits of agreements -0.62 to 0.92. Mean forward flow measured by 2D phase contrast was 4.95ml/s (± 1.96ml/s) and 4DFlowMRI was 4.81ml/s (± 1.82ml/s).
4DFlowMRI quantification was also reproducible, demonstrated by excellent inter-observer agreement for aortic forward flow (ICC 0.98; 95%CI 0.92-0.99) as well as excellent intra-observer agreement (ICC 0.98; 95%CI 0.91-0.99). Multiple measurements also show consistency, with main pulmonary artery flow agreeing with aortic flow (ICC 0.94; 95%CI 0.76-0.99), the sum of branch pulmonary artery flow agreeing with main pulmonary artery flow (ICC 0.89; 95%CI 0.62-0.99), and descending aorta plus superior vena cava flow agreeing with aortic flow (ICC 0.99; 95%CI 0.99-1).
Conclusion: This sedation-free neonatal feed-and-wrap MRI is well tolerated and achievable, with 100% of neonates completing

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the protocol in the incubator. Importantly, non-contrast 4DFlowMRI quantification is comparable to standard validated 2D phase contrast free-breathing imaging, and is reproducible, with high inter-observer and intra-observer agreement.

Keywords: 4D Flow MRI, neonatal, feed and wrap, flow

Neonatal feed-and-wrap MRI

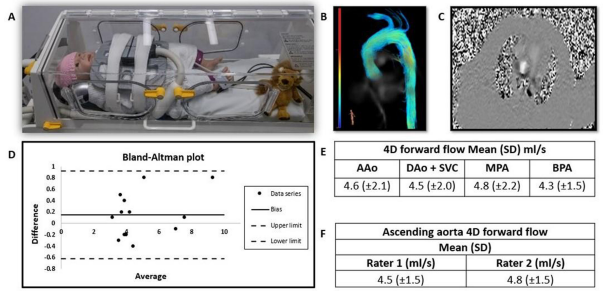

Figure 1: A) MRI-compatible incubator with specialised small coils inside; B) 4D Flow MRI anatomical aortic arch 3D reconstruction; C) 2D Flow MRI ascending aorta at the level of the pulmonary arteries; D) Bland-Altman plot, bias 0.15, 95% limits of agreement -0.62 to 0.92; E) 4D forward flow measurements; AAo, ascending aorta; DAo, descending aorta; SVC, superior vena cava; MPA, main pulmonary artery; BPA, branch pulmonary arteries; F) Ascending aorta 4D flow between two raters.

P-083

Evaluation of cardiac findings in five cases diagnosed with abernethy syndrome

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Background and Aim: Abernethy syndrome (AS), known as congenital extrahepatic portosystemic shunt, is a rare clinical picture in which splanchnic blood is drained into systemic veins through a shunt between the portal and systemic circulation. AS may be associated with cardiovascular, gastrointestinal, genitourinary and skeletal system anomalies.
Methods: In this report, our cases with a diagnosis of AS accompanied by cardiac pathologies, are presented.
Results: Case-1: A 2-year-old girl with Down syndrome was operated for ventricular septal defect. In cardiac catheterization and angiography (CCAG), it was observed that the contrast agent passed from the aneurysmatic portal vein (PV) to the inferior vena cava (IVC) with a fistula. PV and IVC pressures were measured as 14 mmHg and 11 mmHg, respectively. Because the patient was asymptomatic and the portal system pressure was high, follow-up was planned without any interventions.

Case-2: A 15-year-old girl with chronic liver disease and diffuse pulmonary fistulas in the left lung had a history of atrial septal defect (ASD) closure.

Case-3: In CCAG of a 10-year-old girl whose echocardiography (ECO) findings were compatible with polysplenia syndrome (Left atrial isomerism), pulmonary arterio-venous fistulas, IVC interruption, and continuity with hemiazygous vein were detected.
Liver and lung transplantation could not be performed in cases 2 and 3 due to the high perioperative mortality risk.

Case-4: In the balloon occlusion test performed during CCAG to a 15-year-old boy with pulmonary hypertension (PHT) findings, portal vein (PV) pressure exceeded 20 mmHg. Therefore, the portocaval shunt was not closed with percutaneous transcatheter or surgery.

Case-5: Small secundum ASD was observed in the ECO of a 1.5-year-old male patient with Down syndrome, a history of prematurity and elevated liver function tests. Dyspnea was prominent in the physical examination of all cases except case 1. In all cases, portosystemic shunt was detected in PV doppler ultrasonography and abdominal CT angiography examinations.
Conclusion: Since AS can be asymptomatic in childhood, it may not be diagnosed until adulthood. However, AS may be detected during routine cardiac evaluation when investigating the etiology of congenital heart disease or PHT. On the other hand, cardiac screening should be kept in mind in pediatric patients with AS.

Keywords: Abernethy syndrome, cardiac anomaly, portosystemic shunt, pulmonary hypertension

P-084

Assessing medium term cardiac outcomes in children with multi-system inflammatory syndrome

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Background and Aim: Multi-system inflammatory syndrome in children (MIS-C) causes widespread systemic inflammation including a pancarditis in the weeks following a COVID infection. Further coronavirus surges appear inevitable and with vaccination rates lower in young people an understanding of the medium-term cardiac impacts of this condition is important for planning further treatment and understanding the impacts on their health.
Methods: A retrospective single-center study of 67 consecutive patients with MIS-C was performed. Three time points were determined as the point of worst cardiac dysfunction during the acute admission, then at intervals of 6-8 weeks and 6-8 months. Echocardiographic findings were used to evaluate both 2D and 3D measures of cardiac function. Coronary artery measurements were recorded. Corresponding serial ECG findings were evaluated.
Results: The worst cardiac function arose 6.8 ± 2.4 days after the onset of fever. The mean M mode-derived FS was 30.9 ± 8.1% during the acute phase. The mean 3D left ventricle (LV) ejection fraction (EF) was borderline at 50.5 ± 9.8%. A pancarditis was typically present: 46.3% showed cardiac impairment; 31.3% had some pericardial effusion; 26.8% had moderate (or worse) valvar regurgitation and; 26.8% had coronary dilatation. Cardiac function returned to normal in all patients by 6-8 weeks (mean 3D LV EF 61.3 ± 4.4%, $p<0.001$ compared to admission). Coronary dilatation normalized in all but one patient who initially developed large aneurysms at presentation; these continued 6 months later. ECG findings mainly featured T-wave changes resolving at follow-up. There were a small number of adverse events: need for

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ECMO (2), death as an ECMO-related complication (1), subendocardial infarction (1), LV thrombus formation (1).
Conclusion: MIS-C causes a pancarditis with decreased cardiac function and almost a quarter of patients showing coronary changes. In most, discharge from long-term follow-up can be considered as full cardiac recovery is expected by 8 weeks. The exception includes patients with medium sized aneurysms or greater or those with more of a Kawasaki disease phenotype as these require on-going surveillance for persistence of coronary changes.

Keywords: COVID-19, Myocarditis, coronary aneurysms

P-085/Moderated Poster
Validation of various approaches to aorta, pulmonary artery and pulmonary branches measurements in CT scans

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Background and Aim: Computed tomography plays an important role in diagnostic and treatment planning in patients with congenital heart diseases. The aim was to evaluate sources of variability in the measurements of the main thoracic arteries as possible factors influencing study interpretation.
Methods: A total of 755 chest CT scans were done on children between 2015–2020. Based on diagnosis possibly affecting the vessels we excluded: congenital heart defects (141), substantial mediastinal masses, pneumothorax or hydrothorax, chest deformation (49), motion artifacts (7) and lack of anthropomorphic data (234). Statistical software was Wizard 2.0.10.
Results: Eligible for analysis were 324 studies and 304 had complete measurements. There were 20 neonates and infants, 70 patients 1-5 y.o., 73 5-10 y.o., 51 10-15 y.o., and 78 15-18 y.o. The youngest group was excluded from further analysis as underpowered. Seventy-eight (28.1%) studies were done without contrast medium, 200 (71.9%) were contrast enhanced (CE) on SOMATOM Definition Flash (Siemens) and LightSpeed VCT (GE) scanners. The measurements were done by two experienced radiologists in the orthogonal axial plane and repeated after plane correction based on MPR views to adjust for the vessel axis. Aorta (Ao) was measured in sagittal and coronal diameter at the main pulmonary artery (MPA) division level. MPA and its left and right branches (LPA and RPA) were measured once at mid-length. There was excellent agreement between radiologists with interclass correlation with the least ICC coefficient for the RPA diameter – 0.884 and for the rest ranging from 0.974 to 0.994 for singular measurements, and for the averages 0.915 and 0.987 to 0.997 respectively. Cross-sectional plane adjustment analysis revealed no difference for Ao coronal diameter (-0.05 ± 2.9 mm, $p = 0.729$), but slightly different Ao sagittal diameter (-0.3 ± 1.5 mm, $p = 0.002$) and MPA (0.4 ± 2.8 mm, $p = 0.010$). For the analysis of contrast enhancement (CE) effect on the measurements patients older than 5 y.o. were included so that they were no different on baseline characteristics (i.e. age, height, weight, BSA. No statistically significant differences were found for any of the measurements (p 0.065 to 0.781).
Conclusion: Following consistently the methods applied in the making of the reference values is necessary to keep the study interpretation valid and reproducible.

Keywords: aorta, pulmonary artery, pulmonary branches, measurements, CT, Computed tomography

P-086
Left ventricular longitudinal strain in asymptomatic or mildly symptomatic children and young adults with SARS-COV-2 infection

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Background and Aim: Clinical manifestations of children's coronavirus disease-2019 (COVID-19) are generally considered less severe compared with adult patients. The objective of this study was to evaluate cardiac involvement in healthy children with asymptomatic or mildly symptomatic severe acute respiratory syndrome coronavirus-2 (SARS-CoV-2) infection.
Methods: We analysed a cohort of 210 paediatric patients, mean age 7 ± 4 years, who had a confirmed diagnosis of SARS-CoV-2 infection and were asymptomatic or mildly symptomatic for COVID-19. Patients underwent standard transthoracic echocardiogram and speckle tracking echocardiographic study 138 ± 65 days after diagnosis. Seventy-two age, sex, and body surface area comparable healthy subjects were used as control group.
Results: Left ventricular ejection fraction was within normal limits but significantly lower in the cases group compared to controls (62 ± 4% vs. 65 ± 5%; $P = 0.012$). Left ventricular (LV) global longitudinal strain (- 20,91 ± 2,83 % vs. - 22,73 ± 2,51 %; $P < 0.001$) was significantly reduced in cases compared with CTRLs. Regional LV strain analysis showed a significant reduction of the LV mid-wall segments strain among cases compared to controls. Furthermore, in the cases group, there were 25% of subjects with a regional peak systolic strain below -16% (-2.5 Z score in our healthy cohort) in at least two segments. These subjects did not show any difference regarding symptoms or serological findings.
Conclusion: SARS-CoV-2 infection may affect left ventricular deformation in 26% of children despite an asymptomatic or only mildly symptomatic acute illness. A follow-up is needed to verify the reversibility of these alterations and their impact on long-term outcomes.

Keywords: COVID-19, Speckle-tracking analysis

P-087
Can ECHO simulation assist paediatric & neonatal specialists to diagnose acute presentations of complex congenital heart disease? a systematic review

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Background and Aim: Across Europe, recent policy has developed the role of local paediatrician-delivered echocardiography. Thanks to high commitment to learning, agreement of international standards, strong local networks and the EACVI accreditation process, such scans are typically performed with high quality. Nevertheless, the volume of acute and complex cases encountered in local hospitals is naturally lower than within tertiary cardiology units. A training gap is therefore foreseen to

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support these professionals, both to initially develop their skills in this area and to maintain their skills in the years following accreditation.

Methods: A systematic literature review was performed using the PubMed database, EMBASE and Google Scholar. Three search-term groups were developed, each summarized by the terms 'congenital heart disease', 'echocardiography' and 'simulation'. All peer-reviewed articles were included, without date, language or geographic restrictions. Studies were assessed for relevance i) to congenital echocardiography simulation for paediatricians and neonatologists and ii) to outcome measures assessing diagnostic accuracy. Study quality was further assessed using the CEBM 'Level of Evidence' tool.

Results: The search identified 406 studies, of which 7 met pre-agreed inclusion criteria. Five papers were highly relevant to simulation of congenital heart disease outside of a tertiary setting, whilst two others solely demonstrated content and construct validity. Learners in most studies were classed as 'novices' to congenital echo, limiting inferences to paediatricians who have already reached accreditation-equivalent standard. Four papers assessed clinically-relevant outcome measures before and after simulation training. These documented improvements in factual and applied knowledge, probe kinematics, quality of ECHO images, and diagnostic accuracy of new simulated cases. No paper was sufficiently powered to assess effects upon subsequent clinical practice.

Conclusion: Current evidence supports the validity of ECHO simulators as training and assessment tools for diagnosis of complex congenital heart disease, irrespective of previous learner experience. Potential for use within pre-planned learning sessions is also demonstrated, dramatically reducing the cost of real patient demonstrators and offering rare experience in scanning acute and complex lesions. Although simulation has clear benefits in ECHO performance for novices, the effect on more experienced practitioners is not specifically tested. Further investigation is planned to assess the role of simulation in this group.

Keywords: ECHO, simulation, paediatrician, congenital heart disease

P-088

What is the normal range for right ventricular global longitudinal strain in the term neonate?

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Background and Aim: Right ventricular (RV) function plays an important role in various common neonatal conditions, yet its quantitative assessment is complicated by a number of factors. Classical markers of RV function for example are highly sensitive to loading conditions, which change rapidly during normal neonatal transitional physiology. Attempts have been made to assess normative values for neonatal left ventricular speckle-tracking (STE) derived strain, yet no systematic reviews are published for the right ventricle in neonates. This study aims to review available normative datasets for STE-derived RV systolic strain.

Methods: A systematic review was performed using two databases. Search terms were based around word groups summarised by 'neonate', 'echocardiography' and 'strain'. Inclusion criteria allowed any study describing postnatally-acquired STE-derived RV global longitudinal strain (GLS) or systolic strain rate (GLSr), measured from the apical 4-chamber view, in healthy term neonates. All data were reported as positive values. Mean values and confidence intervals (CI) were calculated using a random effects model.

Results: The search identified 254 studies, of which 11 were suitable for inclusion. These described 380 relevant participants. Ten studies were performed prospectively, with seven designed specifically to produce normative data and the remainder taken from 'healthy' control groups of wider studies. Most utilised 2-dimensional imaging, with one using 3-dimensional echocardiography. No significant change was noted during the initial 28 days of life across all studies (figure), though individual studies typically reported significant increase in RV-GLS on follow-up after the first 48 hours of life. Ten reported RV-GLS, totalling 14 time points from birth, with mean average 21.4% (95%CI:19.3-23.5). Five studies reported systolic RV-GLSr, including 6 time points, with mean 1.6/sec (95%CI:1.1-2.1). No study reported a significant effect of residual patent ductus arteriosus or patent foramen ovale on measured strain.

Conclusion: We report a first systematic review of RV STE-derived values of strain and strain rate in healthy term neonates. Despite substantial changes in normal right ventricular haemodynamics, we demonstrate a non-significant trend towards increase in RV-GLS and RV-GLSr across the first four weeks of life. Subgroup analysis suggests that use of different processing software and 3-dimensional imaging were major drivers of heterogeneity between studies.

Keywords: Neonate, strain, speckle-tracking, right ventricle, function

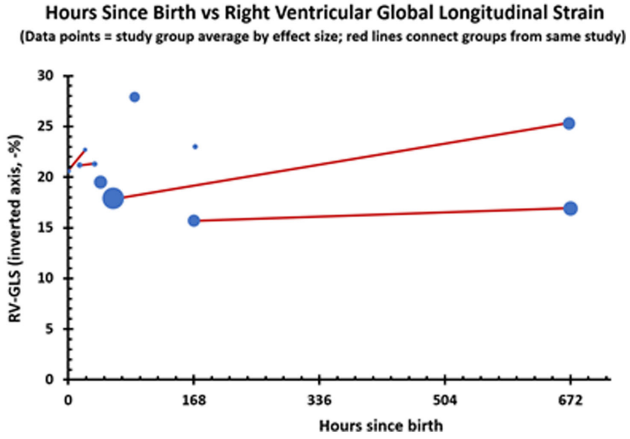

Figure 1 Scatter plot of average reported values of RV-GLS for healthy term neonates in various published study groups. Describes average timing of the measurement (hours) since birth against measured RV-GLS (-%). Size of datapoints equate to relative effect size. Red lines connect two time points of groups followed-up within the same study.

P-089

Left atrial phasic function and atrioventricular coupling in healthy adolescents at exercise under normoxia and altitude simulation

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Background and Aim: Left atrial phasic function can give insight into heart function, especially the diastolic function. Little is known

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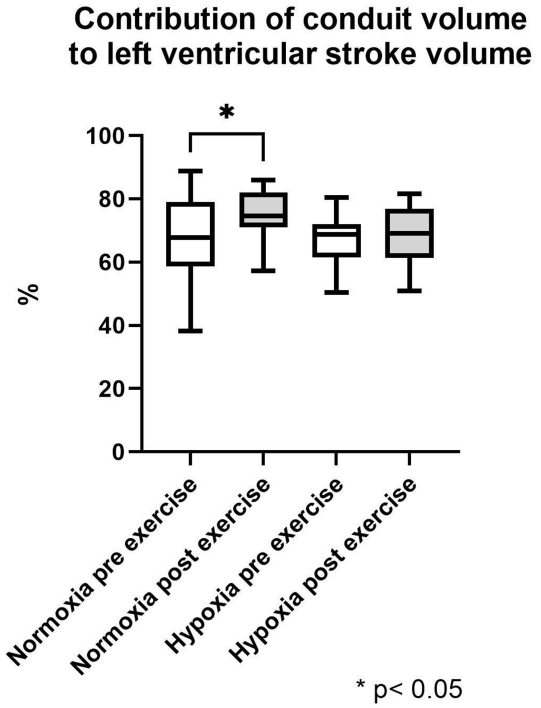
about possible changes of this function during exercise especially under hypoxic conditions simulating altitude. We sought to study the contribution of left atrial phasic function to left ventricular filling before and after exercise testing under normoxia and hypoxia by three-dimensional echocardiography (3DE).
Methods: 19 healthy adolescents and young adults (median 15 years) performed a cycle ergometric test under normoxia and under hypoxia in a normobaric altitude chamber (ambient oxygen saturation 15.2%, corresponding to 2,500 m asl). 3DE was done before and after exercise both under normoxia and hypoxia thus creating four timepoints. 3DE-data sets where analyzed by TomTec 4D LV-Analysis software 3.1. Volume-time curves of the left atrium (LA) and left ventricle (LV) were generated. Conduit volumes (calculated as LV stroke volume – LA emptying volume), passive and active emptying volumes were assessed as well as their contribution to the filling of the LV. One-way ANOVA and Tukey's multiple comparison post hoc test was used to compare timepoints.
Results: Conduit volume contributed most to LV stroke volume at all timepoints. Under normoxia, contribution of conduit volume to LV stroke volume increased significantly after exercise compared to resting conditions. This increase after exercise could not be observed under hypoxia (Figure). Contributions of passive and active emptying did not differ significantly between the timepoints but a tendency of less active emptying contribution after exercise under normoxia could be seen.
Conclusion: Conduit volume contributes most to LV stroke volume under normoxia and hypoxia. Increased conduit flow after exercise under normoxia can be attributed to enhanced diastolic properties of the LA and the LV, e.g. LA compliance, LV relaxation and compliance. That we cannot observe this effect under hypoxia might be a sign of altered diastolic properties under hypoxia. More research is needed to further investigate this observation.

Keywords: left atrial phasic function, atrioventricular coupling, altitude simulation, hypoxia, exercise test



Contribution of conduit volume to left ventricular stroke volume pre and post exercise testing during normoxia and hypoxia

P-090

Spirometry-based reconstruction of real-time cardiac mri: motion control and quantification of heart-lung interactions

Lena Maria Röwer[1], Tobias Uelwer[3], Janina Hußmann[1], Halima Malik[2], Monika Eichinger[4], Dirk Voit[5], Mark Oliver Wielpütz[4], Jens Frahm[5], Stefan Harmeling[3], Dirk Klee[2], Frank Pillekamp[1]
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Background and Aim: Physiological conditions in combination with the best available methods to quantify physiological parameters are necessary to judge heart-lung interactions and to guarantee meaningful cardiac functional examinations. Real-time magnetic resonance imaging (MRI) offers the opportunity to replace the conventional breath-hold technique by imaging during free-breathing. Spirometry is the gold standard to measure pulmonary ventilatory function and can be performed during MRI. To ensure physiological conditions, we tested the feasibility of cardiac real-time MRI in combination with retrospective gating by MR-compatible spirometry, to improve motion control and allow quantification of respiratory-induced changes during free-breathing.
Methods: Cross-sectional real-time MRI (1.5 T; 30 frames/second) using balanced steady-state free precession contrast during free-breathing was combined with MR-compatible spirometry in healthy, adult volunteers (n = 4).
Retrospective binning assigned images to classes that were defined by ECG and spirometry. Left ventricular eccentricity index (EI) as an indicator of septal position and ventricular volumes in different respiratory phases were calculated to assess heart-lung interactions.
Results: Real-time MRI with MR-compatible spirometry is feasible and well tolerated. Spirometry-based binning improved motion control significantly. The end-diastolic epicardial EI increased significantly during inspiration (1.04 +/- 0.04 to 1.19 +/- 0.05; $p<0.05$). During inspiration, right ventricular end-diastolic volume (79 +/- 17 ml/m² to 98 +/- 18 ml/m²), stroke volume (41 +/- 8 ml/m² to 59 +/- 11 ml/m²) and ejection fraction (53 +/- 3% to 60 +/- 1%) increased significantly whereas the end-systolic volume remained almost unchanged. Left ventricular end-diastolic volume, left ventricular stroke volume, and left ventricular ejection fraction decreased during inspiration while the left ventricular end-systolic volume increased. The relationship between stroke volume and end-diastolic volume (Frank-Starling relationship) based on changes induced by respiration allowed for a slope-estimate of the Frank-Starling curve to be 0.9-1.1.
Conclusion: Real-time MRI during free-breathing combined with MR-compatible spirometry and retrospective binning improves image stabilization, allows quantitative image analysis, and most importantly, offers unique opportunities to judge heart-lung interactions.

Keywords: free-breathing examinations, real-time imaging, cardiac function, diagnostic imaging, magnetic resonance imaging, spirometry

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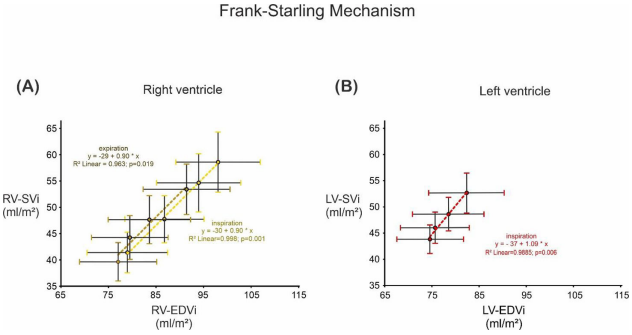

When the end-diastolic volume of the ventricles was modified significantly by respiration (Δ EDVi >5 ml/m^2), the relationship between stroke volume and end-diastolic volume could be analyzed. Linear regression revealed highly significant relationships for (A) the right ventricle, and (B) the left ventricle during inspiration. Circles indicate mean value, whiskers, standard error of the mean. ⋆ $p<0.05$ (linear regression). The unit of x in the linear equations is ml/m^2.

P-092

4D flow mri derived kinetic energy parameters change with age in the tetralogy of fallot disease process

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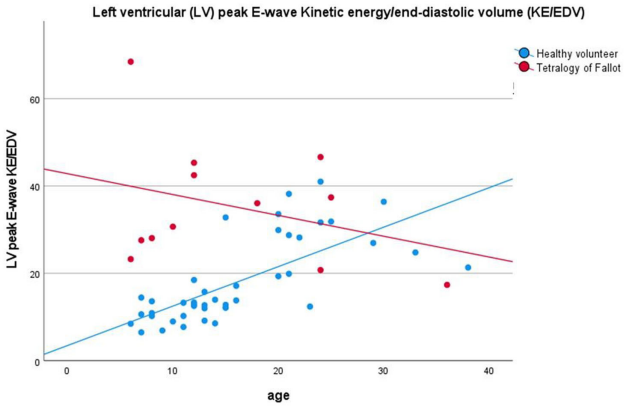
Background and Aim: Optimal timing for pulmonary valve replacement (PVR) in repaired tetralogy of Fallot (rTOF) patients was traditionally based on right ventricular size which poorly correlates with long term mortality and morbidity. Functional parameters especially in the left ventricle (LV) have shown more promise in predicting long term outcome. The primary aim of this study was to assess if ventricular kinetic energy (KE) changes with age are different in rTOF patients compared to healthy volunteers and would therefore potentially be a useful imaging biomarker.
Methods: 55 participants [healthy volunteers (HV) (n = 43, age range 6-38 years); rTOF (n = 12, age range 6-37 years] were prospectively recruited and underwent 4D Flow MRI⋆ and standard cine imaging on 3T clinical scanners (MAGNETOM Trio and MAGNETOM Prisma, Siemens Healthcare, Erlangen, Germany); The following kinetic energy (KE) parameters were extracted using a research analysis software MASS: peak E-wave; peak A-wave; minimum diastolic; maximum systolic. Parameters were indexed to end-diastolic volume (EDV).
⋆prototype sequence
Results: rTOF patients had higher mean LV peak E-wave KE/EDV compared to HV (35 ± 14 vs 18 ± 10 μJ/mL; p = 0.001), lower LV maximum systolic KE/EDV (18 ± 6 vs 29 ± 11 μJ/mL; p<0.001) and lower LV peak A-wave KE/EDV (9 ± 6 vs 22 ± 14 μJ/mL; p<0.001). LV peak E-wave KE/EDV increased with age (r = 0.702) in HV whereas it decreased in rTOF (r = -0.326). This remained significant when indexing E-wave KE to body surface area (HV r = 0.660; rTOF r = -0.157) or stroke volume (HV r = 0.677; rTOF r = -0.208). LV EDV in comparison increased similarly with age in both groups (HV r = 0.653; rTOF r = 0.704).
Conclusion: LV peak E-wave KE/EDV decreases with age in rTOF whereas it increases in childhood in healthy volunteers. This distinct difference suggests that E-wave KE as an imaging biomarker has the potential to help inform timing of PVR and warrants evaluation in larger longitudinal studies.

Keywords: 4D Flow MRI, Tetralogy of Fallot, pulmonary valve replacement



Left ventricular E-wave kinetic energy indexed to end diastolic volume increases with age in healthy volunteers and decreases with age in patients with repaired Tetralogy of Fallot.

P-093

Orthostatic tolerance of paediatric fontan patients and the effect of enalapril treatment

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Background and Aim: It is still unclear how Fontan patients respond to orthostatic stress, especially because the cardiovascular mechanisms that control adaptation to orthostasis may be impaired in these patients. Although many Fontan patients are often treated with angiotensin converting enzyme inhibitors, they may negatively affect orthostatic tolerance and lead to hypotension or reduction of cardiac index (CI). This study aims to assess the orthostatic stress response of paediatric Fontan patients and to determine the effect of enalapril treatment.
Methods: For this study Fontan patients with an extracardiac conduit without pre-existent enalapril treatment were included. Patients were treated with enalapril for 3 months and before and after treatment, cardiovascular response to head-up tilt test

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was evaluated by different parameters including blood pressure, CI, cerebral blood flow, aortic stiffness, and cardiac vagal activity. Healthy controls were included for baseline comparison.
Results: 35 Fontan patients (median age 14.0 years, 5-15 years after Fontan palliation) and 34 healthy controls (median age 12.8 years) were included. At baseline, patients showed a similar CI and vagal activity, but a higher systolic blood pressure and aortic stiffness and lower cerebral blood flow compared to controls. None of the Fontan patients had imminent syncope during head-up tilt while 3 healthy controls (9%) did. During tilt, almost all parameters changed with a similar magnitude in patients and controls, only the percentage increase of systolic blood pressure was lower in Fontan patients (+2.8% vs +6.3%; $p = 0.008$). 27 Fontan patients completed the enalapril study with a mean dosage of 0.3 ± 0.1mg/kg/day for a duration of 13.4 ± 3.2 weeks. Most parameters were unaffected by enalapril. However, treatment did lower systolic (118mmHg vs 114mmHg; $p = 0.004$) and diastolic blood pressure (65mmHg vs 61mmHg; $p = 0.03$) at rest. Furthermore, in reaction to tilt, only the percentage decrease in CI was higher after treatment (-6.7% vs -20.5%; $p = 0.03$), however the absolute CI during tilt was not lower (3.0L/min/m2 versus 2.8L/min/m2; $p = 0.15$).
Conclusion: Paediatric Fontan patients exhibit orthostatic tolerance with adequate maintenance of blood pressure and CI and sufficient vagal response. Enalapril treatment resulted in a lower blood pressure at rest but did not alter the orthostatic stress response.

Keywords: Fontan physiology, Orthostasis, Hemodynamics, Enalapril, ACE inhibition

Table. Cardiovascular parameters during supine rest and head-up tilt in Fontan patients vs controls and before enalapril treatment vs follow-up in Fontan patients

	Supine rest	HUT	% difference	Supine rest	HUT	% difference
	Fontan patients			*Healthy controls*		
HR (bpm)	69.2 (14.6)	88.0 [76.6-94.4] ǂǂ	+22.4 [13.9-44.0]	69.4 (10.3)	89.7 (13.5) ǂǂ	+30.5 [19.5-41.0]
Systolic BP (mmHg)	118.7 (9.2)	121.3 (9.5) ǂ	+2.3 (5.2)	108.3 (8.5) ***	114.8 (7.3) ǂǂ	+6.3 (6.2) ǂǂ
Diastolic BP (mmHg)	65.4 (6.6)	73.5 (7.7) ǂǂ	+12.9 (12.0)	62.1 (7.5)	71.1 (6.3) ǂǂ	+15.8 (14.3)
CI (L/min/m²)	3.4 (0.7)	3.0 (0.6) ǂǂ	-9.7 [-18.3- -4.6]	3.3 (0.6)	2.8 (0.6) ǂǂ	-18.0 [-24.6- -3.1]
Mean flow art. cerebri media (cm/s)	66.8 [51.4-75.1]	60.7 [45.3-70.3] ǂ	-6.4 [-14.0- -2.7]	81.0 (15.6) **	72.0 (17.6) ǂ	-11.5 [-20.2- -3.7]
Aortic stiffness						
Pulse wave velocity aorta (m/s)	5.6 (1.2)	6.8 (1.5) ǂǂ	+23.3 (20.0)	4.4 (0.6) **	5.2 (1.0) ǂǂ	+18.5 (12.2)
Cardiac vagal activity						
RSA (ms)	70.0 [27.4-112.0]	35.3 [12.1-59.6] ǂǂ	-44.0 [-62.7- -25.7]	84.5 [55.8-119.9]	47.7 [36.6-58.5] ǂǂ	-47.0 [-54.3- -31.8]
RMSSD (ms)	60.6 [20.8-107.2]	25.3 [9.0-35.0] ǂǂ	-55.5 [-69.6- -27.1]	62.5 [49.8-103.1]	30.0 [22.7-36.6] ǂǂ	-52.7 [-68.4- -44.3]
SDNN (ms)	79.9 [28.0-126.1]	40.0 [19.2-54.5] ǂǂ	-44.5 [-61.1 -22.0]	70.7 [55.1-97.6]	56.8 [44.8-70.6] ǂ	-18.4 [-35.4- -3.2] ǂǂ
	Pre-enalapril			*Follow-up*		
HR (bpm)	66.6 (14.4)	85.9 (13.9) ǂǂ	+25.7 [18.9-44.2]	68.1 (17.0)	86.3 (16.4) ǂǂ	+27.0 [17.9-38.9]
Systolic BP (mmHg)	118.6 (8.4)	122.0 [116.0-126.5] ǂ	+3.5 (5.1)	114.3 (7.7) **	117.0 [111.0-123.0] ǂ	+3.6 (7.4)
Diastolic BP (mmHg)	65.0 (6.1)	74.7 (7.9) ǂǂ	+17.2 [4.0-24.0]	61.0 [59.0-65.5] *	70.0 [64.5-76.0] ǂǂ	+11.1 [8.9-21.5]
CI (L/min/m²)	3.3 (0.6)	3.0 (0.5) ǂ	-6.7 [-18.2-2.4]	3.5 (0.6)	2.8 (0.7) ǂǂ	-20.5 [-27.6- -6.8] ǂ
Mean flow art. cerebri media (cm/s)	59.7 [50.8-69.0]	58.2 ǂ [40.5-69.2]	-5.1 [21.8- 1.2]	56.9 [48.8-67.6]	55.9 [47.2-60.5]	-2.4 [-18.0-3.8]
Aortic stiffness						
Pulse wave velocity aorta (m/s)	5.4 (0.9)	6.8 (0.9) ǂǂ	+27.7 (18.2)	5.1 (1.1)	6.3 (1.2) ǂǂ	+24.4 (11.5)
Cardiac vagal activity						
RSA (ms)	81.5 [42.2-111.1]	35.3 [18.3-56.8] ǂ	-44.0 [-65.5- -12.3]	66.7 [35.7-88.2]	32.6 [18.0-64.4] ǂ	-34.3 [-61.9- -8.6]
RMSSD (ms)	78.0 [40.8-110.6]	25.3 [16.5-33.1] ǂǂ	-57.6 [-76.0- -28.8]	68.8 [30.0-94.8]	23.2 [11.2-41.8] ǂǂ	-52.2 [-71.1- -27.4]
SDNN (ms)	86.9 [46.4-132.4]	40.0 [21.7-55.4] ǂǂ	-42.9 (28.1)	79.2 [43.0-100.7]	41.8 [27.0-56.4] ǂ	-30.8 (29.8)

Data expressed as mean (±SD), and median [Q1-Q3].
P-value: *<0.05, **<0.01, ***<0.001 for differences in supine rest between Fontan patients and healthy controls or pre-enalapril treatment and follow-up;
P-value: ǂ <0.05, ǂǂ <0.01, ǂǂǂ <0.001 for difference between supine and HUT parameters in each group of subjects (Fontan patients and controls or pre-enalapril treatment and follow-up;
P-value: ǂ <0.05, ǂǂ<0.01, ǂǂǂ <0.001 for difference in percentage change in parameters from supine to HUT between patients and controls or pre-enalapril treatment and follow-up.
BP= Blood pressure; CI= cardiac index; HUT= head-up tilt; RMSSD= root mean square of successive differences between normal sinus beats; RSA= respiratory sinus arrhythmia; SDNN= standard deviation of the inter-beat interval of normal sinus beats;

P-094

Growing pains in young patients with univentricular heart palliated with total cavo-pulmonary connection

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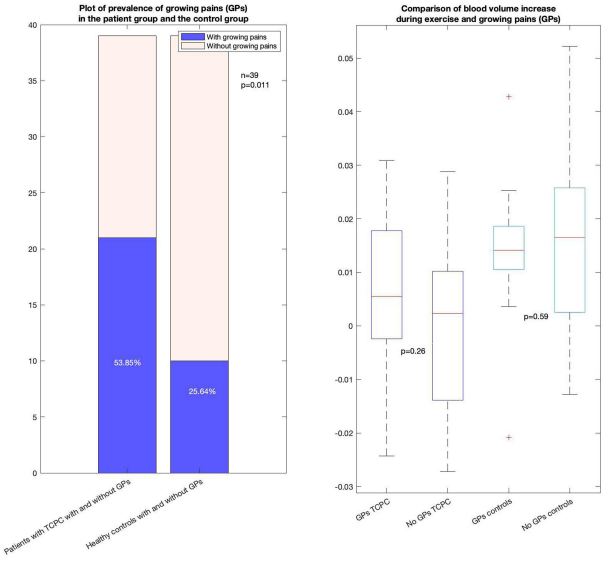
Background and Aim: Growing pains is a condition in children characterized by intermittent non-articular pains in the lower limbs, most commonly occurring in the evening or during night-time. Studies have shown that young patients with total cavo-pulmonary connection (TCPC) have lower muscle mass, impaired muscle strength in the lower limb muscles, and they also exhibit an altered muscle metabolism compared to healthy controls. Our primary aim was to determine prevalence of growing pains in children with TCPC compared to a cohort of age and sex matched healthy controls. The second aim was to investigate if muscle mass, bone density, isometric muscle strength, muscle endurance or muscle metabolism is associated with growing pains.
Methods: Thirty-nine patients with univentricular heart defects surgically palliated with TCPC were recruited. Exclusion criteria were age younger than 6 years and older than 19 years, cognitive impairment, or co-morbidity making participation in the muscle testing impossible. The patients were compared with 39 age and sex-matched healthy control subjects. A questionnaire including questions on leg pains, prior or present was used, and participants were asked to briefly describe their pains. Body composition was analysed with dual-energy X-ray absorptiometry, isometric muscle strength, muscle endurance capacity was assessed, and calf muscle metabolism during exercise measured with near-infrared spectroscopy (NIRS) in patients and controls.
Results: Growing pains was present in 54% of the TCPC group and in 26% in the control group, $p = 0.011$, (Fig 1). There was no difference between patients and controls regarding body composition or muscle function tests. Yet, there might be a tendency towards slower rate of increase in total hemoglobin (blood volume) analyzed with NIRS comparing the patients reporting growing pains and the patients without growing pains (Fig 1).
Conclusion: Growing pains are more common in young patients palliated with TCPC compared to healthy peers. A slower increase of blood volume for the patients with growing pains could indicate presence of obstruction due to increased intramuscular pressure. Larger studies are needed to further assess any differences.

Keywords: Growing pains, univentricular heart, Total cavo-pulmonary connection, TCPC, Fontan, children

Results_TCPC_GP



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P-095

Is pulmonary hypertension a risk factor for poor outcomes in bronchiolitis?

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Background and Aim: Bronchiolitis is the most common lower respiratory tract infection in children. Pulmonary hypertension (PH) has been reported as a crucial factor in the pathophysiology of severe bronchiolitis. The aim of this study was to assess PH and right ventricle (RV) function in patients with bronchiolitis and to analyze their correlation with clinical outcomes.
Methods: This prospective cohort study included infants less than 12 months of age hospitalized with bronchiolitis. The cohort was divided in mild and severe cases based on the need for positive pressure respiratory support (PPRS). PH was considered when one of the following: RV acceleration/ejection time ratio (AT/ET) ≤0.3, RV isovolumic relaxation time (IVRT)>40 msec, eccentricity index (EI)≥ 1.1. RV/left ventricle ejection time ratio (RV/LVET) was also analyzed. Statistics: bivariant analysis, significant differences $p<0.05$.
Results: One hundred eighty-one patients were included. Median age was 2 [1- 4] months. 73 (40%) required PPRS. Hospital length of stay (HLOS) was 9.5 [7-14] days for severe cases and 4 [3-5.7] for mild cases ($p = <0.001$). Patients requiring PPRS had increased NT-proBNP values compared to mild cases (1615 [763-3716] vs. 510 [243-1269] pg/ml; $p = 0.007$). Ninety-seven (53%) patients had at least one parameter of PH, 52 mild and 45 severe cases. Only 21 (11%) cases had more than one parameter, 13 mild and 8 severe cases. Sixty-six (36%) patients had an AT/ET ≤0.3, and 14 (7%) patients had IVRT >40 msec, neither of which were different between mild and severe groups ($p = 0.67$ and $p = 0.68$, respectively). There was no difference for diastolic or systolic EI ($p = 0.23$ and $P = 0.66$, respectively). RV/LVET was 1.06 for mild and 1.1 for severe cases ($p = 0.06$). Patients requiring PPRS had worse RV systolic function [TAPSE 11 vs 13mm, ($p<0.001$) and TDI S wave 9.3vs10cm/s, ($p = 0.05$)]. There was no correlation between AT/ET and TAPSE, NT proBNP or HLOS.
Conclusion: In our cohort patients with severe bronchiolitis had poorer RV systolic function but, interestingly, PH was not clearly evident. Although 36% of patients had an AT/ET≤0.3, this finding was not reinforced with other echocardiographic parameters of PH or associated to worse RV function or adverse clinical outcomes. Further investigation is required to better understand the pathophysiologic basis for this observed RV dysfunction.

Keywords: Bronchiolitis, Pulmonary hypertension, Right ventricle dysfunction

P-096

Outcome after surgery for membranous subaortic stenosis in children; long-term follow-up, recurrence and reintervention

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Background and Aim: Membranous subaortic stenosis (mSAS) is a congenital heart disease (CHD) that has a high rate of recurrence despite surgical treatment. The purpose of this study was to examine the background and outcome for operated mSAS-patients, identify predictors for recurrence and potentially propose indications for intervention.
Methods: A retrospective chart review from January 1994 until December 2019 was conducted for all surgically treated mSAS-patients (n = 38) in our institution up until 18 years of age. Patients with complex concomitant cardiac anomalies were excluded. The primary outcomes were recurrence, reintervention and mortality. Secondary outcomes were different predictors of recurrence and reintervention. Recurrence was defined as an increase of peak gradient (PG) > 50% during follow-up compared postoperative (post) PG and a minimum PG-value of 16 mmHg.
Results: Mean age at diagnosis, initial intervention and last follow-up was 3.4 ± 3.7(SD), 5.6 ± 3.6 and 14.8 ± 4.7 years, respectively. Mean follow-up time was 9.4 ± 5.6 years (range 1.1-18.4 years). 71% were males and 55% had other associated CHD. Death occurred in one patient (3%). Mean value of PG and mean gradient (MG) were at initial diagnosis, preoperative (pre), post and at last follow-up, 56/40, 83/48, 18/9 and 25/16 mmHg ($p<0.0001$ pre/post). 12 patients (56%) developed recurrence without receiving reintervention, while 7 (19%) developed recurrence with reintervention. Three patients were lost during follow-up. PrePG and postPG were linearly correlated, decreasing approximately by 80% pre-/postop. ($p<0.01$). Development of symptoms and the prePG correlated with a threshold-value of >90mmHg ($p<0.001$). The distance between the SAS-membrane and the aortic valve was inversely correlated to prePG ($p<0.01$). Age<5years at first intervention increased the likelihood of reintervention. High postPG/postMG as well as a presence of congenital aortic stenosis increased the likelihood of recurrence. A preop. value of PG/MG ≥ 60/40 mmHg should warrant a consideration of intervention.
Conclusion: MSAS-surgery is an effective therapy, but has a high rate of recurrence and reintervention. A clear correlation exists between high pre- and postPG. Low postPG seems vital for avoiding recurrence.

Keywords: Membranous subaortic stenosis, paediatric, surgery, recurrence, intervention, indications

P-097

Assessment of left ventricular dyssynchrony by speckle tracking echocardiography in children with duchenne muscular dystrophy

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Background and Aim: Prognosis of Duchenne muscular dystrophy (DMD) is related to cardiac dysfunction. Two dimensional-speckle tracking echocardiography (2D-STE) has recently

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emerged as a non-invasive functional biomarker for early detection of DMD-related cardiomyopathy. This study aimed to determine, in DMD children, the existence of a left ventricle (LV) dyssynchrony using 2D-STE analysis.

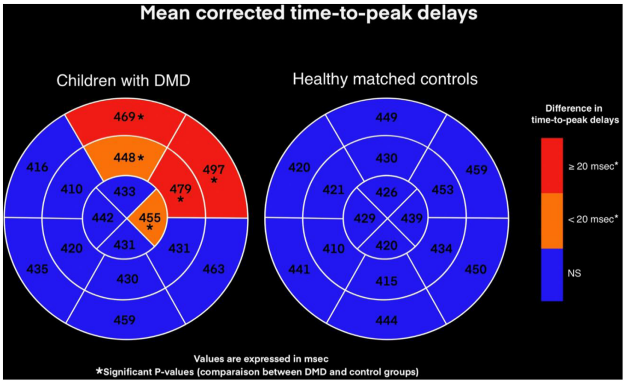
Methods: This prospective controlled study enrolled 25 boys with DMD (mean age 11.0 ± 3.5 years) with normal LV ejection fraction and 50 age-matched controls. Three measures were performed to assess LV mechanical dyssynchrony: the opposing-wall delays (longitudinal and radial analyses), the modified Yu index, and the time-to-peak delays of each segment. Feasibility and reproducibility of 2D-STE dyssynchrony were evaluated.

Results: All three mechanical dyssynchrony criteria were significantly higher in the DMD group than in healthy subjects: (1) opposing-wall delays in basal inferoseptal to basal anterolateral segments (61.4 ± 45.3 msec vs. 18.3 ± 50.4 msec, P<0.001, respectively) and in mid inferoseptal to mid anterolateral segments (58.6 ± 35.3 msec vs. 42.4 ± 36.4 msec, P<0.05, respectively), (2) modified Yu index (33.3 ± 10.1 msec vs. 28.5 ± 8.1 msec, P<0.05, respectively), and (3) most of time-to-peak values, especially in basal and mid anterolateral segments. Feasibility was excellent and reliability was moderate to excellent, with ICC values ranging from 0.49 to 0.97.

Conclusion: Detection of LV mechanical dyssynchrony using 2D-STE analysis is an easily and reproducible method in pediatrics. The existence of an early LV mechanical dyssynchrony visualized using 2D-STE analysis in children with DMD before the onset of cardiomyopathy represents a perspective for future pediatric drug trials in the DMD-related cardiomyopathy prevention.

Keywords: cardiac dyssynchrony, speckle tracking, DMD, heart failure, pediatrics

Comparison of LV longitudinal time-to-peak delays between DMD and control groups.



The two figures represent, for each group, the time-to-peak delay of each LV segment, corrected by heart rate (using Bazett's formula, expressed in msec), using the 16-segment bull's eye model of myocardial segmentation. The boxes in red represent the segments for which time-to-peak delays were significantly different (P<0.05) between the two groups with a magnitude of the difference ≥20 msec (basal anterolateral, mid anterolateral, and basal anterior segments). The boxes in orange represent the segments for which time-to-peak delays were significantly different (P<0.05) between the two groups with a magnitude of the difference <20 msec (mid anterior and apical lateral segments). The boxes in blue represent the segments for which time-to-peak delays were not significantly different between the two groups (NS, non-significant).

P-098

Preoperative assessment of pulmonary atresia with VSD from emerging nations: 3D reconstruction is a major help

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Background and Aim: Mécénat-Chirurgie-Cardiaque (MCC) is a French non-governmental organisation which has been bringing, since 1996, 3,800 children with congenital heart disease from 66 different emerging countries to France for surgical purposes. Only 4% of the 3,800 children returned to their country without surgery because of bad screening, but 23% of 132 Pulmonary Atresia with VSD (PA/VSD) for intractable pulmonary branch anatomy and MAPCA or fixed pulmonary hypertension, often impossible to know with local echocardiography.

To avoid this problem, we sought to improve the pre-surgical assessment of PA/VSD by a three-dimensional reconstruction (3DR) of a preoperative cardiac CT scan.

Methods: Since June 2021, all children diagnosed locally with PA/VSD were assessed with a cardiac CT scan with a special acquisition protocol, performed in their country of origin before bringing them to France for surgery. Dicom data were transferred to MCC and a 3DR was performed. The models obtained were analysed during the joint medico-surgical meeting of MCC which decides on the patient's arrival according to his operability (according to the size of the pulmonary arteries and MAPCAs).

Results: Eleven of the 12 cardiac CT scans from 8 different African countries could be reconstructed in 3D (one inadequate image protocol).

Unfortunately, only 3 children were considered for surgery and 8 were turned away due to pulmonary arteries and MAPCAs anatomy (see figure).

For 4 of these 8 rejected, however, the echo data appeared to be favourable, and for the remaining 4, echo data for the pulmonary branches was not available.

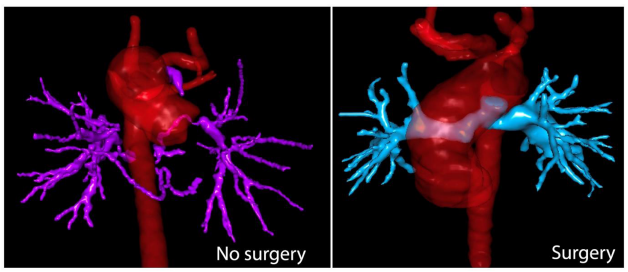
For 2 of the 3 accepted, we also did not have echo data for the pulmonary branches.

Without 3DR, we could have accepted for surgery 4 children with falsely favourable echo data and rejected 2 falsely unfavourable.

Conclusion: Local CT scan followed by 3D reconstruction was of great help in identifying patients with PA/VSD who could be brought to France and operated safely via MCC. The use of modern technology in emerging countries is becoming a reality.

Keywords: 3D reconstruction; PA/VSD; pulmonary arteries anatomy; MAPCAs anatomy; surgical planning; humanitarian medicine

No Surgery/Surgery



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P-099

Accuracy of current echocardiographic scores for biventricular repair risk prediction in borderline left ventricle: a critical review

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Background and Aim: Several echocardiographic formulas are used to predict biventricular repair (BVR) risk prediction for varying etiologies of complex congenital heart disease (CHD) with borderline left ventricle (LV) including critical aortic stenosis (AS), critical left ventricle outflow (LVOT) obstruction at any level, hypoplastic left heart complex/syndrome (HLHC/HLHS), and unbalnaced atrioventricular septal defects (uAVSD). Despite their widespread availability further augmented by online calculators, their effective use remains limited and accuracy debated. Tools are often erroneously employed interchangeably regardless of the etiology of the defect associated with LV hypoplasia. Given these limitations, the aim of the present review was to critically assess known echocardiographic BVR prediction models for borderline left ventricle in complex congenital CHD.
Methods: In September 2021 a systematic search in the National Library of Medicine for Medical Subject Headings and free text terms including echocardiography, CHD, and scores, was performed. The search was refined by adding keywords for critical AS, borderline LV, complex LVOT obstruction, HLHS/HLHC, uAVSD.
Results: (HLHS/HLHC), unbalanced atrio-ventricular septal defects (uAVSD). 15 studies were selected for the final analysis. We outlined what echocardiographic scores for different types of complex CHD with diminutive LV are available. Scores for CHD with LVOT obstruction including critical AS, HLHS/HLHC and aortic arch hypoplasia, have been validated and implemented by several studies. Scores for uAVSD with right ventricle (RV) dominance have also been established and implemented, the first being the atrioventricular valve index (AVVI). In addition to AVII, both LV/RV inflow angle and LV inflow index have all been validated for the prediction of BVR. We conclude with a discussion of limitations in the development and validation of each of these scores, including retrospective design during score development, heterogeneity in echocardiographic parameters evaluated, variability in the definition of outcomes, differences in adopted surgical and Interventional strategies, and institutional differences. Furthermore, scores developed in the past two decades may have little clinical relevance now.
Conclusion: In summary, we provide a review of echocardiographic scores for BVR in complex CHD with a diminutive LV that may serve as a guide for use in modern clinical practice.

Keywords: echocardiography, borderline left ventricle, calculators

Prediction models for BVR in Borderline LV

Prediction models for BVR in Borderline LV			
Score	**Author**	**Echo projection**	**Formula**
Rhodes	Rhodes et al., 1991	4Ch and PLa	14.0 (BSA) +0.943 (ROOTi) +4.78 (LAR) + 0.157 (MVAi) -12.03
Parsons	Parsons et al., 1991	4Ch and PLa	MV anulus, LVEDd, LV cross sectional area
CHSS1	Lofland et al., 2001	PLa	Survival benefit = 30.55 (inverse of age at study entry [d]−1) − 6.20 (aortic root z-score) − 12.14 (echocardiographic grade of EFE) − 23.33 (logarithm of Asc Ao [mm]) − 28.30 (presence of moderate or severe TR) − 0.70 (LV long-axis length z score) − 86.47
Discriminant Score	Colan et al., 2006	PLa	10.98 (BSA) - 0.56 (AoV z-score) - 5.89 (LAR) - 0.79 (presence of grade 2 or 3 EFE) - 6.78.
CHSS2	Hickey et al, 2007	4Ch and PLa	Multivariable regression equation using BSA, MV z score, indexed heart long axis, minimum LVOT diameter, mid aortic arch indexed
*2-V Score**	Mart and Eckhauser, 2014	4Ch, PLa and SA	[(MV annulus in 4Ch/AoV annulus)/ (LV length 4Ch/RV length 4Ch) + MPA diameter]/BSA
MV to TV ratio and AV ratio	Plymale et al., 2017	4Ch, PLa and SA, suprasternal view	Maximal systolic AoV, Maximal systolic PV, Maximal diastolic MV, Maximal diastolic TV, , Maximal systolic aortic arch , including Asc Ao *
Prediction models for BVR in uAVSD			
Index	**Author**	**Echo projection**	**Formula**
LV Volume	Van Son et al., 1997 (17)	estimated LV volume from the SA LV area and 4Ch	LV Volume = 0.85*SA area*long dimension
AV Valve Index (AVVI)	Cohen et al., 1996 (10)	30° left anterior oblique view diastole	area of the smaller AV valve component /common AV valve area
Modified AVVI (mAVVI)	Jegatheeswaran et al., 2015 (14)	30° left anterior oblique view, diastole	LAVV area/ common AV valve area
RV/LV inflow Angle	Cohen et al., 2013 (11)	4Ch, the angle of RV/LV inflow is measured from the crest of the IVS to each AVV hinge point. The angle is derived at the crest of the IVS.	Not Applicable
LV inflow index (LVII)	Szwast et al., 2011	4Ch, diastole	secondary colour inflow diameter/AVV annulus diameter.
LAVRI	Schleiger et al. 2021	left anterior oblique view at end-diastole and bisecting the common AV valve along a connecting line from muscular to infundibular IVS	Left AVV area after complete cleft closure/BSA (Du Bois formula)

Asc Ao = ascending aorta, AoV = aortic valve; AVV = atrioventricular valve, BSA = Body Surface Area; BVR = biventricular repair; DescAo = descending aorta; CA = carotid artery; -i = indexed by BSA; DT = Distal Transverse Arch; IA = innominate artery; EFE = Endocardial FibroElastosis; IVS = interventricular septum; LA = Left Atrium; LAR = LV long-axis to heart long-axis ratio; LCA = left carotid artery; LV = Left Ventricle; LVEDd = Left Ventricle End Diastolic Diameter; LVOT = Left Ventricle Output Tract; MPA = Main Pulmonary Artery; MV = Mitral Valve; MVA = Mitral Valve Area; RA = Right Atrium; RSA = right subclavian artery, PLa = parasternal long axis; PV = Pulmonary Valve; s, TR = Tricuspid Regurgitation; TV = Tricuspid Valve; 4 Ch = apical four chamber view; uAVSD = unbalanced atrio-ventricular septal defect.

P-100

Characterization of aortic flow patterns by newly high-frame rate blood speckle tracking echocardiography in healthy children

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Background and Aim: High frame rate blood speckle tracking (BST) is a newly echocardiographic technique allowing for direct visualization of blood flow patterns into cardiac chambers and vessels. In healthy subject's aortic flow should be laminar, but helical motions and vortices may be present.
AIM: to investigate flow patterns in the aorta by BST echocardiography in healthy children.
Methods: 60 healthy children (32 males) (mean age 9.19 ±4.92 years, median 9.47 years; interquartile range-IQR- 5.87-13.13 years; median body surface area 1.1 m2, IQR 0.83-1.48 m2) were prospectively recruited. BST imaging was obtained from a focused and zoomed view of the aortic root, and of the ascending aorta in

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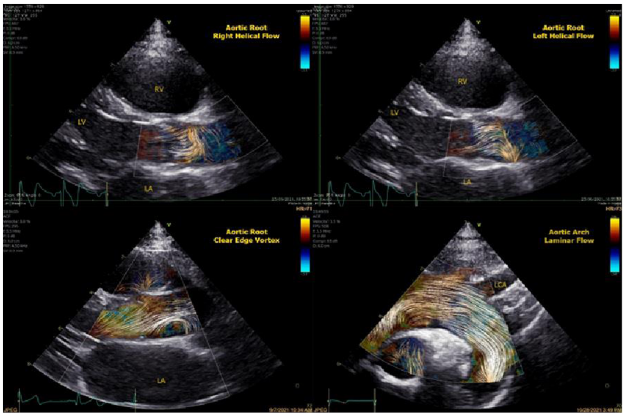
parasternal long-axis view. The flow characteristics were defined as follows: laminar (parallel to the major axis), helical (particles moving in a spiral), vortex (circular or swirling motion).
Results: Feasibility for BST analysis was 100%. A laminar flow was visible in all the cases. An anterograde systolic helical motion was visible in 55 out of 60 subjects (91.7%), whilst clear-edge vortices were identified in 19 out of 60 (31.7%). A single helical flow was visualized in 28 out of 55 cases (50.9%) and was directed toward the right coronary sinus in 20 (36.3%) and toward the left coronary sinus in 8 (14.5%). In 27 of 55 cases (49.1 %) a double helical flow motion (one to the right and one the left coronary sinus) was observed. Vortices were seen in early-diastole (12 out of 19,63.2%), or late-systole (7 out of 19,36.8%). Vortices were mainly visualized close to sino-tubular junction (STJ) (at a mean of 124 ± 63% of the total distance from aortic valve to STJ) and closer to left aortic wall (at a mean of 85.7 ± 24.7% of the total aortic diameter at the level of the STJ). Vortices had a mean area of 0.279 ± 0.134 cm2 (0.293 ± 0.103 cm2/m2).
Conclusion: We showed how in children aortic flow pattern characterization by newly BST echocardiography is highly feasible. Data on normal subjects may serve as baseline for the evaluation of flow pattern in children with congenital and acquired heart disease.

Keywords: Blood speckle tracking, aorta, children,

Blood speckle tracking sample of vortical, helical and laminar flow in aortic root and arch



LA = Left Atrium; LCA = Left Carotid Artery; LV = Left Ventricle; RV = Right Ventricle

P-102

Exercise tolerance and physical activity short after intensive treatment in patients with childhood cancer

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Background and Aim: Patients with childhood cancer are confronted with exercise intolerance (EI (VO_2peak<85% predicted)) after treatment, with a detrimental effect on quality of life and mortality. Knowledge on the limiting factor(s) for this EI and its relation with physical activity (PA) is essential in order to prescribe individually tailored rehabilitation and to stimulate physical and social reintegration.
Methods: 41 patients with childhood cancer (13 ± 3 years; 71% boys; BMI: 20 ± 4 kg/m²), diagnosed with leukemia/lymphoma (61%), solid tumor (32%) or brain tumor (7%) and recently finalized their oncology-related treatment (duration cancer treatment: 216 [168-270] days) were included in the study. Patients performed a maximal symptom-limited cardiopulmonary exercise test on a treadmill (4.8 km/h; +2% elevation/min). PA was recorded with a 3-axial accelerometer (Dynaport MoveMonitor, McRoberts, The Hague), that patients wore for 7 consecutive days. Active time (standing and walking), sedentary time and steps were withheld.
Results: Exercise tolerance (VO_2 peak: 29.7 ± 7.8 ml/min/kg (67 ± 16% predicted)) was markedly reduced in patients with childhood cancer compared to healthy peers. EI was present in 88% of patients. The majority of patients were peripherally limited (83%). A cardiac limitation was present in 71% of patients and was predominantly due to a reduced oxygen pulse (97%). Hyperventilation (32%) and a ventilatory limitation (12%) were less prevalent. (Figure 1) PA data of 13 patients were available (Active time: 178 ± 67 minutes/day; sedentary time: 515 ± 113 minutes/day; steps: 6411 [4458-6838]).
Conclusion: Exercise tolerance is markedly reduced in patients with childhood cancer short after intensive treatment and mainly caused by deconditioning of peripheral muscles and reduced oxygen pulse. Further research is necessary to study the link with physical activity.

Keywords: Exercise tolerance, childhood cancer, maximal symptom-limited cardiopulmonary exercise test, physical activity

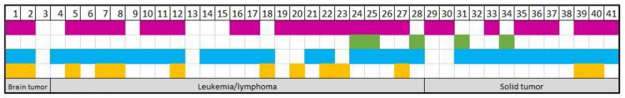

Figure 1: Individual limitation (s) of maximal symptom-limited cardiopulmonary exercise test based on cancer type. : Cardiac limitation; : Ventilatory limitation; : Peripheral limitation and : Hyperventilation.

P-103

Exploration of fetal cardiovascular mri for assessment of congenital heart disease at 20 to 28 weeks' gestation

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Background and Aim: Motion-corrected 'black-blood' fetal cardiac magnetic resonance imaging (MRI) provides excellent three-dimensional (3D) visualisation of suspected vascular anomalies in the third trimester but is severely limited by fetal motion and size at earlier gestational age (GA). We recently developed a novel technique using deep learning and landmark-based reorientation which allows correction of severe fetal motion and has shown promising results in healthy controls. In this study we explore

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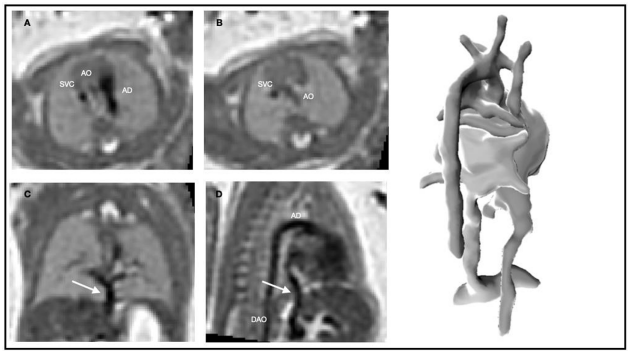
the diagnostic potential of this new fetal MRI tool in second trimester fetuses with suspected congenital heart disease (CHD).
Methods: Seven fetal MRI datasets (mean GA 24 weeks, GA 20-28 weeks) with known cardiovascular anomalies and three healthy control scans were identified retrospectively and reconstructed to 3D volumes using the new motion-correction technique. Two observers blinded to prenatal diagnosis and outcome subsequently assessed all ten fetal MRI datasets using standard segmental assessment of four vascular areas: systemic venous return, pulmonary arterial supply, pulmonary venous return, aortic and ductal arch anatomy (i.e. 40 areas per observer).
Results: Overall image quality was scored as "high" in 5/10, "adequate" in 4/10, "poor" in 1/10 and no reconstruction failed. In total, the two observers were unable to visualise structures in only 13/80 areas (16%), of which 2/80 for systemic venous return, 3/80 pulmonary arterial supply and 4/80 both pulmonary venous return and arch anatomy. In abnormal cases (example shown in Figure 1), pathognomonic vascular features were correctly identified in 6/7 (86%) by at least one observer, with only one vascular anomaly (aberrant right subclavian artery) not identified by either observer. In one healthy control aortic coarctation was suspected by one observer which was not present after birth.
Conclusion: This is the first report assessing feasibility and diagnostic potential of 3D motion-correction methods in the second trimester of pregnancy, demonstrating overall good performance for visualisation of vascular anomalies in these retrospective datasets. This has important implications for use at earlier gestation than is currently feasible using existing methods. Future work will focus on technical improvements and prospective evaluation of diagnostic accuracy.

Keywords: Congenital Heart Disease, Magnetic Resonance Imaging, Fetal



High quality motion-corrected 3D fetal CMR volume (resolution 0.7mm) of fetus (GA 23+5 weeks) with anomalous pulmonary venous drainage and ventricular and great artery disproportion: A) three vessel trachea view demonstrating great artery asymmetry B) transverse view demonstrating aortic arch hypoplasia C) coronal view of left and right lower pulmonary veins to descending vertical vein (arrow) D) sagittal view of vertical vein (arrow) to portal venous system E) 3D heart model of generated using semi-automatic segmentation (posterior view)

P-104

A rare case of a large coronary fistula with a giant aneurysm in a newborn

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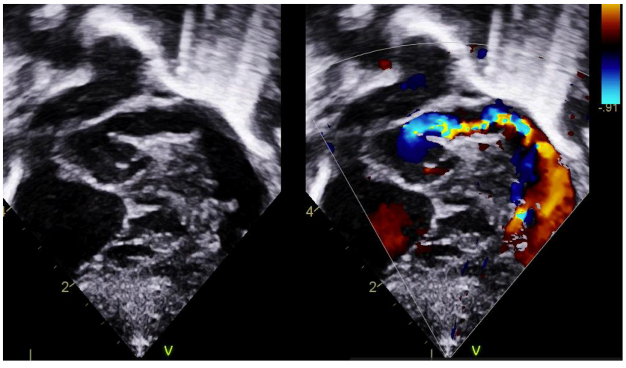
Background and Aim: Coronary fistulae (CAF) are persistent and abnormal connection between the coronary artery and one of the cardiac chambers or pulmonary arteries. less commonly they may be large and impose a significant hemodynamic burden or future risk for cardiovascular events
Methods: We are reporting a case of a left coronary fistula associated with a giant aneurysm which we managed. On day two of life a transthoracic echo was done showed, (large coronary artery fistula from left coronary cusp with large tunnel like structure, measures 10-11.5 mm with large opening to right ventricle apex below level of moderator band, opening is large measures 11.5 mm). A trial of device closure was done. But the child continued to be in heart failure due to severe leak around the device. A week later, underwent a cardiac surgery. Under cardiopulmonary bypass, the fistula was opened at the place with no coronaries. The device was removed, the opening to right ventricle was closed by Gore-Tex patch.
Results: The child remained asymptomatic and was followed up with serial echocardiography scans. The distal end of the fistula showed aneurysmal dilatation with a progressive course. At the age of 10 months the aneurysmal distal end was 30 mm x 27 mm.At the age of 11 months the child underwent a cardiac cath with insertion of an ASD device at the neck of the aneurysm. This stopped the aneurysm further expansion.
Conclusion: Although surgery was successful in eliminating the shunt and heart failure symptoms, the distal portion of the fistula, due to persistent exposure to aortic pressure and flow, continued to dilate. This increased the risk of complications especially thrombosis and rupture. Hence another intervention was indicated to close the neck of the aneurysm.

Keywords: Coronary fistula, Congenital heart anomaly, intervention

Coronary artery fistula subcostal view



A large vascular structure vessel is seen arising from the left coronary aortic sinus.

P-105

A rare case of combined congenital absent pericardium and diaphragmatic hernia mimicking cardiac tamponade

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Background and Aim: Congenital absent pericardium (CAP) combined with diaphragmatic hernia is a rare condition and a fatal complication could occur if it is not recognised. We report a case presented with suspected cardiac tamponade following repair of

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recurrent congenital diaphragmatic hernia. A lifesaving sternotomy was performed for drainage of mediastinal fluid collection and a CAP was identified intraoperatively
Methods: 4-year-old boy was brought to the local hospital emergency department with acute abdominal syndrome. He was known to have had a repair of congenital diaphragmatic hernia needing ECMO as a new born baby. He had underwent emergency bowel resection for an obstruction on the day of admission. He became hemodynamically unstable following abdominal operation and was transferred to the cardiac center with suspected cardiac tamponade evidenced by significant amount of fluid and air bubbles around the heart on echocardiogram. Fluid in association with air bubbles were considered to be in the mediastinum as there were no echocardiographic signs of tamponade and the cardiac shadow could only be visualised from the back.. Upon arrival to thecardiac surgical centre he was in a critical condition with systemic BP of 60 and CVP between 30 and 40 mmHg. An emergency xipho-sternotomy, followed by a full sternotomy was performed. There was a significant hemodynamic improvement upon opening of the chest with BP raising to 80 and CVP dropping to 15 mmHg. Intraoperatively, it was found that the retrosternal space was filled with the right lung. The heart was displaced to the left pleural cavity and the left pericardium was found to be completely absent. There was approximately 150mls of sero-sanguinous fluid in the left pleural cavity.
Results: The patient was stable following the procedure and was transferred back to the local hospital where he made an uneventful recovery and was discharged home soon after the surgery.
Conclusion: CAP combined with congenital diaphragmatic hernia can mimic tamponade and should be considered in the differential diagnosis of any case presenting with haemodynamic collapse, and mediastinal fluid collection

Keywords: Congenital absent pericardium, Diaphragmatic Hernia, Cardiac tamponade

P-106
Surveillance for ventricular dysfunction in duchenne muscular dystrophy

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Background and Aim: Recent studies have indicated the superiority of cardiac magnetic resonance imaging (CMR) comparative to trans-thoracic echocardiography (ECHO) in the detection of early signs of cardiac dysfunction in patients with Duchenne Muscular Dystrophy (DMD). Review of current practice in Royal Belfast Hospital for Sick Children (RBHSC) was prompted by local observation that few DMD patients had previously underwent CMR.
Our study aimed to review practice in RBHSC in the clinical assessment, investigation and management of cardiac dysfunction in DMD. We aspired to subsequently develop a regional protocol for cardiac management and imaging of this patient group.
Methods: Patients were identified using Northern Ireland (NI) DMD database. We performed a retrospective review of each patient's cardiac care using local data resources including Electronic Care Record (ECR) and NI Picture Archiving and Communication System (NIPACS). We assessed the parameters used to indicate dysfunction on clinical review and through ECHO surveillance and compared results to that seen on CMR. Comparison was then made of current imaging surveillance practice in RBHSC versus published guidelines in 2018 Journal of Pediatrics.
Results: 45 patients from the NI DMD database met inclusion criteria. 51% of paediatric DMD parents are on cardiac medication. 65% of these patients are on cardiac medication secondary to ECHO findings. 87% of ECHO findings prompting commencement on cardiac medication are based on subjective assessment. 67% of these patients had poor ECHO windows. 11.1% of DMD patients attending cardiology have underwent CMR, a further 4.4% are awaiting CMR. Findings correlated between ECHO and CMR in only 20% of cases.
Conclusion: Majority of ECHO findings prompting commencement on cardiac medication are based on subjective "eyeball" assessment. There was poor correlation between ECHO and CMR findings, however interpretation is limited by patient numbers. Increased utilisation of CMR in the assessment of this patient group is indicated.

Keywords: Duchenne Muscular Dystrophy, Cardiac MRI,

P-107
Cardiac involvement in pediatric inflammatory multisystem syndrome temporally associated with SARS-COV-2 (PIMS-TS; MIS-C)

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Background and Aim: Pediatric inflammatory multisystem syndrome temporally associated with SARS-CoV-2 (PIMS-TS) is a postimmunological reaction after SARS-CoV-2 infection. Various cardiac manifestations of PIMS-TS have been reported, namely pericardial effusion, ventricular arrhythmia, myocarditis, valvular regurgitation, and pericarditis. The aim of this study was to analyze clinical and laboratory features to distinguish any possible prediction for cardiac involvement in children with PIMS.
Methods: The PIMS patients under 18 years old treated in our center between July 2020 and December 2021 were included. Data of the patients were retrospectively obtained from their medical records.
Results: A total of 46 patients with PIMS were examined during the study period. The mean age of study group was 9.4 ± 4.6 years, 18/46 were female and 3 groups were formed according to their age ranges. Among them, seventeen patients (37%) had cardiac involvement with mean age was 8.7 years. Impaired cardiac functions were seen more in male patients (n: 10/17). Coronary artery dilatation seen in seven patients especially with mean age of 5.2 years (Age group 1,2,3; %36.4,%14.3,%0; $p = 0.033$; respectively) and especially related to high troponin T levels ($p = 0.006$). In our study group, cardiac involvement was shown more related to ProBNP and Troponin T ($p = 0.008$; $p = 0.003$). The cut-off values of proBNP and troponin T for predicting in cardiac dysfunction were 2759 pg/mL (95% confidence interval (CI), 0,83-1; sensitivity, 0.86; specificity, 0.93; AUC:0.92, $p < 0,001$).
Conclusion: Although there is a wide variability of symptoms, MIS-C is a rare, severe, less understood complication of COVID-19 that may cause multisystemic involvement in the patients. Clinicians should be aware of this condition in children with persistent fever and a family history of COVID-19. Cardiac involvement in children with PIMS may strongly be predicted by levels of Troponin T and ProBNP. Further more younger age and high Troponin T levels are the independent predictors for the coronary artery dilatation among children with PIMS.

Keywords: Pediatric inflammatory multisystem syndrome / PIMS, Cardiac Involvement, Echocardiography, Multisystem Inflammatory Syndrome in Children / MIS-C

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P-108

Infantile type of scimitar syndrome with persistent pulmonary hypertension, biventricular dysfunction and contralateral pulmonary vein stenosis

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Background and Aim: Scimitar syndrome (SS) is a rare condition consisting of anomalous pulmonary venous drainage from the right lung to the inferior vena cava (IVC). Various cardiac and respiratory anomalies have been reported in association with SS.
Methods: We present a case of a 4-month-old female infant, referred to our institution due to respiratory distress and desaturation to high 70s. She was diagnosed with SS with concomitant moderate secundum atrial septal defect (ASD), a large perimembranous ventricular septal defect (VSD), left superior vena cave to coronary sinus and abdominal arterial supply to the right lower lobe. Systolic biventricular function was impaired, correlated with chronic volume overload. Her condition was exacerbated by obstruction of the IVC between the entry sites of scimitar vein and hepatic veins, and entry of IVC to right atrium (RA) causing high pressure in the scimitar vein and low IVC, with much of the lower body venous return draining through collaterals to the azygos vein. At presentation, she was tachypnoeic and tachycardic with gallop. To assess reduced biventricular function, myocarditis screening was performed, and patient was tested positive for enterovirus and rhinovirus. She was intubated and ventilated and started on milrinone. She underwent cardiac catheterization and coil embolization of one aortopulmonary collateral. Computed tomography showed a right upper lobe tracheal bronchus and a narrowing of carina and right proximal bronchus intermedius and left main bronchus behind the right pulmonary artery. As a first surgical stage, fenestrated closure of ASD and VSD patch closure along with IVC obstruction relief was performed without intra-operative complications.
Results: Post operatively, she was treated for ongoing pulmonary hypertension (PH) with high flow oxygen and Sildenafil. Residual IVC-RA obstruction, mild left-sided pulmonary venous stenosis (PVS) and trachea-bronchomalacia complicated our case. Patient underwent transcatheter balloon dilatation of IVC to mitigate IVC-RA stenosis in 6 weeks after operation. Active management of near systemic PH, residual cardiac and respiratory lesions remain challenging.
Conclusion: Association between infantile-type SS and contralateral PVS is rarely reported and usually associated with dismal outcomes. Co-existing refractory pulmonary hypertension, persistent IVC-RA stenosis and poor ventricular function impose even higher morbidity and mortality rate.

Keywords: Infantile type, Scimitar syndrome, pulmonary hypertension

P-109

Dynamic change of an electrocardiogram in myocarditis following mRNA COVID-19 vaccination

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Background and Aim: Coronavirus disease (COVID-19) is a pandemic infectious disease caused by the SARS-CoV-2 virus. At the beginning of 2021, the Food and Drug Administration (FDA) of the United States issued an emergency use authorization (EUA) for the Pfizer-BioNTech COVID-19 vaccine to prevent COVID-19 infection. Cases of myocarditis and pericarditis after the vaccination in adolescents and young adults have been reported, especially more often after the second dose. There are very few case reports in Asia. We aim to present a case of confirmed myocarditis after the Pfizer-BioNTech COVID-19 vaccine in Thailand and the remarkable dynamic change in an electrocardiogram (ECG).
Methods: Case report. We reported the case and reviewed serial electrocardiogram and echocardiographic findings, including cardiac magnetic resonance imaging (MRI) in a case of confirmed myocarditis after the Pfizer-BioNTech COVID-19 vaccine.
Results: A previously healthy 13-year-old boy presented with chest pain and shortness of breath within 20 hours following the second dose of the Pfizer-BioNTech COVID-19 vaccine. Electrocardiogram (ECG) revealed diffuse ST-segment elevation with significant improvement within 3 hours. The peak level of high sensitivity cardiac troponin T was 1,546 ng/L. No alternative etiology of myocarditis was identified. Echocardiogram revealed mildly depressed left ventricular septal wall motion. Cardiac MRI showed abnormal native T1, T2 mapping, and extracellular volume (ECV) that were compatible with myocarditis. His symptoms were relieved with ibuprofen. He was discharged on the fifth day of admission. In a 1-week follow-up, the ECG showed an incomplete right bundle branch block.
Conclusion: This case illustrates the potential of myocarditis following mRNA COVID-19 vaccination with striking dynamic change and transition of the ECG. The possible mechanism for myocarditis after vaccination is molecular mimicry between viral spike protein and cardiac protein. More data and long-term follow-up are needed to understand the association between the COVID-19 vaccine and myopericarditis.

Keywords: myocarditis, mRNA covid-19 vaccination

P-110

Dietetic modification effect on left ventricular mass and function in patients with glycogen storage disease III: a longitudinal study

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Background and Aim: Cardiac involvement in Glycogen Storage Disease type III, (GSD III) typically manifests as left ventricular hypertrophy (LVH) which may progress to symptomatic hypertrophic cardiomyopathy and life-threatening cardiac arrhythmias. The aim of this study was to assess the LV systolic function in patients with GSD III using both conventional and novel echocardiographic indices of LV systolic and diastolic performance. It also sought to investigate if a high protein diet could be beneficial in those children.
Methods: Ten patients with GSD III, ages 4 months-10 years, were serially assessed by echocardiography for wall thickness, LV mass, ejection and shortening fractions, LV 2D longitudinal deformation, systolic tissue Doppler of the lateral and septal LV basal segments and LV diastolic function.
Results: At baseline the IVSd z score was 1.87 ± 1.44 (children with z score >2, n = 4), the LVPWd z score was 2.05 ± 1.15 (children with z score >2, n = 4). The LV mass z score at baseline was 0.70 ± 1.63 (children with z score >2, n = 2). The baseline 2D longitudinal strain was $-20.78 \pm 1.39\%$, (appeared to be reduced in two patients). Both children with reduced 2D longitudinal strain

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had z scores >2 for IVSd, LVPWd and LV mass, but normal LVEF and FS. At baseline the S'med z score was -1.19 ± 1.96 and S'lat was -0.44 ± 0.78, (all children had z score <2). The baseline E/A and E/E' z scores of those children were -0.26 ± 0.10 (all children had z score <2) and 1.20 ± 1.62 (children with z score >2, n = 2), respectively. The subsequent serial echocardiograms did not identify a significant effect on LV wall thickness, LV mass or LV systolic and diastolic performance after dietary interventions. Three children had low 2D strain at follow up as one more child had low 2D LV longitudinal strain in addition to the two children with abnormal strain at baseline.

Conclusion: The current study has confirmed that the incidence of LVH in paediatric patients with GSD III is common. However, conventional and novel echocardiographic indices of LV systolic and diastolic performance appeared to remain unaffected despite the dietary modifications during the follow-up period.

Keywords: GSD III: Glycogen Storage Disease type III

P-111

Role of restrictive pattern in cardiopulmonary exercise testing in adolescent with repaired tetralogy of fallot

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Background and Aim: Restrictive lung function (RLF) is considered a risk factor for reduced exercise performance in adults with repaired congenital heart disease. Patients with repaired tetralogy of Fallot (rTOF) present a higher RLF rate than the rest of the population. The aim of the study was to compare cardiopulmonary exercise test parameters between two subgroups (normal lung function and restrictive lung function) of adolescents with rTOF.

Methods: Multicentre retrospective review of patients with repaired tetralogy of Fallot who underwent a cardiopulmonary exercise testing (CPET) on a stationary cycle from 2017 to 2021. Inclusion criteria included a respiratory exchange ratio (RER) ≥ 1 and/or peak heart rate ≥ 85% of the predicted value. Exclusion criteria included an obstructive spirometry pattern. To classify lung function, forced vital capacity (FVC) and forced expiratory volume in 1st second of forceful expiration (FEV1) were used. Demographic variables, vital signs, and exercise test parameters were compared between normal lung function group and restrictive lung function group.

Results: Of the total 70 patients screened, a cohort of 59 patients met the inclusion criteria (51% female, mean age 14.3 ± 2.1 years); 37 with normal lung function (63%) and 22 with RLF (37%). Demographic variables and vital signs at rest and at the maximum level were similar in both groups. Scoliosis was more frequent in RLF patients (18% vs 5%, p = 0.02). In CPET parameters, exercise time (ET) and maximum workload (Watts) were also similar between groups. Individuals with RLF presented a statistically significant decrease in minute ventilation at peak exercise (VEmax) (48.1 ± 11.0 vs 58.1 ± 17.2, p = 0.01). Moreover, RLF patients had lower predicted peak oxygen uptake (%VO2), ventilatory anaerobic threshold (VAT), VE/VCO2 slope, breathing reserve (BR), and OUES100, although neither of these parameters had statistical significance.

Conclusion: Our study shows a tendency of rTOF adolescents with RLF to develop a decreased exercise capacity compared to those with normal lung function, although statistically significant differences were not reached. However, the contribution of RLF to decreased exercise capacity in adolescents may seem smaller than in adults. Further studies with a larger sample size are needed.

Keywords: Tetralogy of Fallot; restrictive lung function; cardiopulmonary exercise test.

Lung function	Normal (n=37)	Restrictive (n=22)	p value
Sex (female)	17 (46%)	12 (60%)	0.33
Age at CPET (years)	14.2 ± 2.2	14.4 ± 1.9	0.71
Weight	56.0 ± 17.2	49.2 ± 13.0	0.11
Height	158.7 ± 13.0	157.0 ± 9.0	0.74
BMI	21.75 ± 4.4	19.8 ± 3.8	0.06
Age at repair (months)	9.9 ± 4.6	9.7 ± 15.9	0.96
Scoliosis	5.3%	18%	0.02
Sternotomies (nº)	1.4 ± 0.7	1.75 ± 0.9	0.25
Exercise time (min:s)	8:43 ± 2:25	8:44 ± 3:47	0.48
Watts/kg	2.1 ± 0.5	2.1 ± 0.5	0.83
RER	1.12 ± 0.10	1.08 ± 0.05	0.06
VO2	28.8 ± 7.3	28.4 ± 6.4	0.86
%VO2	74.8 ± 13.7	69.3 ± 11.0	0.10
VAT	15.7 ± 4.0	14.6 ± 3.5	0.38
%VAT	40.6 ± 8.7	38.0 ± 8.8	0.28
VE/VCO2 slope	30.6 ± 6.4	28.6 ± 2.7	0.31
VE/VCO2 at VAT	28.1 ± 3.6	28.0 ± 3.0	0.98
%OXYPULSE	85.6 ± 16.3	88.5 ± 26.5	0.39
Breathing reserve	36.8 ± 14.9	32.5 ± 14.0	0.15
VEmax %	58.1 ± 17.2	48.1 ± 11.0	0.01
PETCO2	36.6 ± 3.8	36.6 ± 3.2	0.75
OUES100	1726 ± 499	1640 ± 390	0.40

CPET cardiopulmonary exercise test, *BMI* body mass index, *RER* respiratory exchange ratio, *VO2* peak volume of oxygen consumption, *VAT* ventilator anaerobic threshold, *VE* minute ventilation, *PETCO2* end-tidal carbon dioxide, *OUES100* oxygen uptake efficiency slope.

Table 1. Comparison of demographic variables and exercise test parameters in patients with normal and restrictive lung function

P-113

Benign idiopathic transient severe isolated right ventricular hypertrophy (BITSI RVH) in neonates - a case series

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Background and Aim: Isolated severe right ventricular hypertrophy (RVH) is a rare finding in neonates. Although a few etiological/causative factors have been described for secondary RVH in neonates, idiopathic RVH is even rarer. The aim of the study is to analyze the history, clinical findings, echocardiography findings, management and follow up of neonates with benign idiopathic transient severe isolated right ventricular hypertrophy, and propose criteria for diagnosis.

Methods: Of all the neonates (n = 3632) undergoing echocardiography study at a single tertiary pediatric cardiology referral centre, from February 2013 to December 2019, neonates with a diagnosis of isolated RVH (n = 34) were included in the case series. Neonates with structural heart defects or other causes known to cause RVH (pulmonary stenosis/atresia, persistent pulmonary hypertension of newborn, infants of diabetic mother, inborn errors of metabolism, etc.) were excluded. The antenatal history, birth details, symptomatology, clinical and echocardiography findings, and follow up details were collected and analyzed.

Results: Irrespective of the gestational age, mode of delivery, gender and clinical status, certain echocardiography findings were present in all the 34 neonates- severe right ventricular hypertrophy, inter-atrial communication shunting either right to left or bidirectional, mild to moderate tricuspid regurgitation, mild or no pulmonary regurgitation, no right or left ventricular outflow

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tract obstruction, normal pulmonary artery pressures for age, and normal left ventricular dimensions.

Conclusion: Benign idiopathic transient severe isolated right ventricular hypertrophy (BITSI RVH) is an entity seen in neonates who have isolated severe RVH without any discernible cause (idiopathic), which by its presence per se doesn't cause any major hemodynamic problem (benign), and resolves on its own without any treatment within 6-12 weeks of life (transient). It is a distinct cause of (transient) cyanosis in neonates. Not only is it underdiagnosed, but also underreported. Hence criteria for diagnosis of BITSI RVH have been proposed for ease of diagnosis, management and prognostication.

Keywords: cyanosis, echocardiography, neonates, severe right ventricular hypertrophy

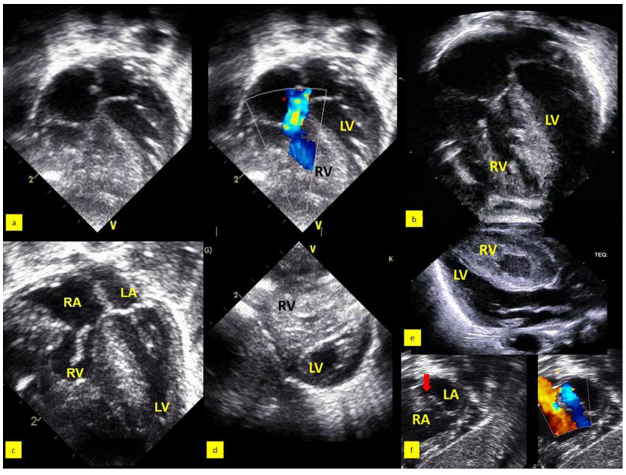

Figure 1 Representative images of few neonates, showing severe right ventricular hypertrophy. a,b,c: Apical 4 chamber views showing severe right ventricular hypertrophy, obliteration of RV apex (bipartite RV), tricuspid regurgitation,and normal left ventricular dimensions. d- parasternal short axis view, and e-parasternal long axis view: showing severe right ventricular hypertrophy and normal left ventricular dimensions. f- subcostal view showing right to left shunting across patent foramen ovale (red arrow).

P-114

Standardising 4D flow MRI derived kinetic energy in paediatric healthy volunteers - a validation study

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Background and Aim: Left ventricular (LV) and right ventricular (RV) blood flow kinetic energy (KE) is a potentially useful marker to further characterise cardiac performance and function. In adults KE is mainly normalised to end diastolic volume (EDV) but no gold standard, age-independent parameter has been systematically investigated and defined in the paediatric population. We aimed to investigate which indexed KE parameter has the least correlation with age in a healthy paediatric population.

Methods: 28 prospectively recruited healthy volunteers aged 6-16 years (Mean age = 11.41, ± 3.016 SD) underwent a prototype 4Dflow MRI scan at a 3 Tesla Trio (Siemens Healthcare Erlangen) as well as standard cine imaging. Research software MASS was used to calculate maximum systolic KE, minimum diastolic KE, peak E-wave KE and peak A-wave KE. Each KE value obtained was normalised to body surface area (BSA), end diastolic volume (EDV), stroke volume (SV), BSA indexed end diastolic volume (EDVi) and BSA indexed stroke volume (SVi). Pearson correlation coefficients and regression analysis were used to evaluate the correlations and associations of the indexed KE values with age.

Results: KE indexed to BSA had the least correlation with age (LV maximum systolic KE r = 0.346: LV minimum diastolic KE r = 0.351; LV peak E-wave KE r = 0.048; LV peak A-wave KE r = 0.034; RV maximum systolic KE r = 0.235: RV minimum diastolic KE r = 0.044; RV peak E-wave KE r = 0.335; RV peak A-wave KE r = -0.043) with the smallest regression coefficient in all KE values in both ventricles [see table; comparison example LV maximum systolic KE/BSA regression coefficient = 0.002 (95% CI 0-0.004), p value = 0.071]. Interestingly KE/EDV [LV maximum systolic KE/EDV regression coefficient = 0.243 (95% CI 0.039,0.446), p value = 0.021] and KE/SV [LV maximum systolic KE/SV regression coefficient = 0.17 (95% CI 0.039,0.302), p value = 0.013] showed a stronger association with age suggesting a bigger impact of age on these indexed parameters.

Conclusion: When assessing a paediatric population for age independent outcomes, kinetic energy should be indexed to BSA to minimise the bias of age in the comparison.

Keywords: 4D Flow MRI, kinetic energy, validation

Results table

		Left Ventricle					Right Ventricle				
		Indexed to BSA	Indexed to EDV	Indexed to SV	Indexed to EDVi	Indexed to SVi	Indexed to BSA	Indexed to EDV	Indexed to SV	Indexed to EDVi	Indexed to SVi
Max systolic KE	Regression coefficient	0.002	0.243	0.17	0.209	0.133	0.001	0.231	0.131	0.224	0.129
	p value	0.071	0.021	0.013	<0.01	<0.01	0.229	0.074	0.069	<0.01	<0.01
	Confidence interval	(0.000,0.004)	(0.039,0.446)	(0.039,0.302)	(0.150,0.268)	(0.096,0.170)	(-0.001,0.003)	(-0.024,0.486)	(-0.011,0.272)	(0.166,0.282)	(0.094,0.165)
	Pearson correlation	0.346	0.433	0.463	0.819	0.825	0.235	0.343	0.349	0.842	0.83
Min diastolic KE	Regression coefficient	0.021	2.065	1.167	1.846	1.072	0.001	0.11	0.083	0.727	0.383
	p value	0.067	0.043	0.054	<0.01	<0.01	0.825	0.853	0.802	0.046	0.056
	Confidence interval	(-0.002,0.044)	(0.073,4.057)	(-0.024,2.357)	(0.980,2.712)	(0.539,1.605)	(-0.011,0.014)	(-1.099,1.319)	(-0.593,0.759)	(0.015,1.438)	(-0.011,0.778)
	Pearson correlation	0.351	0.386	0.367	0.652	0.63	0.044	0.037	0.05	0.381	0.365
Peak E-wave KE	Regression coefficient	0.003	0.018	0.015	0.121	0.065	0.003	0.409	0.232	0.365	0.218
	p value	0.081	0.796	0.703	<0.01	<0.01	0.081	0.026	0.028	<0.01	<0.01
	Confidence interval	(0.000,0.007)	(-0.122,0.158)	(-0.064,0.094)	(0.062,0.180)	(0.030,0.100)	(0.000,0.007)	(0.053,0.766)	(0.027,0.437)	(0.251,0.479)	(0.150,0.286)
	Pearson correlation	0.048	0.051	0.075	0.636	0.599	0.335	0.42	0.416	0.791	0.791
Peak A-wave KE	Regression coefficient	0	0.03	0.031	0.117	0.075	-0.001	0.03	0.042	0.248	0.168
	p value	0.865	0.832	0.705	0.083	0.062	0.827	0.899	0.781	<0.01	<0.01
	Confidence interval	(-0.003,0.004)	(-0.258,0.318)	(-0.137,0.2)	(-0.016,0.25)	(-0.004,0.155)	(-0.006,0.005)	(-0.447,0.507)	(-0.268,0.353)	(0.073,0.424)	(0.060,0.277)
	Pearson correlation	0.034	0.042	0.075	0.333	0.358	-0.043	0.025	0.055	0.495	0.53

KE = kinetic energy; BSA = body surface area; EDV = end diastolic volume; SV = stroke volume; EDVi = end diastolic volume indexed to BSA; SVi = stroke volume indexed to BSA

P-115

Influence of subendocardial perfusion on ventricular function after norwood-SANO palliation

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Background and Aim: The hypoplastic left ventricle (LV) is present in different cardiac single ventricle anomalies. Palliation of

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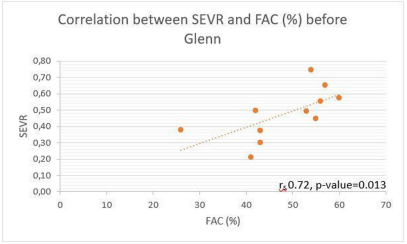
hypoplastic LV requires a challenging three-staged surgical pathway which begins with Norwood intervention and continues with two operations (Glenn and Fontan) to create the final univentricular Fontan physiology. After Norwood intervention, the single ventricle is exposed to cyanosis and volume overload. The coronary perfusion may be impaired, causing silent myocardial ischemia and ventricular dysfunction. Coronary anomalies and coronary artery fistula are not unusual in the HLHS. The type of pulmonary blood supply used in Norwood intervention (modified Taussig-Bing shunt, mBTS, or right ventricle to pulmonary artery conduit, RVPAC) also influences the adequacy of myocardial perfusion. However, even in case of RVPAC the coronary perfusion is impaired. We hypothesized that abnormal myocardial oxygen supply–demand balance after Norwood-Sano could influence the systemic ventricular function.
Methods: We retrospectively evaluated all patients diagnosed with hypoplastic LV who survived to Norwood-Sano operation and reached stage II procedures at IRCCS Policlinico San Donato. We used subendocardial viability ratio (SEVR) as indirect index of myocardial oxygen supply-demand balance, calculating it during cardiac catheterization pre-stage II surgery. We used fractional area change (FAC) as index of right ventricular function, and we assessed FAC before and after Glenn intervention. We investigated the correlation between SEVR and FAC.
Results: From March 2019 to December 2021, we measured SEVR on 11 patients before second stage operation. Eight patients (73%) had hypoplastic left heart syndrome, 1 patient (9%) hypoplastic left heart complex, 1 patient (9%) ventricular septal defect with aortic arch interruption, 1 patient (9%) double outlet right ventricle with aortic atresia. The median SEVR was 0.49 (IQR: 0.21-0.75). Median FAC before Glenn was 53 % (IQR: 26-60%), after Glenn 50% (IQR: 36-57 %). Lower values of SEVR were associated with lower values of FAC both before (rs 0.72, p-value = 0.013) and after stage II surgery (rs 0.66, p-value = 0.028).
Conclusion: In patients with hypoplastic LV treated with Norwood-Sano procedure, the myocardial oxygen supply-demand imbalance influences the single ventricular function. Future studies should investigate the effectiveness of surgical and medical interventions in improving the myocardial perfusion, and therefore the ventricular function.

Keywords: Hypoplastic left ventricle, Norwood-Sano operation, coronary perfusion, SEVR

Correlation between Subendocardial Viability Ratio (SEVR) and Fractional Area Change (FAC) before and after Glenn intervention.



P-116
Cardiac manifestations of multisystem inflammatory syndrome in children

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[1]Department of Pediatric Cardiology, Ankara University, Ankara, Turkey; [2]Department of Intensive Care Unit, Ankara University, Ankara, Turkey; [3]Department of Radiology, Ankara University, Ankara, Turkey; [4]Department of Infectious Disease, Ankara University, Ankara, Turkey

Background and Aim: We aimed to evaluate cardiac manifestations of the multisystem inflammatory syndrome in children(MIS-C) and the changes in cardiac function during one year of follow-up.
Methods: All children diagnosed as MIS-C with cardiac involvement were enrolled in this prospective study. The diagnosis and severity of the disease of MIS-C was made according to the Centers for Disease Control and World Health Organization guidelines. Clinical findings, laboratory parameters including cardiac markers, electrocardiographic and echocardiographic findings at the time of diagnosis and during follow-up were evaluated. Cardiac magnetic resonance imaging (MRI) was performed on all children with echocardiographic abnormality.
Results: Between April 1st 2020 and December 1st 2021,71 children were diagnosed with MIS-C and 44 of these patients had cardiac involvement (25 male and 19 female). 24 patients were followed up in the intensive care unit and all of these patients had myocardial involvement. All the patients had elevated NT-proBNP levels (median:5893pg/ml) whereas troponin-T levels were above upper limit in 13 patients. A significant positive correlation was found between troponin-T and NT-proBNP ($p<0.01$). The NT-proBNP levels were also positively correlated with the severity of MIS-C ($p<0.05$). On admission 22 patients had tachycardia and atrioventricular conduction disturbances and supraventricular tachycardia developed in 5 of these patients during follow-up. Bradycardia was observed in 18(40%) patients during hospitalization (4 of these occurred after tachycardia). Although 26 patients had an echocardiographic abnormality, only twelve patients had systolic dysfunction (9 with mild and 3 with moderate) and two patients had diastolic dysfunction. NT-proBNP and troponin-T were negatively correlated with ejection fraction ve fractional shortening (respectively, $p = 0.003$, $p = 0.013$). Cardiac MRI was normal in all patients except 3 patients who had myocardial late gadolinium enhancement of left ventricle. Pericardial effusion was observed in 14 patients. The echocardiographic abnormalities disappeared in 42 patients during follow-up, one patient died on the second day of hospitalization and and 1 patient has ongoing LV systolic dysfunction.
Conclusion: Bradycardia and myocardial involvement is common during MIS-C. Although myocardial dysfunction can be observed during acute disease, commonly the disease does not cause permanent damage during one year of follow-up.

Keywords: MIS-C, cardiac MRI, bradycardia

P-117
Coronary artery and immunological status in PostCOVID-19 children in sarajevo

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Background and Aim: The Coronavirus disease 2019 /COVID-19/ exerts an unprecedented global impact on public health and health

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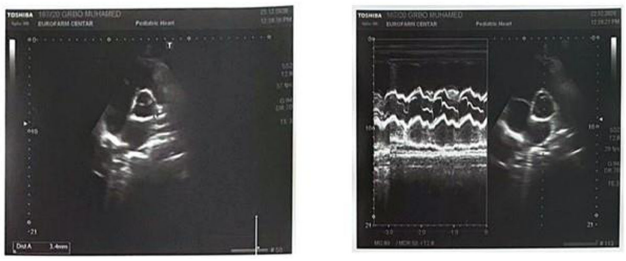
care delivery. The aim of this study was to evaluate the knowledge on SARS-CoV-2, epidemiology, clinical presentation including cardiovascular and immunological status in postCovid children.
Methods: A group of 70 children /previously healthy or with no pre-existing heart disease/ from Sarajevo with positive postcovid history, formed this study. Patients were evaluated at the Polyclinic Eurofarm in Sarajevo, from October 2020 till April 2021. Following history and epidemiological data, a detailed cardiovascular examination has been performed including oxygen saturation, pulse, blood pressure, electrocardiogram /ECG/, values of polymerase chain reaction (PCR), serological tests for corona, laboratory blood tests and echocardiography.
Results: The group consisted of 70 children /40 boys/: infants: 10, 1-5 years: 20; 6-10:12; 11-15:21; 16-18 years: 7; forming five groups. Symptoms differ depending on age group, younger children had no or mild symptoms in comparison to the older group of children. The values of immunoglobulin G were significantly higher in the older group of children with ($p < 0.05$; $p = 0.043$) indicating that the immune system with age is more responsive to the virus. PCR test was negative in 9/70 children. The majority of children /64.3 %/ were asymptomatic. Two boys aged 14 years, had palpitation on exertion, shortness of breath, ECG changes, lower oxygen saturation /91% and 94%/, elevated creatinine phosphokinase miofibrilae /CPKMB/: 38 and 45, in one patient the diameter of left coronary artery /LCA/ was enlarged up do 3.8mm, no aneurysm, no skin changes, with normal ejection fraction of left ventricle. They were on short period /10-15days/ of treatment with nonsteroids including low doses of Aspirin, vitamins /C and D/, rest and no sport activities. After treatment and a regime of no activities, they were fully recovered, free of symptoms, with normal oxygen saturation, normal values of CPKMB, diameter of LCA was within a normal range according to age and body weight of the patient.
Conclusion: Practitioners should consider the possibility of COVID-19 in children with atypical symptomatology and positive or suspicious epidemiological survey, paying special attention to coronary and immunological status.

Keywords: Post-COVID-19, children, coronary artery and immunological status

Figure 1. A: 2D echocardiogram, short axis view, level of aorta, showing dilated diameter of left coronary artery /LCA/, and **B:** M and B mod view of the same patient showing normal opening of three leaflet coronary artery.



P-118

Resolution of severe haemolytic anemia after cardiac surgery in a child with complex congenital heart disease and trisomy 21

Elio Caruso[1], Silvia Farruggio[1], Davide Calvaruso[1], Hadeed Khaled[2], Salvatore Agati[1], Philippe Acar[2]
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Background and Aim: We described a 23-months-old female infant, affected from trisomy 21 syndrome and a complex congenital heart disease (CHD) characterized by mitral valve cleft (MVC) with severe regurgitation, huge posterior ventricular septal defect (VSD) and tricuspid valve straddling (TVS) running through the left outflow, who was referred us for severe haemolytic anemia (HA) and low growth retardation.
No extracardiac anomalies were hypothyroidism, double left renal district with a vesico-ureteral reflux of II grade.
We excluded all differential diagnosis of HA and we suppose the ethiopathogenesis of HA due to MVC, TVS and pulmonary banding.
Methods: Echocardiogram at birth showed situs solitus, levocardia, normal systemic and pulmonary venous returns, dilated left atrium, AV and VA concordance, severe mitral regurgitation due to the cleft, TVS with almost three chordae tendinae going from septal and posterior tricuspid leaflet through the VSD and inhabiting the left outflow and inlet ventricle tracts. One chorda tendinae was attached to the anterior mitral leaflet, two to the anterior papillary muscle and one to the lateral left ventricular wall. Dilated left ventricle, normal ejection fraction, huge posterior VSD with anterior extension, dilated pulmonary trunk (PT).
At 8 months of age and BW 5 kg, due to the complexity of CHD and low BW, a palliative cardiac surgery attempted was PT banding and mitral valve commissuroplasty.
One year later from the cardiac surgery the patient was hospitalized for fever, paleness, vomit and asthenia; blood tests in emergency room showed a severe HA (Hb 4,6 g/dl, BW: 7,6 Kg) and splenomegaly. Patient needed blood transfusions every 5 days. Echocardiography confirmed previous anatomy and showed a setting of TVS causing anterior mitral leaflet prolapse and worsening severely the regurgitation due to the cleft, left chamber dilatation, maximum gradient on PT banding of 60 mmHg.
Results: After have excluded all other causes of HA, the patient underwent to definitive surgical treatment, and surgical view confirmed echocardiographical diagnosis.
Conclusion: We suspect turbulence and mechanical damage related to MVC, pulmonary banding, TVS caused HA which resolved after definitive surgery. To our knowledge this is a very rare case for both anatomy and anemia resolution.

Keywords: Hemolytic anemia, mitral valve cleft, straddling tricuspid valve, ventricular septal defect, pulmonary banding.

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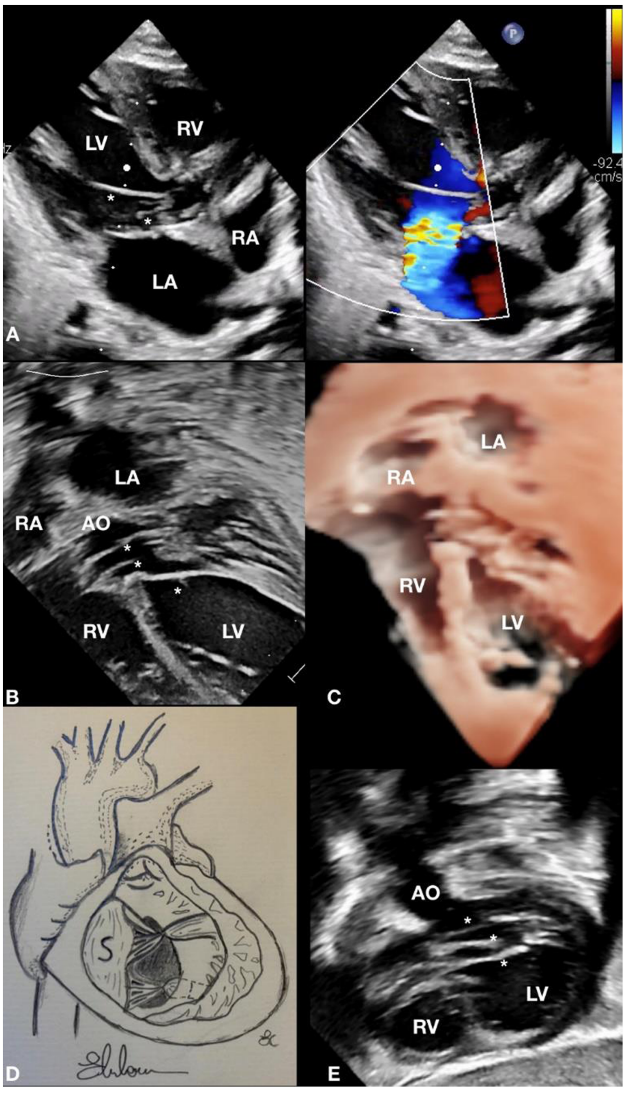
Straddling of tricuspid valve through the ventricular septal defect



A. Parasternal off-axis view shows severe mitral valve regurgitation and attachment of one chorda tendinae to the anterior mitral leaflet and to the anterior papillary muscle. B. Five chamber view shows straddling of tricuspid valve through the subaortic ventricular septal defect with the attachment of chordae tendinae to the anterior mitral leaflet and to the anterior papillary muscle. C. Three dimensional echocardiography four chamber view shows the posterior extension of ventricular septal defect and straddling of tricuspid valve. D. Courtesy of Elio Caruso. Manual drawing of surgical view of the straddling of tricuspid valve. E. Subcostal ventricular short axis view shows the attachment of chordae tendinae from posterior and septal tricuspid valve to the anterior papillary muscle and to the lateral left ventricular wall.

P-119/ Moderated Poster

Myocardial strain response to a preload challenge in the fontan circulation

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Background and Aim: Limitation of cardiac output reserve in the Fontan circulation is well documented, but the mechanisms are not fully understood. We aimed to explore the role of myocardial function by investigating echocardiographic strain measurements in response to an acute preload increase in adolescent patients with Fontan circulation.

Methods: We included 20 patients (median age 16.6 years, range 16.1-17.7 years, 6 females) during pre-transition diagnostic work-up. Echocardiographic imaging for longitudinal strain analysis was performed during heart catherization, both at baseline (rest), during a rapid 5 ml/kg body weight saline infusion and repeatedly until 6 minutes post infusion. We measured myocardial peak global strain (GLS) in a longitudinal 4 chamber view before saline infusion (rest), at 1.00-2.00 minutes after saline infusion and at 6.00 minutes (steady state). Measurements of central venous (CVP) and ventricular end diastolic (VEDP) pressures were obtained simultaneously. A CVP of 18 mmHg or higher as well as a VEDP of 15 mmHg or higher at any stage were considered elevated.

Results: Mean GLS was -17.4% (±4.5%) at baseline, -18.0% (±3.8%) after saline infusion, and -17.2% (±4.1%) at steady state. In more than half of the patients, there was no change or even a decline in GLS after saline infusion. In six patients, the lowest GLS after saline infusion was equal to or more than 20% reduced compared to baseline. Ventricular end diastolic pressure was elevated in three patients and CVP was elevated in two patients. Univariate analysis demonstrated that the degree of GLS decline correlated significantly with VEDP after volume load ($P = 0.002$). There was no difference between morphologic right (RV) and morphologic left (LV) ventricles. Decline in GLS did not correlate with baseline CVP.

Conclusion: Myocardial deformation imaging during volume load demonstrates lack of improvement or even worsening of systolic function by GLS, and the degree correlates with VEDP. Volume load thereby reveals the incompetence of preload recruitable stroke work in the Fontan circulation.

Keywords: Fontan, univentricular heart defect, echocardiography, strain, catheterization, invasive hemodynamics

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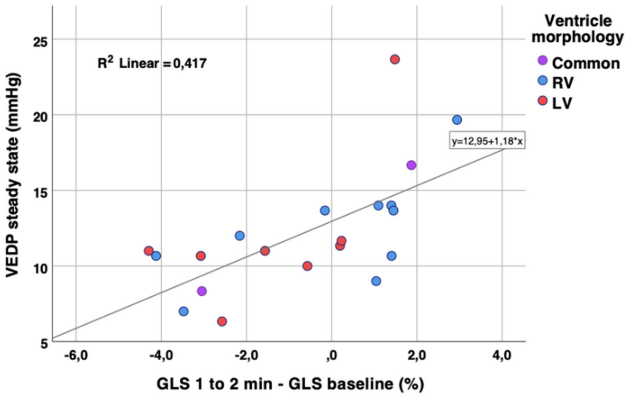

Figure 1 Correlation of change in global longitudinal strain (GLS) by echocardiography during volume load (x-axis) and invasive measurement of ventricular end diastolic pressure at steady state (y-axis). RV, right ventricle. LV, left ventricle.

P-120

Quantitative deformation echocardiography of ventricular function in pulmonary hypertension due to uncorrected congenital shunts

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Background and Aim: Prognosis and survival are much better in patients with pulmonary arterial hypertension (PAH) due to uncorrected congenital heart disease (CHD) with left to right shunts including Eisenmenger syndrome compared to other forms of pulmonary hypertension despite the presence of significant pressure and volume overload. The aim of this study was to assess morphology and function of right atrium (RA), right ventricle (RV) and left ventricle (LV) in such patients using 2D standard and speckle tracking echocardiography and examine the clinical association of various parameters.
Methods: All enrolled patients underwent clinical assessment, 6-minute walking test, cardiopulmonary exercise test, NT-proBNP measurement and 2D standard and speckle tracking echocardiography for RV and LV function.
Results: A total of 24 patients, aged 37 ± 11 (9-56) years, 14 males, were enrolled in the study. Diagnoses included ventricular septal defect (12 patients), patent ductus arteriosus (1 patient), aortopulmonary window (3 patients) and complex CHD with PAH (8 patients). Patients were in NYHA II-III and all were on pulmonary vasodilator therapy with endothelin antagonists. A total of 7 patients were additionally treated with PDE-5 inhibitors, while 1 patient received triple therapy with the addition of selexipag. RA area was marginal at 17 ± 8cm2, while RV was dilated with RV mid diameter 40 ± 2mm and RV basal diameter 45 ± 3mm. TAPSE, RV pulsed Doppler S wave and RV free wall 2D strain were relatively preserved at 19 ± 2mm, 13 ± 1cm/sec and -21 ± 3% (Table).
Conclusion: Patients with PAH due to congenital shunts including Eisenmenger syndrome exhibit relatively preserved RV and LV function compared to other forms of PAH despite the long-standing severe pressure and volume overload and chamber dilation. These findings suggest a favourable RV remodelling that is instrumental in the better prognosis and survival in this entity, while deformation mechanics of the ventricles may be used as a serial follow up tool in these patients.

Keywords: Pulmonary Hypertension, Uncorrected Congenital Shunts, Quantitative Deformation Echocardiography

Table. Echocardiography Data

	RA area (cm²)	RV mid diameter (mm)	RV basal diameter (mm)	TAPSE (mm)	PW S wave (cm/sec)	RV free wall 2D strain (%)
Patient population	17±8	40±2	45±3	19±2	13±1	-21±3
Abnormal threshold	>18	>35	>41	<17	<9.5	>-20

Comparison of our population data and the abnormal thresholds of various parameters derived from the American Society of Echocardiography Guidelines.

P-121

Pulmonary valve replacement in repaired congenital heart disease improves ventricular function by quantitative deformation echocardiography

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Background and Aim: Pulmonary insufficiency and stenosis after congenital heart disease (CHD) repair leads to right ventricular (RV) dilation and dysfunction necessitating eventual surgical or transcatheter pulmonary valve replacement (PVR) in the majority of patients. The timing of PVR is still disputed and mainly driven by the RV size and function on cardiac magnetic resonance (CMR) imaging. This study examines morphology and function of right atrium (RA), RV and left ventricle (LV) in such patients using 2D standard and speckle tracking echocardiography before and serially after PVR, attempting to identify clinically significant parameters.
Methods: All enrolled patients underwent clinical assessment, cardiopulmonary exercise test (CPET), CMR, NT-proBNP measurement and 2D standard and speckle tracking echocardiography for RV and LV function before, immediately after and 4 months after PVR.
Results: A total of 37 patients, aged 26 ± 12 (12-65) years, 24 males, were enrolled in the study. Diagnoses were Tetralogy of Fallot in 24 patients and operated complex CHD with prosthetic valves or conduits in the pulmonary position, such as Ross operation, in the remaining patients. Patients were in NYHA II-III and fulfilled the current criteria for PVR. PVR was surgical in 21 and transcatheter in 16 patients. 2D standard and speckle tracking echocardiography before and 4 months after PVR showed marginal RA area decrease from 17 ± 4 to 15 ± 4cm2, TAPSE increase from 13 ± 4 to 16 ± 5mm, RV pulsed Doppler S wave increase from 7 ± 1 to 9 ± 2 cm/sec and RV free wall 2D strain increase from -16 ± 6 to -21 ± 7% (Table).
Conclusion: Pulmonary insufficiency and stenosis after CHD repair adversely affects RV and LV function and PVR leads to improvement and ventricular remodelling, at least over midterm follow up. Serial measurements of RV and LV deformational parameters may

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be useful in the follow up of these patients and help guide towards the optimal timing of PVR in this complex patient population.

Keywords: Repaired Congenital Heart Disease, Pul monary Valve Replacement, Quantitative Deformation Echocardiography

Table. Echocardiography Data

	RA area (cm^2)		TAPSE (mm)		PW S wave (cm/sec)		RV free wall 2D strain (%)	
	Pre PVR	Post PVR	Pre PVR	Post PVR	Pre PVR	Post PVR	Pre PVR	Post PVR
Patient population	17±4	15±4	13±4	16±5	7±1	9±2	-16± 6	-21±7
Abnormal threshold	>18		<17		<9.5		>-20	

Caption. Comparison of our population echocardiographic data before and after PVR as well as the abnormal threshold of various parameters derived from the American Society of Echocardiography Guidelines.

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Management and outcomes of coronary fistulae in children and young adults

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Background and Aim: The objective of this study was to assess anatomical characteristics, management and oucome of coronary artery fistulaes (CAF) diagnosed at childhood age.
Methods: Retrospective observational single-centre study included all pediatric patients diagnosed with CAF from 2011 to 2014. Demographics, clinical, echocardiographic data, therapeutic management and outcomes were assessed.
Results: Eighty five patients were included in the study, 51 males and 34 females, diagnosed at a mean age of 6.5 years. Only one coronary artery was involved in 93% of the cases and 91% connected coronary artery and right heart (50% right ventricle, 35% main pulmonary artery and 6% right atrium. CAF was congenital in 89% of cases, acquired after surgery in 11%. CAF was isolated in 58% or associated with congenital heart disease in 42% of the cases. Coronary artery was enlarged in 27% and increased pulmonary flow was present in 8% of the cases. Symptoms and/or heart murmur were present in respectively 3 cases (4%) and 51% of the cases. Diagnosis was made by echocardiographic and Doppler evaluation. CT scan was performed to reach diagnosis in 11% of patients and coronary angiography in 6%. No patient died. Only 2 patients (2.3%) underwent closure of CAF (1 interventional procedure and 1 surgery).
Conclusion: Coronary artery fistulae natural history is uncomplicated and uneventful in the vast majority of cases.

Keywords: coronary artery, fistulae, natural history, prognosis

P-123

Experience with restrictive cardiomyopathy in childhood age

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Background and Aim: The objective of this study was to assess the clinical outcomes of children diagnosed with restrictive cardiomyopathy (RCM).
Methods: Retrospective data analysis of patients diagnosed with RCM at age less than 18y. Age at diagnosis, clinical oucome, therapeutics and survival were collected. Prognosis factors were assessed.
Results: Twenty four patients (15 males = 62.5%) were diagnosed with RCM, at the age of 5.5 ± 4.8y (median 4.2y, range 1week to 14.8y). All patients had normal LV and RV systolic function. Events occurring over time included: supraventricular tachycardia in 5 cases (20%), atrioventricular block in 1 (4%), right heart failure in 10 (40%), stroke in 1 (4%) and syncope in 3 (12%). Pulmonary hypertension occurred in 4 cases (16%). Follow-up was uneventful in 4 patients at the time of analysis (16%). Treatment included: diuretics in 13 cases (52%), beta-receptor antagonists in 11 (44%), amiodarone in 4 (16%), and 2 patients had no treatment (8%). NYHA class I, II, III and IV range was respectively: 2 cases (8%), 11 (44%), 4 (16%) and 7 (28%). Death occurred in 5 cases (20%), at the age of 2 ± 1.7y (median 1.5y, range 0.5 to 4.8y) and 0.6 ± 0.7y after diagnosis (median 0.5y, 1week to 2y), from HF in 1 and sudden death in 4. Ten patients (40%) underwent heart transplantation, at the age of 12.5 ± 6.7y (median 14.8y, range 3.7 to 22.1y) and 5 ± 7y after diagnosis (median 2y, 0.6 to 19.4y). Overall death or transplantation occurred in 60% of the cases. Age at diagnosis was a predictive factor for death (1.4y in deceased patients versus 6.6y in alive cases, p = 0.03).
Conclusion: RCM prognosis is poor in children with high rates of death or transplantation. Infant age at diagnosis is a predictive factor of bad outcome.

Keywords: restrictive cardiomyopathy, children, pronostic factor, outcome

P-124

End diastolic forward flow in the RV outflow tract is a novel marker for LV diastolic dysfunction in the patients with repaired tetralogy of fallot

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Background and Aim: Accumulating evidence suggested end-diastolic forward flow in the right ventricular outflow tract (EDFF) is a marker for increased diastolic stiffness of the right ventricle (RV), which is developed either by concentric or eccentric remodeling, in the patients with repaired tetralogy of Fallot (TOF). As the right ventricular remodeling can affect function of the left ventricle (LV) through interventricular cross-talk, we tested our hypothesis that EDFF can be a marker for LV diastolic dysfunction in the patients with repaired TOF.
Methods: Consecutive 143 patients who underwent cardiac catheterization after TOF repair were enrolled in this study. The EDFF was measured at the central echo lab in our institute and its interactions with the catheter based ventricular property were analyzed.
Results: The age of TOF repair and catheter exam in the patients with EDFF (E-TOF, N = 73) and those without (N-TOF, N = 70) were 2.5 ± 3.3, 3.4 ± 4.4 years (p = 0.14), and 10.3 ± 11.3, 9.7 ± 8.6 years (p = 0.73), respectively. While hemodynamic index including cardiac index, heart rate, central venous pressure, end diastolic pressure (EDP) and volume (EDV) were similar between two groups, RV volume (169 ± 62, 145 ± 47%N, p = 0.013) and pulmonary regurgitant fraction (32 ± 20, 22 ± 28%, p = 0.023) in E-TOF were higher than those in N-TOF. Although LVEDV was positively corelated with RVEDV (p = 0.0006), it was markedly suppressed when augmented EDFF velocity-time integral (p = 0.036) and longer EDFF time duration (p = 0.044) were observed in E-TOF. Intriguingly,

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augmented EDFF velocity was strongly corelated with augmented LVEDP (p = 0.0076). EDFF was independent of plasma levels of BNP, NT-pro BNP or HANP.
Conclusion: While appearance of EDFF represents RV volume is in the volume range of steepened end-diastolic pressure-volume relationship in the patients with repaired TOF, waveform analyses of EDFF might provide novel information on LV volume intolerance due to interventricular cross-talk, which is independent of serum levels of natriuretic peptides. Since excessive RV volume loading coupled with LV diastolic dysfunction indicates strong candidacy of reintervention for pulmonary valve in the patients with postoperative TOF, prospective research to assess the role of EDFF in determining reintervention would be warranted.

Keywords: End Diastolic Forward Flow,Tetralogy of Fallot

P-126
Quantitative analysis of left ventricular energetic performance in children undergoing anthracycline chemotherapy

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Background and Aim: Anthracyclines are widely used to treat childhood cancers; however, their cardiotoxicity is the leading non-oncological cause of mortality in childhood cancer survivors (CCS). It is well-understood that adverse cardiac remodelling can develop from anthracycline cardiotoxicity, and may precede the development of ventricular dysfunction and heart failure. Vector flow mapping (VFM) is a novel echocardiographic modality which allows for the quantitative assessment of intracardiac fluid dynamics and energetics as markers of abnormal structural alterations in the heart and therefore may be a more sensitive investigation to identify cardiotoxicity. Hence, to enhance the current risk stratification and echocardiographic monitoring in children undergoing anthracycline chemotherapy, this study aimed at assessing the performance of various echocardiographic modalities, including VFM, and correlating them with cardiac biomarkers as potential clinical tools for the quantitative assessment of early adverse cardiac remodelling for early diagnosis of anthracycline cardiotoxicity.
Methods: This was a single-centre, prospective quantitative serial study of 50 children at a tertiary paediatric oncology centre from 2019-2021. Echocardiographic parameters investigated from 269 echocardiograms included M-Mode, standard 2D, tissue Doppler imaging, and speckle-tracking echocardiography, with high sensitivity assay for troponin-T (hs-TnT) and N-terminal of pro-B-type natriuretic peptide (NT-proBNP) as cardiac biomarkers. VFM was utilised to visualise intra-cardiac vortex parameters, energy loss and intraventricular pressure gradient.
Results: Commensurate with previously described epidemiology, the majority (n = 30/50, 60.0%) of patients were diagnosed with leukaemia at a mean age at diagnosis of 9.07 years old. 2 (4.0%) patients developed cardiotoxicity (left ventricular fractional shortening <28%) with a mean cumulative doxorubicin dose of 152.29 mg/m2. The mortality rate was 6.0% (n = 3/50). Our echocardiographic and biomarker results will be presented during the conference.
Conclusion: The mortality and cardiotoxicity rate were commensurate with existing literature.

Keywords: Energy Loss, Echocardiography, Cardio-oncology, Intraventricular Pressure Difference, Vortex, Relative Pressure Imaging

P-127
Use of "3D flow" MRI for accurate and rapid flow measurements in a TCPC phantom

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Background and Aim: 4D flow cardiac magnetic resonance imaging (MRI) provides a non-invasive method to measure hemodynamic conditions including blood flow rate and wall shear stress (WSS). Gated 4D flow acquisitions produce flow vs. time measurements over the cardiac cycle. Venous flow, such as within the total cavopulmonary connection (TCPC) in single ventricle patients, does not vary significantly as a function of time. Foregoing cardiac gating when assessing venous flow and acquiring data as a single time averaged phase reduces scan times. This "3-dimensional (3D) flow" method captures velocity encoding averaged over the entire cardiac cycle and can be utilized for measuring non-pulsatile flow.
Methods: Patient specific MRI data was used to segment and 3D print TCPC vessels and acquire flow profiles. An MR conditional flow pump, providing a constant flow rate, was used to simulate venous flow and valves were used to control flow distribution. The models were then scanned with 4D and 3D flow sequences using a Siemens Magnetom Aera 1.5 T MRI scanner. A 150 BPM simulated heartbeat was used for 4D flow cardiac gating. Models were scanned with a 1.3 mm and a 1.1 mm slice thickness to assess the impact of spatial resolution. The models were segmented and measured using the iTFlow software (Cardio Flow Designs, Tokyo Japan). Flow rates for each vessel and wall shear stress across the entire model were collected.
Results: 7 models were compared accounting for 28 flow rate measurements. 3D flow scans were completed in 2 minutes and 37 seconds. There was no significant difference (p = 0.50) between flow rates measured by 3D flow or 4D flow methods. Reducing the slice thickness of the 3D flow measurements from 1.1 mm to 1.3 mm did not significantly alter the flow rate measurements (p = 0.32). Additionally, there was no significant difference between the maximum and average WSS values (p = 0.94 and p = 0.69 respectively).
Conclusion: 3D Flow can be used for flow rate and WSS measurements in non-pulsatile flow conditions, reducing scanning time. This functionality has been demonstrated in TCPC phantom

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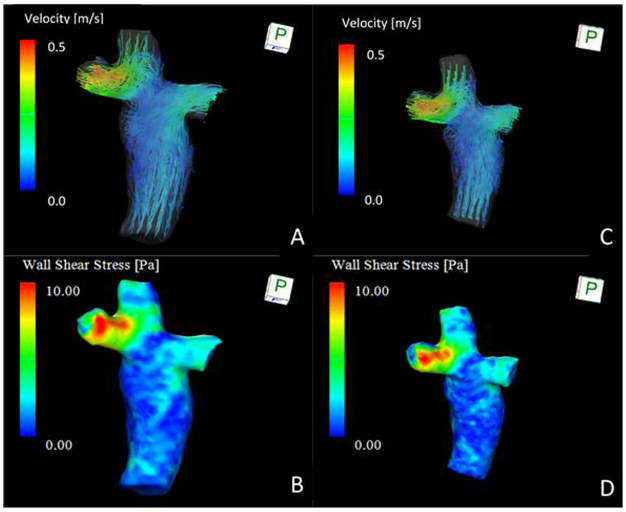
models, and further research is needed to establish this process's feasibility in clinical wall shear stress measurements.

Keywords: 4D Flow, Fontan, MRI, Pediatric Cardiology

Comparison of 4D and 3D flows



Examples of (A) 4D flow velocity flow lines, (B) 4D flow wall shear stress distribution, (C) 3D flow velocity flow lines, and (D) 3D flow wall shear stress distribution.

P-128

Clinical features, electrocardiographic, and echocardiographic findings in pediatric patients with isolated left ventricular non-compaction

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Background and Aim: Left ventricular non-compaction is rare cardiomyopathy that occurs following an early arrest in endomyocardial morphogenesis. This study presents clinical and electrocardiographic (ECG) findings, diagnostic features, treatment, and follow-up of pediatric patients diagnosed with LVNC.

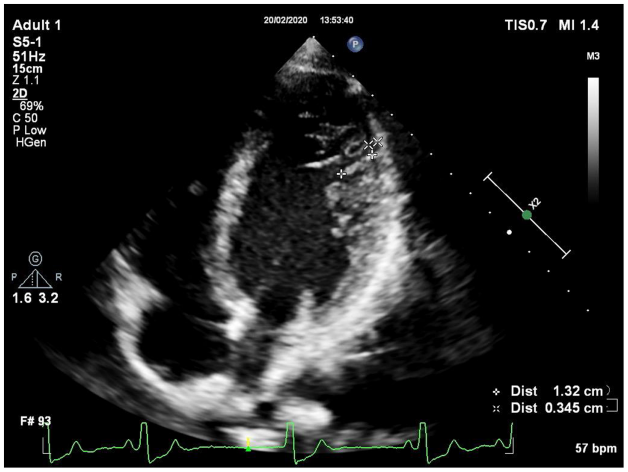
Methods: We retrospectively reviewed children with left ventricular noncompaction from January 2010 to June 2020 in a tertiary referral center. Patients with congenital heart disease, metabolic illnesses, systemic diseases, neuromuscular diseases were excluded.

Results: Fifty-five children were diagnosed with left ventricular noncompaction over the study period. 32 patients (58.2%) were male, the median age of presentation was 8.5years (1 month-17.9 years), and 13 patients (23.6%) were admitted under the age of 1 year. The median follow-up of the study was 19 months (1-121 months). Fourteen (25.5%) presented with cardiac dysfunction (EF <45%), and 2 presenting with resuscitated/aborted cardiac arrest. ECG abnormalities were present in 78.2%. Fragmented QRS was observed in the ECGs of six patients, and the QTc duration was 450ms and above in 17 patients (30.9%). Premature ventricular contractions were observed in 10 patients, non-sustained VT in 1, and sustained VT in 8 patients. Ventricular fibrillation developed in half of the 8 patients with sustained VT. ICDs were placed in a total of 6 patients to prevent sudden cardiac death. One patient with a complete AV block and one patient with Long QT syndrome and severe bradycardia underwent permanent pacemaker implantation.The mean end-systolic LV ratio of non-compacted to compacted myocardium was 2.5, and the mean EF was 58.8% (28%-78%) at echocardiography. 32 patients were diagnosed as LVNC with cardiac MRI findings. However, the data of only 16 patients with sufficient image quality could be evaluated, retrospectively. The mean EF was 58.5%±9,7 and only 4 patients had a decreased EF (< 53%). One patient had Late gadolinium enhancement (LGE) sequence, and only one patient had wall motion abnormalities. Five (9.1%) patients died. All patients with sudden death (n = 5) had abnormal cardiac dimensions or cardiac dys function. One patient with increased cardiac dimensions and normal function diagnosed as Long QT syndrome died suddenly.

Conclusion: Left ventricular noncompaction has a high mortality rate and is strongly associated with arrhythmias in children. Preceding cardiac dysfunction or ventricular arrhythmias are associated with increased mortality. Children with normal cardiac dimensions and normal function are at low risk for sudden death.

Keywords: left ventricular noncompaction, arrhythmias, cardiomyopathies, children

Image 1



Apical 4 chamber image demonstrates non-compacted myocardium with prominent trabeculations and deep sinuses.

Preventive

P-129

Physical activity of single ventricle patients during the covid-19 pandemic

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Background and Aim: It is well known that Fontan patients tend to a sedentary lifestyle with low physical activity levels and their objective exercise capacity is reduced. Currently the COVID-19

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pandemic has a significant impact on the physical activity levels of the whole population. This study investigates physical activity levels of Fontan patients during the COVID-pandemic, and their relationship to exercise capacity, heart rates, cardiac function at cardiovascular magnetic resonance (CMR), and biomarkers.
Methods: CMR, exercise testing, 24h-ECG, and blood samples were prospectively performed in 38 Fontan patients, of which 15 were females (40%) and 18 (47%) had a single left ventricle (LV). Time interval from Fontan operation was in median (IQR) 10 (8-15) years. Physical activity was assessed by accelerometer during 7 consecutive days of regular school/work. Moderate intensity physical activity was defined as > 2296 counts/minute and vigorous physical activity as > 4012 counts/minute. Patients with moderate-to-vigorous physical activity (MVPA) below 60minutes per day were categorized as inactive as recommended by the WHO. Parameters of exercise capacity included maximal oxygen uptake, maximum work rate, and maximal heart rate.
Results: Daily MVPA was in median (IQR) 40.2 (27.6-56.5) minutes and 7/18 (39%) patients reached the recommended 60minutes/day of MVPA. Daily minutes of MVPA did not correlate with gender, age, single left ventricle, years from Fontan surgery, mean heart rate, ventricular volumes, and ejection fraction at CMR, cardiac biomarkers, or exercise capacity.
Conclusion: Only 39% of the Fontan patients meet the recommendation for daily MVPA. The MVPA during the pandemic in our patients' cohort is lower than the one reported in Fontan patients before the pandemic. These current changes in behavioral habits may increase the risk for habitual sedentary lifestyles in Fontan patients.

Keywords: single ventricle, Fontan, physical activity, COVID-19

Imaging / Functional assessment

P-130 / Moderated Poster
Risk factors for perioperative brain lesions on MRI in infants with severe congenital heart disease: a european collaboration

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Background and Aim: Infants with Congenital Heart Disease (CHD) are at risk for perioperative brain injury. The aim was to identify risk factors for perioperative brain lesions in infants with CHD.
Methods: Infants with Transposition of the Great Arteries (TGA), Single Ventricle Physiology (SVP) and Left Ventricular Outflow Tract and/or aortic arch obstruction (LVOTO) undergoing cardiac surgery within 6 weeks after birth [gestational weeks at birth median (range) = 39.0 (34.9-41.9)] from 3 European prospective cohorts were included. White matter injury (WMI), arterial ischemic stroke (AIS) and cerebral sinovenous thrombosis (CSVT) were scored on pre- and postoperative MRI [postnatal age at surgery in days median (range) = 10 (3-41); days from surgery to MRI median (range) = 10 (4-76)]. Risk factors were assessed with univariate group differences followed by stepwise multivariate logistic regression. Results are presented as adjusted Odds Ratios (OR) with 95% confidence intervals (CI).
Results: 180 infants underwent preoperative brain MRI of whom 146 also underwent postoperative imaging. Induced vaginal delivery (OR 2.23, 95% CI 1.06-4.70) was a risk factor for preoperative WMI and remained significant when including gestational age at birth in the model. Balloon atrial septostomy was associated with preoperative brain injury (WMI OR 2.51, 95% CI 1.23-5.20; AIS OR 4.49, 95% CI 1.20-21.49). Postoperatively, SVP was a risk factor for new WMI (OR 2.88 95% CI 1.20-6.95). Independent risk factors for new postoperative AIS included younger postnatal age at surgery (OR 1.18, 95% CI 1.05-1.33) and selective cerebral perfusion, particularly at deep hypothermia (OR 13.46, 95% CI 3.58-67.10). New postoperative CSVT was linked to TGA diagnosis (OR 13.47, 95% CI 2.28-95.66), delayed sternal closure (OR 3.47 95% CI 1.08-13.06) and lower minimum intraoperative temperatures (OR 1.22, 95% CI 1.07-1.36).
Conclusion: Modifiable risk factors for brain injury in CHD include delivery planning and timing of surgery. This may present opportunities to personalize treatment to minimize the risk of perioperative brain injury. In keeping with previous literature, BAS was

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associated with increased risk of preoperative brain injury and infants with SVP were at higher risk for postoperative WMI. Further research is needed to optimize neonatal cerebral perfusion techniques and to confirm the relationship between CSVT and perioperative factors.

Keywords: Neonatal, Brain Injury, Risk Factors

Preventive

P-131

The impact of gestational diabetes in pregnancy on the cardio-vascular system of children at one year of age

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Background and Aim: Gestational diabetes mellitus (GDM) is a common complication in pregnancy. The effect of GDM on the cardiovascular system after birth is still unclear.
Methods: 205 pregnant women were included in the prospective controlled observational study between August 2015 and December 2018. Patients with GDM were assigned to the study group (n = 99), whereas (n = 107) healthy women served as controls. Postnatal follow up of their offspring was performed at 12 months of age. All included children (n = 125) underwent a specific standardized protocol including anthropometric data such as weight, height, body mass index (BMI), blood pressure (BP) recordings and ultrasound measurements of the abdominal aortic Intima-Media-Thickness (IMT). Furthermore, at least 10 minutes 3-channel electrocardiogram recording was done to evaluate the autonomic nervous system (ANS) by Phase Rectified Signal Averaging.
Results: There were no significant differences in anthropometric data between the groups, neither in the blood pressure nor in the Intima-Media-Thickness of the Aorta abdominals. However, in the study group, significantly lower average acceleration capacity (AAC) (study group -20,10 ± 3,04 ms, control group -18,87 ± 4,00 ms, p = 0,02) was found, indicating ANS activation at one year of age.
Conclusion: Further studies are required to determine if these results are persistent and if these findings have long-term effects.

Keywords: gestational diabetes, pregnancy, cardiovascular outcome, autonomous nervous system, fetal programming

P-132

First in man data on diving physiology in fontan patients

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Background and Aim: Swimming and diving are popular recreational activities. As congenital heart disease, especially patients with univentricular hearts after Fontan palliation are thought to have reduced physiological capacities for compensation of submersion associated physiological demands, current guidelines put restraints on this group of patients. Although these restrictions on doctoral advice place a significant burden on affected patients, it is especially interesting that these guideline recommendations are merely based on physiological assumptions, ie. expert consensus. A recent study by Paech et al. presented the first in vivo data on the effects of immersion in Fontan patients, stating no major adverse events in their study group as well as comparable physiologic adaption as reported in the literatur for healthy people. Yet, submersion was not reflected in this study, this current study therefore aimed to conduct a first study for the evaluation of the effects of submersion and apnea diving in Fontan patients.
Methods: : A control group of healthy adults as well as patients recruited from the Heart Center Leipzig, Department of pediatric cardiology, underwent standardized diving protocol including a static as well as dynamic apnea phase. Physiologic data were recorded.
Results: This study presents the first structured data on diving physiology in Fontan patients compared to healthy probands. Data are shown in illustrative diagrams. There were no adverse events. The physiologic response to diving seems to be comparable between healthy probands and Fontan patients.
Conclusion: Although, healthy probands did reach a much better performance, the basic mechanisms of physiological adaption seem comparable between healthy probands and Fontan patients.

Keywords: congenital heart disease, Fontan, diving, swimming, physiology, sports cardiology

P-133

Physical activity in high- risk pregnancies associated with assisted reproductive technology and fetal congenital heart disease

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Background and Aim: Physical activity before and during pregnancy is associated with health benefits for both, mother and fetus. The majority of pregnant woman are not physically active in pregnancy and the WHO guidelines do not include information on physical activity (PA) for specific target groups.
The aim of our study was to elaborate whether assisted reproduction technologies (ART) and/or the diagnosis of fetal congenital heart defects (CHD) influences the level of PA in pregnant women and if there is a difference between the PA behavior before and during pregnancy.
Methods: A non-interventional, cross-sectional, monocentric study based on two standardized questionnaires on physical activity was conducted.
Results: In total, N = 158 pregnant women were included. The following four groups were compared to each other: 'ART' (n = 18), 'CHD' (n = 25), 'ART and CHD' (n = 8) and 'healthy controls' (n = 107). Women after ART showed a significantly reduced level of physical activity (p = .014) during pregnancy compared to women who got pregnant naturally. Additionally,

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less (p <.001) and lighter (p = .002) physical activity was observed in all groups during pregnancy compared to those before pregnancy. An increase in maternal age increases the likelihood of CHD (p <.001) and decreases the level of physical activity before pregnancy (p = .012).
Conclusion: The overall physical activity level decreased during high- risk pregnancies. Further research for the specific target groups is highly recommended in order to promote and increase physical activity in ART and CHD pregnancies.

Keywords: Assisted reproductive technology, congenital heart disease, pregnancy, fetal, physical activity

P-134 / Moderated Poster
Common carotid artery characteristics in patients with repaired aortic coarctation compared to other cardiovascular risk factors

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Background and Aim: Increased common carotid artery (CCA) intima media thickness (cIMT) is a well-known risk factor for cardiovascular morbidity and mortality. cIMT thickening has been described in patients with repaired aortic coarctation (CoA), but data on mechanism and clinical relevance in this population are scarce. Our aim was to gain mechanistic insights into cIMT thickening of patients with repaired CoA by comparing their wall architecture to patients with coronary artery disease (CAD), other congenital heart diseases (oCHD), and healthy controls.
Methods: This is a retrospective analysis of right CCA ultrasound images comparing morphology in patients with repaired CoA, oCHD, and CAD to healthy controls. CIMT and lumen diameter (LD) were determined using semiautomated analysis software. Linear regression analyses were performed correcting for relevant covariates.
Results: A total of 310 subjects were included (CoA (n = 58), oCHD (n = 96), CAD (n = 68) and healthy controls (n = 88)). Both, patients with repaired CoA and CAD had significantly increased cIMT (B = 0.1, p<0.001; B = 0.15, p<0.001) and cIMT/LD ratios (B = 0.01, p = 0.012; B = 0.03, p<0.001), while LD was increased only in CoA patients (B = 0.37, p = 0.001; B = 0.08, p = 0.528). Comparing CoA to CAD patients, LD was significantly larger (B = 0.3, p = 0.021) and IMT/LD less (B = -0.01, p = 0.005), but IMT was not significantly different between thr groups. Furthermore, patients with repaired CoA had decreased CCA stiffness (p = 0.005). CCA characteristics in the oCHD group were not significantly different from controls.
Conclusion: The mechanism of cIMT thickening in patients with repaired CoA may differ from CAD. While there is concentric remodeling in the latter, we see predominant eccentric remodeling in the CoA group, which could be due to increased flow as a result of compliance mismatch at the CoA repair site. We therefore suggest that the prognostic value of cIMT in post-CoA patients should be validated separately prior to using it to guide clinical management in this group.

Keywords: Aortic coarctation, arterial stiffness, common carotid artery, congenital heart disease, dimension, intima media thickness

P-135 / Moderated Poster
Does pulse wave analysis by digital plethysmography reliably assess arterial stiffness in patients with congenital heart disease?

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Background and Aim: Arterial stiffness is an important predictor of cardiovascular morbidity and mortality in the general population. More recently, researchers have become interested in arterial changes of patients with congenital heart disease. While abnormalities are seen in several lesions, mechanism, location and extent of arterial stiffening in this population appear to be diverse. In addition, the prognostic value of arterial abnormalities in CHD remains unclear. It was the aim of this study to evaluate whether pulse wave analysis (PWA) by photoplethysmography provides reliable results that correspond to non-invasive PWA of the central arterial wave form.
Methods: We included 225 subjects (77 controls, 54 bicuspid aortic valve, 46 aortic coarctation, 19 Marfan syndrome, 29 single ventricle(SV)). SphygmoCor XCEL (Atcor Medical) was used to measure central augmentation index (cAix) and carotid-femoral pulse wave velocity (PWV). Digital plethysmography (DPA, Meridian) provides variables based on the pulse curve and acceleration curve which represent different phases of the cardiac cycle. The so-called aging index (AI) is a global index arterial aging which is based on the acceleration curve. All measurements were performed in duplicate in the same physiological state. First we assessed intraclass correlation coefficient for repeated measurements. Variables with ICC>0.9 (p<0.001) were used for subgroup analyses. Correlation analyses were performed.
Results: CAIx and PWV showed optimal reproducibility with ICC >0.96 (p<0.001). Acceleration curve parameters of the DPA also had excellent reproducibility (ICC 0.9-0.95, p<0.001). However, none of the DPA parameters derived from the basic pulse curve had acceptable reliability (all ICC<0.9). Reproducibility was similar in all groups except the SV group, which was therefore excluded from further correlation analyses. CAIx correlated strongly with AI (R = 0.722, p<0.001) and other indices of the acceleration curve (R = 0.6-0.7, p<0.001) while PWV had weak correlations with DPA indices (R = 0.2-0.4, p<0.003).
Conclusion: Patients with a biventricular circulation and controls have excellent reproducibility of cAIX, PWV and DPA parameters of the acceleration curve, esp. AI. Correlation of AI with cAIx is strong, and could be used as a surrogate. PWV by contrast correlates weakly with DPA measurements. In SV patients, right arm measurements of arterial stiffness appear to be unreliable, possibly due to prior modified BT-shunt.

Keywords: arteriell stiffness, augmentation index, pulse wave velocity, digital plethysmography

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P-136
CHIC-D - cardiovascular health in children with type 1 diabetes

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Background and Aim: Patients with type 1 diabetes (T1D) have an increased risk for cardiovascular disease, with the impact of metabolic control and blood pressure on the cardiovascular system starting already in childhood. Our objective is to use novel, highly sensitive methods for early cardiovascular risk evaluation and treatment monitoring for paediatric patients with T1D, to establish a detailed phenotype, time course and determinants for cardiovascular changes in children with T1D to facilitate cardiovascular prevention.
Methods: Children (6-15,99yr) with T1D duration of ≥5 years were randomly selected from the pediatric diabetes registry SWEDIABKIDS. We use ultra-high frequency ultrasound, enabling separate visualization of the layers in the arterial wall combined with measurements of pulse wave velocity (PWV), endothelial function, blood pressure and baroreceptor sensitivity (BRS) as a measure of autonomic dysfunction. 50 children with T1D and 29 healthy controls have been included thus far.
Results: Preliminary results show an approximately 11% increase in dorsal pedal (DP) intima- media thickness (IMT) among the children with T1D ($p = 0.05$). A tendency towards increased intima- thickness (IT) in the radial artery and media- thickness (MT) in DP among the children with T1D was also seen ($p = 0.09$, $p = 0.09$) as well as a negative correlation between DP IMT and GFR (-0.32 $p = 0.05$) and a positive correlation between DP IMT and aortic PWV and HbA1c (0.38 $p = 0.01$, 0.32 $p = 0.04$). There were no significant differences in baroreceptor sensitivity between the two study groups.
Conclusion: Increased DP IMT in this well treated cohort of children with T1D (HbA1c 48.1 ± 5.9 mmol/mol) may be an important marker for early vascular impact. The correlation between DP IMT and HbA1c further supports the importance of metabolic control and striving for normoglycemia. The correlation between PWV and GFR indicates a connection between micro-and macro vascular disease. Using our sensitive methods preventive treatment strategies may be tested in the future.

Keywords: Cardiovascular, Prevention, Ultrasound, Type 1 Diabetes

Table and correlations

	T1D patients n=46	Healthy controls n=26	p- value
Age (years)	12.0 ± 2.3	11.2 ± 2.7	0,156
Weight (kg)	48.3 ± 14.3	45.1 ± 14.6	0,358
Height (cm)	156.4 ± 14.8	151.6 ± 17.6	0,230
T1D duration (years)	7.7 ± 1.8	---	---
HbA1c (IFCC mmol/mol)	48.1 ± 5.9	30.6 ± 2.5	<0.001
HbA1c (DCCT %)	6.6 ± 0.5	4.9 ± 0.2	<0.001
Time in range%(B-glc 4-10 mmol/mol)	64.0 ± 8.5	---	---
Time in target%(B-glc 4-7.8 mmol/mol)	42.6 ± 9.8	---	---
SBP (mmHg)	105.8 ± 6.9	103.5 ± 8.3	0,217
DBP (mmHg)	65.4 ± 4.0	62.1 ± 5.4	0,004

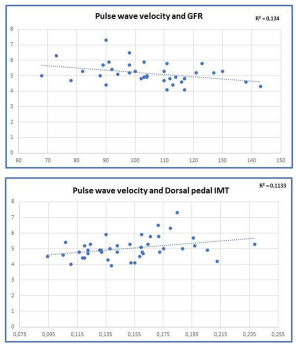

Table including results and demographic data, scatterplots illustrating correlations.

P-137
A case of interrupted aortic arch in a 4 day old with abnormal pulse oximetry screening

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Background and Aim: Screening for congenital heart disease involves a cardiac examination and pulse oximetry prior to discharge and ideally 24 hrs after the birth of a newborn. Unfortunately these processes are less likely to diagnose certain lesions including aortic arch anomalies.
Methods: The clinical information and investigations were reviewed at the local hospital. Final diagnosis and further imaging was obtained from the tertiary unit where the patient was transferred.
Results: A 4 day old term male infant was reviewed on the postnatal ward with abnormally low discharge pulse oximetry. He remained an inpatient at day of life 4 due to a maternal history of pyrexia following delivery. A normal discharge examination had been completed on day of life 2. The antenatal course was uncomplicated and there was a normal anatomy scan. On initial examination the infant was pink and comfortable. At the time of discharge on day 4 post duct saturations were 80% recorded by post natal staff and alerted paediatric team. He was peripherally cool and had a hyperactive precordium with diminished peripheral pulses. Pre and post ductal saturations were normal on repeat. A blood pressure differential >20mmHg systolic existed between the right arm and the remaining limbs. An arterial blood gas showed a metabolic acidosis. A chest x-ray was normal. An ECG performed showed extreme Superior axis with normal sinus rhythm. The patient was discussed with a tertiary paediatric cardiology centre and transferred there on prostaglandin. The final diagnosis was confirmed by ECHO and CT angiogram showing a type B interrupted aortic arch with the left subclavian arising from the ductus, a hypo plastic left ventricular and a peri membranous VSD. He underwent a Yasui procedure at day of life 8 and had an uncomplicated post-operative course. Further surgery is planned for 6-8 months of life.
Conclusion: Interrupted aortic arch is a cardiac emergency that can lead to neonatal cardiac failure. Prompt diagnosis is crucial in initiating prostaglandin as a bridge to surgical intervention. Our case represents the importance of mandatory thorough clinical exam and pulse oximetry screening prior to discharge helps in diagnosing left sided obstructive lesions as the ductus arteriosus closes.

Keywords: Interrupted aortic arch, aortic arch anomalies

P-138 / Moderated Poster
Lack of improvement in the postnatal detection of heart defects

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Background and Aim: Early detection of severe congenital heart defects (CHDs) is crucial for treatment and outcome. This study investigated the impact of new pre- and postnatal screening methods for detection of severe CHDs and reports national data.
Methods: Data on all pregnancies and live-born children with severe CHDs in Norway from 2004- 2005 and 2017-2020 were extracted from the Medical Birth Registry of Norway and Oslo

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University Hospital's Clinical Registry for CHDs. The screening program in Norway includes prenatal ultrasound in week 18-20 and clinical examination on day one or two after birth. The three vessel and trachea view in prenatal ultrasound and pulse oximetry screening within the first 24 hours after birth were gradually implemented in Norway during the study period. The indications for cardiac examination after birth were retrieved from medical records.

Results: The prenatal detection rate increased from 31 to 66% from 2004-2005 to 2017-2020 (35% increase, 95% CI 29%-40%). Double amount of time was required in the late cohort to collect an equivalent number of live-born children with postnatal detected severe CHD (169/2 years vs 174/4 years). There was no significant improvement in detection of CHDs in live-born children before postnatal discharge (69 vs 74%, 95% CI -3%-13%). Symptoms and comorbidity were the indications for cardiac examination and diagnosis before discharge in 40% of the cases in 2004-2005 and in 38% in 2017-2020. Approximately one quarter of the children with severe CHDs were discharged from hospital undiagnosed (29% in 2004-2005 and 24% in 2017-2020). The contribution of pulse oximetry was limited (2 vs 8 %,) Figure 1 presents the registered causes of postnatal detection.

Conclusion: Prenatal diagnosis of severe CHDs has markedly increased in Norway since 2005. The detection rate of severe CHDs before postnatal discharge did not change despite nationwide implementation of pulse oximetry screening during the period. About one quarter of live-born children with severe CHDs still leave hospital undiagnosed after birth.

Keywords: Congential heart defect, detection

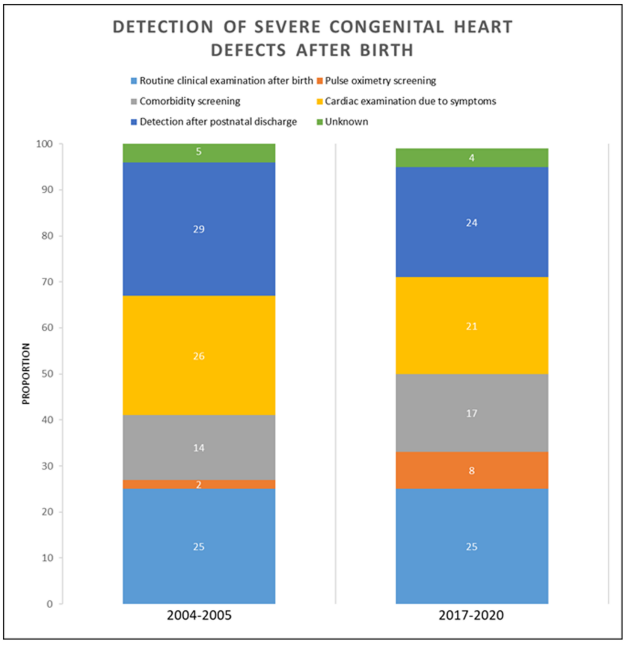

Figure 1.

P-139 / Moderated Poster

Preclinical aortic atherosclerosis in adolescents with chronic disease

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Background and Aim: Adolescents with chronic disease are often exposed to inflammatory, metabolic, and hemodynamic risk factors for early atherosclerosis. Since post-mortem studies have shown that atherogenesis starts in the aorta, the 'Cardiovascular Disease in Adolescents with Chronic Disease' (CDACD) study investigated preclinical aortic atherosclerosis in adolescents with chronic disease, and studied the association with disease-associated risk factors.

Methods: The cross-sectional CDACD study enrolled 114 adolescents 12-18 years old with chronic disorders including juvenile idiopathic arthritis, cystic fibrosis, obesity, corrected coarctation of the aorta, and healthy controls. Cardiovascular Magnetic Resonance (CMR) was employed to assess aortic pulse wave velocity (PWV) and aortic wall thickness (AWT), as established aortic measures of preclinical atherosclerosis. Conventional carotid intima-media thickness (cIMT) was measured using ultrasonography.

Results: CMR showed a higher aortic PWV, which reflects aortic stiffness, and higher AWT in all adolescent chronic disease groups, compared to controls ($p<0.05$). Diastolic blood pressure ($\beta = 0.310$), fasting glucose ($\beta = 0.253$), and classical monocyte numbers ($\beta = 0.198$) were identified as predictors for aortic PWV, using multivariable regression analysis. AWT was predicted by body mass index ($\beta = 0.230$) and fasting glucose ($\beta = 0.211$), next to aortic lumen area ($\beta = 0.422$). cIMT was only higher in adolescents with CoA, compared to controls ($p<0.001$).

Conclusion: Adolescents with chronic disease showed enhanced aortic stiffness and wall thickness, compared to controls. These aortic measures of preclinical atherosclerosis may indicate accelerated atherogenesis. Our findings emphasize the importance of the aorta for the assessment of early atherosclerosis, and the need for tailored cardiovascular follow-up of children with chronic disease.

Keywords: Atherosclerosis, children, adolescents, chronic disease, inflammation, cardiovascular magnetic resonance

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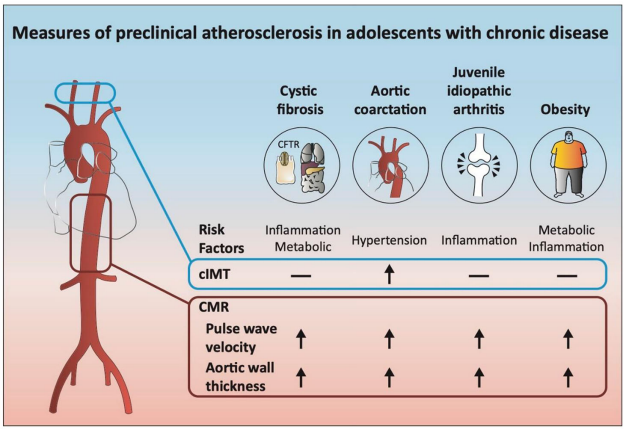
Graphical abstract



Summary of the CDACD study findings following CMR assessment of the aortic pulse wave velocity and aortic wall thickness, next to conventional carotid intima-media thickness measurements using ultrasonography, in adolescents with chronic disease. Upward arrows (↑) indicate an increase compared to the healthy controls, while downward arrows (↓) indicate a decrease, and hyphens (-) indicate no difference

P-140

Ventricle morphology determines physical performance in young single ventricle patients

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Background and Aim: In surgically palliated single ventricle Fontan patients (FP) ventricle preload is dependent on systemic venous pressure and pulmonary vascular resistance. In FPs muscle mass positively correlates with maximum oxygen consumption (maxVO2), and long-lasting exercise intervention improves anaerobic threshold (AT). We studied whether systemic ventricle morphology (SVM) and size of pulmonary arteries (PAs) influence cardiopulmonary performance.
Methods: 28 pediatric stable FPs with right (RV-SVM) and left ventricle morphology (LV-SVM), and normal peripheral oxygen saturation underwent cardiopulmonary exercise testing. AT, maxVO2, forced vital capacity (FVC), forced expiratory volume in 1 second (FEV1) and VE/VCO2 slope were determined. These data were correlated with indexed pulmonary resistance prior to Fontan conversion, and combined branch pulmonary artery size index at the latest magnetic resonance imaging (McGoon index, McGI). Non-parametric Mann-Whitney U-test and multiple regression analysis tested statistical difference.
Results: In multiple regression analysis using maxVO2 and McGI as dependent variables, only SVM was predictive ($R2 = 0.803$, $p<0.03$ and $R2 = 0.720$, $p<0.025$, respectively).
Conclusion: Young Fontan patients have about 40% lower than normal cardiopulmonary performance with LV-SVM superior to RV-SVM. All patients had restrictive lungs but with RV-SVM this finding was significantly inferior. Ways of lung recruitment should be included in exercise protocols of young Fontan patients.

Keywords: univentricular heart, Fontan circulation, maximal oxygen uptake, cardiopulmonary exercise testing, pulmonary vascular imaging

AEPC_ABSTRACT_TABLE

	All	RV-SVM	LV-SVM	p
Combined PA-branch size (Z, n=26)	0.18 ± 1.2	0.12 ± 1.17	0.33 ± 1.3	0.513
McGoon index (n=26)	1.9 ± 0.4	1.8 ± 0.4	2.2 ± 0.4	0.035
PVRI (Wood units, n=17)	2.1 ± 0.9	2.0 ± 0.7	2.3 ± 1.2	0.814
MaxVO2 (ml/kg/min, n=28)	28.5 ± 6.2	26.3 ± 5.5	32.5 ± 5.6	0.004
MaxVO2, % of normal	59,6 ± 14.1	57.5 ± 11.3	63.3 ± 18.1	0.443
AT (ml/kg/min, n=28)	18.9 ± 5.0	17.5 ± 4.3	21.3 ± 5.0	0.074
AT, % of VO2	42,6 ± 11.2	39,5 ± 10,9	48,0 ± 9,0	0.045
FVC (Z, n= 26)	-2.1 ± 1.6	-2.6 ± 1.6	-1.1 ± 1.2	0.008
FEV1 (Z, n=26)	-2.4 ± 2.1	-3.2 ± 2.1	-0.9 ± 1.2	0.002
VE/VCO2 (n= 28)	32.4 ± 6.3	32.3 ± 6.1	32.7 ± 7.1	0.822

Results from cardiopulmonary exercise test and pulmonary vascular imaging

P-141

A case of mRNA COVID-19 vaccine-associated myocarditis in a child

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Background and Aim: Accumulation of cases is needed to determine whether vaccines should be recommended for children because of their potential to cause myocarditis in healthy children.
Methods: We report a case in which changes in laboratory data, electrocardiogram (ECG), and magnetic resonance imaging (MRI) were tracked at our hospital.
Results: A 12-year-old girl developed fever a day after receiving the second dose of the COVID-19 vaccine. Three days after vaccination, she also developed chest pain and went to a hospital. ECG showed ST-T segment elevation. However, the symptoms were mild, and she was treated with antipyretics and analgesics. The next day, she visited the hospital again because she had mild chest pain. ECG showed a negative T-wave, and she was referred to our hospital. Her real-time reverse-transcription polymerase chain reaction tests for COVID-19 yielded negative results. Computed tomography revealed no anatomical abnormalities of the coronary arteries. The serum concentration of troponin T was elevated by 131 ng/L. Echocardiography showed the left ventricular ejection fraction to be 64%. MRI showed a normal T2 value on T2-weighted imaging; however, extracellular volume increased by 33%. Although the Lake Louise criteria was not met, we diagnosed the condition as myocarditis. She was hospitalized for 2 days and discharged without the need for steroids or gamma globulin treatment to relieve her symptoms. Although these findings improved 17 days after vaccination, late gadolinium enhancement was noticed on MRI.
Conclusion: The COVID-19 vaccine-related myocarditis (C-VAM) in this case was mild and like as cases in Europe and the United States. The risk of COVID-19 associated myocarditis is more than three times the risk of C-VAM. In addition, the mortality rate for COVID-19 associated myocarditis is higher than that for C-VAM. The need for a vaccine to protect populations from COVID-19 should be properly recognized. However, because the symptoms of C-VAM are mild, there may be many potential

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patients with C-VAM. Therefore, it may be advisable to avoid strenuous exercise for approximately 1 week after vaccination. Further research is needed to determine the long-term outcomes of C-VAM because of the late enhancement identified on MRI.

Keywords: COVID-19, myocarditis, mRNA vaccine, electrocardiogram, magnetic resonance imaging

P-143

The extreme athlete's heart: echocardiographic data from elite endurance winter athletes

Isabelle Schöffl[1], Jan Wüstenfeld[2], Othmar Moser[3], Volker Schöffl[4], Lukas Zimmermann[5], Martin Braun[6], Sven Dittrich[1], Paul Zimmermann[6]

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Background and Aim: As ski mountaineering (skimo), biathlon and nordic skiing involve the whole body, are performed at altitude, and involve uphill locomotion, they represent the most endurant sports imaginable. Because of the physical demands of endurance sports, pronounced cardiac remodeling in the form of hypertrophy of the LV can be expected. Therefore, the hearts of elite endurance winter athletes represent an ideal example for evaluating such adaptations. One readily available method for evaluating this, is two-dimensional echocardiography including speckle tracking analysis and ventricular global longitudinal strain (GLS).
Methods: We were able to evaluate the echocardiographic data from 32 elite world winter sports athletes, which were obtained between 2020 and 2021 during the annual medical examination in a multicenter approach. The age and sex matched data of the elite winter sports athletes, i.e. skimo, biathlon, and nordic skiing (14 women, 18 male athletes, age: 18-35 years) were compared for different echocardiographic parameters.
Results: The cardiac dimension of all three groups were comparable for most cardiac dimensions. However, systolic left ventricular function (LV-EF), intraventricular septum diameter (IVSd), left ventricular posterior wall diameter (LVPWd), and left atrial (LA) remodeling, were significantly lower in Skimo compared to the other athletes. The same could be observed for GLS. With respect to normal values, the LA size was much larger, whereas most of the other measurements were at the upper end of normal.
Conclusion: The echocardiographic two-dimensional analysis of elite winter sport athletes was able to show significant physiological differences in cardiac dimensions. Especially the nordic skiers and biathletes showed pronounced atrial remodeling as well as a more pronounced beginning eccentric hypertrophy of the left ventricle demonstrated in the speckle tracking analysis for the left ventricular global longitudinal strain (GLS). Since skimo athletes focus more on endurance training these differences could be be a consequence of differing training strategies. On the other hand, since nordic skiing and especially biathlon are traditional winter sports in Germany with a high media attention, the accumulation of the lifetime training quota leading to cardiac adaptations was probably lower in the less represented skimo athletes.

Keywords: hypertrophy, cardiomyopathy, exercise physiology, endurance sports

P-144 / Moderated Poster

Impact of lockdown on cardiopulmonary function in children with heart disease

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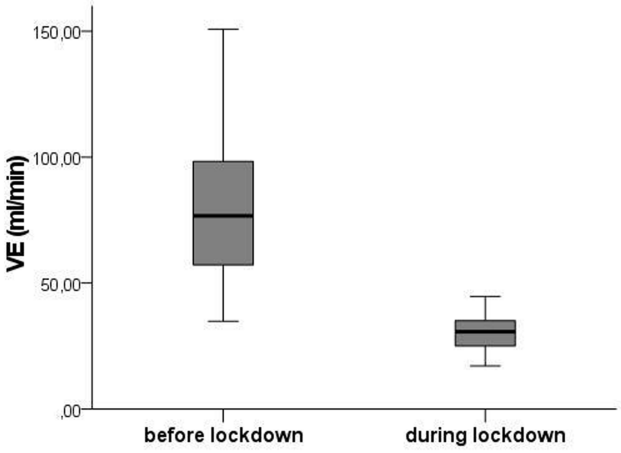
Background and Aim: The COVID-19 lockdown in Germany has led to the closure of sports facilities and to the closure of schools with their curricular physical activities (PA). However, physical activity is essential for improving or at least maintaining cardiopulmonary function assessed by VO2peak. VO2peak represents the best predictor for mortality and morbidity in patients with congenital heart disease (CHD). It is therefore essential to evaluate the effect of lockdowns on this important parameter in children with heart disease.
Methods: We evaluated data from cardiopulmonary exercise data from before lockdown with twin-paired data from during lockdown. The statistically approved twin-pairing was achieved by matching patients with similar heart disease, age, sex, and test method. The subjects consisted of a variety of patients who had received cardiopulmonary exercise testing in our department in the past 3 years.
Results: We were able to twin-pair 52 tests leading to 27 twins. A mean RER of more than 1.1 was achieved in both groups with comparable exercise times. There was a significant decrease of cardiopulmonary function (VO2peak: 35.7 ± 9.8 vs.30.4 ± 10.6) in our patient cohort along with a significant decrease in peak O2pulse (13.3 ± 4.1 vs. 11.4 ± 4.5), a surrogate parameter for stroke volume and most pronounced in peak minute ventilation (VE: 83.05 ± 29.08 vs.71,49 ± 24.96).
Conclusion: We observed a significant decline of V O_2peak during lockdown. This involved a loss of cardiac function assessed by O2pulse as well as a loss of pulmonary function assessed by V E. We believe that the decline of these important predictive parameters could be explained by the limited access to sports facilities as well as the restriction of regular daily movement as a consequence of closing schools and thus curricular PA. Measures need to be established to ensure access to physical activity for children suffering from heart disease during lockdowns.

Keywords: Corona, COVID-19, VO2peak, CPET, physical activity, sports

Peak minute ventilation (VE in ml/min)



The loss of cardiopulmonary function was most pronounced in peak minute ventilation as depicted here.

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P-145 / Moderated Poster

Investigating the relation between hemoglobin a1c with left ventricular remodeling patterns in children with type 1 diabetes mellitus

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Background and Aim: Pathological left ventricular (LV) remodeling is a well-known consequence of diabetes mellitus. However, data regarding the association between glycosylated hemoglobin (HbA1c) and LV remodeling patterns in children with type 1 diabetes (T1D) are limited. This study was aimed to investigate the relation between HbA1c with LV remodeling patterns in children with T1D.

Methods: A total of thirty children with T1D (aged 10 - 18 years; gender M (17) / F (13); duration of T1D ≥ 5 years; absence of hypertension, insulin therapy) were submitted to clinical evaluation, laboratory tests (HbA1c level), electrocardiogram, and echocardiography (left ventricular function and structure).

Results: According to the results, the types of pathological remodeling were distributed as follows: 5% (n = 2) – concentric hypertrophy, 10% (n = 4) – concentric remodeling and 5% (n = 2) – eccentric hypertrophy ((80.0% (n = 22) participants showed a normal LV geometry pattern).

The correlational study between the HbA1c and the LV parameters revealed a statistically significant positive correlation coefficient with aortic root diameter (mm) ($r = 0.7^{**}$, $p < 0.001$), left atrium (mm) ($r = 0.8^{**}$, $p < 0.001$), LV diastolic diameter (mm) ($r = 0.7^{**}$, $p < 0.001$), LV systolic diameter (mm) ($r = 0.7^{**}$, $p < 0.001$), 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) ($r = 0.5^{*}$, $p = 0.038$), LV Mass Index ($r = 0.5^{*}$, $p = 0.038$), LV diastolic volume (ml) ($r = 0.5^{*}$, $p = 0.025$), LV systolic volume (ml) ($r = 0.6^{**}$, $p = 0.01$), LV ejection fraction (%) ($r = 0.7^{**}$, $p = 0.001$), LV fractional shortening (%) ($r = 0.6^{**}$, $p = 0.002$).

Conclusion: The results of the study confirm that concentric remodeling, concentric and eccentric hypertrophy were detected in children with type 1 diabetes, and the increased value of the HbA1c was associated with a consensual and proportional increase in the values of the parameters of the left ventricle remodeling.

Keywords: type 1 diabetes, left ventricular remodeling, children

P-146

The influence of steroid treatment on lipid profile in pediatric patients with duchenne muscular dystrophy

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Background and Aim: Duchenne muscular dystrophy (DMD) is a rare X-linked recessive disorder. Progressive muscular weakness leads to loss of ambulation, cardiomyopathy and respiratory failure. Steroids are commonly used to slow down the disease progression. Additionally sedimentary lifestyle and inappropriate diet lead to higher risk of developing metabolic complications, such as dyslipidemia further incising cardiac risk, but this has not been sufficiently studied so far in this disease.

Methods: A prospective, observational, longitudinal study approved by the local ethics committee was conducted. The inclusion criteria were the genetically confirmed diagnosis of DMD with typical clinical presentation and age of five at least. Ninety-three patients and 312 observations were included. Each observation was considered a separate datapoint. Wizard 2.0.10 was used for statistical analysis using non-parametric tests appropriately for data distribution. Results are given as median (range) in mg/dl or n (percentage).

Results: In the majority of instances 230 (63.0%) patients were treated with steroids: 111 (30.4%) with prednisone and 119 (32.6%) with deflazacort. Significant differences were found in the lipid profiles dependently on lacking or having steroid treatment: triglycerides (TG, mg/dl) 75.00 (40.00 – 204.00) vs 98.00 (32.00 – 628.00), ($p<0.001$) and total cholesterol (TC) 155.00 (99.00 – 218.00) vs 169.00 (99.00 – 279.00), ($p<0.001$) but not for HDL-C 45.00 (27.00 – 66.00) vs 48.00 (26.00 – 91.00), ($p = 0.082$) and LDL-C 93.00 (34.00 – 174.00) vs 100.00 (33.00 – 205.00) ($p = 0.102$). We have also found significantly greater elevation of TG 108.00 (54.00 – 269.00) vs 93.00 (32.00 – 628.00) ($p = 0.021$) and TC 174.00 (114.00 – 279.00) vs 163.00 (99.00 – 248.00) ($p = 0.035$) but not for the HDL-C and LDL-C ($p = 0.808$ and $p = 0.112$ respectively) comparing deflazacort and prednisone. This meant higher prevalence of elevated TG if taking steroids: 2 (3%) vs 33 (21%), $p<0.001$ but the prevalence of abnormalities of the remaining parameters (5 (7%) vs 27 (17%), $p = 0.061$, 2 (3%) vs 12 (7%) $p = 0.187$ and 15 (22%) vs 28 (18%), 0.394 respectively) was not significantly different, possibly due to small numbers.

Conclusion: Steroid treatment leads to high prevalence of lipid profile abnormalities in young DMD patients, and the clinical consequences of this phenomenon need further investigation.

Keywords: steroids, lipid profile, Duchenne muscular dystrophy, DMD, prednisone, deflazacort

P-148 / Moderated Poster

Different profiles of lipoprotein particles associate various degree of cardiac involvement in adolescents with morbid obesity

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Background and Aim: Dyslipidemia secondary to obesity is a risk factor related to cardiovascular disease events, however a pathological conventional lipid profile (CLP) is infrequently found in obese children. The objective of our study is to evaluate the lipoprotein subclass profile (LSP) and its relationship with cardiac changes, metabolic syndrome and inflammatory markers in a population of morbidly obese adolescents with normal CLP and without type 2 diabetes mellitus, the most common scenario in obese adolescents.

Methods: Prospective case-control research of 42 morbidly obese adolescents and 25 normal-weight adolescents, whose left ventricle (LV) morphology and function had been assessed. The LSP was obtained by proton nuclear magnetic resonance spectrometry, and the results were compared according to the degree of cardiac involvement (normal heart, mild LV changes and severe LV changes, specifically LV remodeling and systolic dysfunction) and related to the highly-sensitive C-reactive protein (hs-CPR) and the homeostatic model assessment for insulin-resistance (HOMA-IR) as markers of inflammation and insulin-resistance.

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A second analysis was performed to compare our results with the predominant LSP when only body mass index and metabolic syndrome criteria were considered.
Results: The three cardiac involvement groups showed significant differences in HOMA-IR and hs-CPR ($p<0.01$). A significant increase was also observed in the ratio Total-P/HDL-P (1.41 vs 1.44 vs 1.49, p0.01) and the ratio LDL-P/HDL-P (40.0 vs 43.9 vs 47.1, p0.01), while when mild and severe LV change groups were compared, a not statistically significant elevation in the small LDL-P (565.0 vs 625.1 nmol/L, p0.07) was identified. The significant independent variables associated with severe LV changes were small LDL-P (OR 1.0 [1.0-1.0], $p<0.05$) and HOMA-IR (OR 1.3 [1.0-1.6], $p<0.01$). In the second analysis, an atherosclerotic LSP was detected in morbidly obese subjects, characterized by a significant increase in large VLDL-P, small LDL-P, ratio Total-P/HDL-P, ratio LDL-P/HDL-P and triglyceride-enriched HDL-P ratio. Subjects that meet criteria for metabolic syndrome presented overall worse LSP and remnant cholesterol values.
Conclusion: The overage of VLDL-P and LDL-P compared to HDL-P and, particularly, the increase in small LDL-P, appear to be key LSP's parameters involved in LV changes. Morbidly obese adolescents without pathological CLP show an atherosclerotic LSP.

Keywords: morbid obesity, adolescents, lipoprotein particles, cardiac changes, systolic dysfunction, metabolic syndrome

P-149

What about the lungs? body plethysmography in patients with congenital heart disease

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Background and Aim: Lung volumes are usually examined as forced vital capacity (FVC) and forced expiratory volume in one second (FEV1). However, these values can only give the first hint on patients' lung volumes. With body plethysmography, additional volumes (total lung capacity, TLC or residual volume, RV) can be measured. While this test is commonly performed in patients with lung diseases, it is scarcely used in patients with congenital heart disease (CHD). This study aims to evaluate extended lung volumes in children, adolescents, and adults with different CHD.
Methods: A total sample of 215 children, adolescents, and adults (26 ± 12 years, 93 females) underwent spirometry (FVC, FEV1 and its ratio) along with body plethysmography (TLC). Data were compared with the current reference values from the Global Lung Initiative 2012 for FVC and FVC/FEV1 and Stocks et al., 1995 for TLC. Z-scores with < -1.64 in FVC are described as "restrictive pattern" and in FEV1/FVC as obstructive. In a subsequent step, TLC was added to differentiate true restriction from obstruction with hyperinflation (restrictive pattern but normal TLC).
Results: Overall ($n = 215$), mean z-FVC was -1.11 ± 1.26; FEV1/FVC was -0.10 ± 0.99, TLC -0.72 ± 1.79 and RV/TLC 0.53 ± 1.17, representing a normal lung function in 63%. CHDs were distributed as follows (% with abnormal results): 16 with left heart obstruction (6%), 114 with right heart obstruction (45%), 38 with transposition of the great arteries (26%), 19 with an isolated shunt (33%), 13 with a Fontan palliation (23%), two cyanotic (none) and 13 miscellaneous CHDs (54%). Conspicuous results were: 15% with an obstructive lung function, 21% with a restrictive lung function, and 1% with a mixed pattern. In 8% of the patients, the obstruction could only be detected owing to body plethysmography.
Conclusion: Abnormal lung volumes are seen in many patients with CHD. Body plethysmography improves the diagnosis and unveils findings that need to be treated (e.g., obstruction). We recommend that further clarification with body plethysmography should be performed, whenever a decreased FVC with normal FEV1/FVC in spirometry is seen.

Keywords: Lung function, Lung volume, CHD

P-150 / Moderated Poster

Motor function in children and adolescents with CHD in a direct twin comparison – same same, but different?

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Background and Aim: Newborns and infants with congenital heart defect (CHD) often show delayed neurodevelopment. It seems obvious that the operations and treatments of the heart defect, especially on motor function, are responsible for this delay. It is unclear how they would have developed without the CHD; Therefore, we compared motor function of twin siblings with and without CHD.
Methods: Motor function was measured as part of the nationwide ongoing twin study "SameSame", investigating neurodevelopment (cognitive function, motor function, mental health and health-related quality of life). 49 pairs of twins (one with CHD and the other without, 51 girls (52.0%), 8.99 ± 4.3 years) have completed the Movement Assessment Battery for Children-II so far. The different age groups (A: 3–6 years, $n = 42$; B: 7–10 years, $n = 26$ and C: 11–16 years, $n = 30$) consist of eight age-specific tasks covering the areas of manual dexterity, ball skills, static and dynamic balance as well as a total motor score. The data of the twin siblings among each other were compared using the student t-test for paired samples.
Results: Children with CHD (6.1% simple/42.9% moderate/51% complex CHD severity) showed significantly reduced scores in manual dexterity with a mean difference (MD) of 3.2 points ($p = 0.010$) and total motor score (MD 7.6 points, $p = 0.002$) in the direct comparison with their healthy twin. Regarding the different age groups, the youngest group A there was no difference between the twins in all areas, group B a significant difference in manual dexterity (MD 6.2 points, $p = 0.020$) and group C in manual dexterity (MD 4.3 points, $p = 0.030$), ball skills (MD 4.2 points, $p = 0.026$), static and dynamic balance (MD 3.6 points, $p = 0.047$) as well as a total motor score (MD 12.1 points, $p = 0.002$) in the burden on children with CHD.
Conclusion: Children and especially adolescents with CHD show deficits in motor function compared to their healthy twin siblings. But not the youngest, by contrast, adolescents showed clear deficits in all motor function areas. This in turn demonstrates that patients with CHD need lifelong support and assistance to avoid or compensate for such performance deficits as well as to be supported to lead an active life.

Keywords: neurodevelopment CHD motor function twins siblings

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P-151

Recovery kinetics of gas exchange parameters and heart rate after maximal exercise in children with repaired coarctatio aortae compared to controls

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Background and Aim: Children after succesful repair of Coarctatio Aortae (rCoA) still demonstrate a decreased exercise capacity. Although the physiological processes during exercise are thoroughly investigated, little research exists on the recovery after exercise. In this study we evaluate the recovery kinetics of VO_2, VCO_2 and HR after maximal exercise in children with rCoA compared to healthy controls.

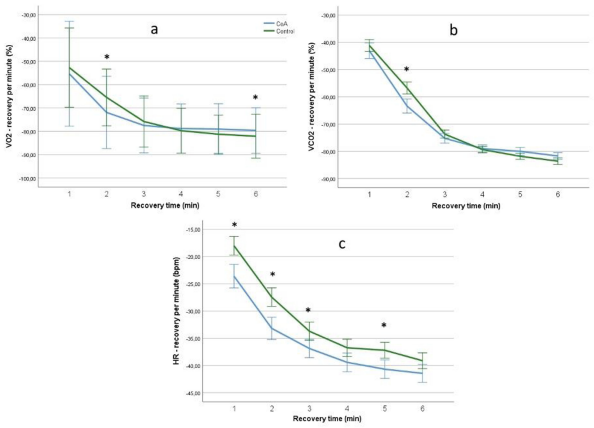
Methods: 65 children after rCoA and 65 matched controls performed a maximal cardiopulmonary exercise test. In the subsequent 6 minute recovery period, kinetics of VO_2, VCO_2 and HR were analysed. The half-life time (T½) of the exponential decay and the patterns of percentages drop per minute were compared between groups.

Results: Exercise performance was reduced in the rCoA group with lower VO_2peak (40.0 ± 7.7 vs. 41.1 ± 8.8 ml/min/kg; p = 0.012) and lower percentages of predicted value of VO_2peak (89.0 ± 18.4 vs. 100.3 ± 13.7 %) and maximal load (79.8 ± 19.9 vs. 93.0 ± 18.0 %). Maximal HR (183 ± 15 vs 190 ± 12 bpm; p = 0.003) was also lower in the rCoA patients. The recovery kinetics of VO_2 and VCO_2 was faster in rCoA patients with lower T½ values compared to controls (T½VO_2: 38.1 ± 11.8 vs. 44.9 ± 9.4 s; p<0.001 and T½VCO_2: 55.5 ± 15.2 vs. 61.8 ± 10.9 s; p<0.001). Examining the patterns of percentages drop revealed a significant interaction (p<0.001) between group and time, indicating that the presence of rCoA altered the recovery course. T½HR was lower in the rCoA group (54.1 ± 21.4 vs. 68.3 ± 20.7 s), demonstrating faster HR recovery. The values of percentages drop were also higher in the rCoA group. Looking at the recovery pattern, a significant effect of group as well as interaction of group and time (p<0.001) was found, confirming the faster recovery in the rCoA patients.

Conclusion: Despite a lower exercise tolerance, rCoA patients exhibit faster recovery kinetics of VO_2, VCO_2 and HR after maximal exercise. An altered oxygen supply-demand balance might induce a higher local muscle metabolism leading to faster recovery. Further research will need to clarify the underpinning mechanisms leading to faster recovery in rCoA patients.

Keywords: Coarctatio Aortae, maximal exercise, CPET, recovery

Recovery of VO2, VCO2 and HR after maximal exercise in children with repaired Coarctio Aortae and healthy controls.



Recovery of VO_2 (a), VCO_2 (b) and HR (c) after maximal exercise in children with repaired Coarctio Aortae (blue) and healthy controls (green).
VO_2 and VCO_2 are expressed as percentage drop from peak value and HR is expressed in absolute drop in bpm from maximum HR.
Error bars indicate mean ± 1.96SD. * indicates statistically significant difference at p < 0.05.

P-152

Evaluation of motor competence, physical activity and motivation in children and adolescents with a univentricular heart

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Background and Aim: Physical activity (PA) and fitness are important predictors of health in later life. To achieve a satisfactory level of activity and fitness, one is dependent on adequate motor competence (MC), i.e., the degree of proficiency in performing a wide array of motor skills as well as the underlying processes such as coordination, control, and quality of movement. Development of MC requires dedicated practice and active exploration, preferably in organized sports (or PE) settings. Given their medical history and possible risk of overprotection by the environment, individuals with a univentricular heart (UVH) may be at risk of reduced MC. The current study set out to examine MC, PA and level of intrinsic motivation towards sports in a sample of children and adolescents with a UVH.

Methods: Twenty participants with UVH (mean age: 10.6, range: 4-17), monitored by the Pediatric Cardiology unit of the Ghent University Hospital, were assessed with the Movement Assessment Battery for Children 2 (MABC2) and surveyed with the Flemish Physical Activity Questionnaire (FPAQ) and an age-appropriate version of the Behavioral Regulation in Exercise Questionnaire (BREQ). The results of the MABC2 were compared against the published reference values, those of the FPAQ and BREQ with an age-matched control sample of 104 children from our own database.

Results: The average MABC2 score of the participants with UVH was at the 23rd percentile (range: 0.1-75). In 13 of them (60%), motor competence was "at risk of a motor problem".

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Furthermore, the degree of PA (32 minutes/day, range: 5-70) was lower than the control group (63 minutes/day, range: 11-195; $t(120) = 3.604$, $p<0.001$). Only 2 participants with UVH (10%) met the WHO guideline of 60 minutes PA per day compared with 50% of the control group. Finally, intrinsic motivation towards sport was relatively high in the UVH group (3.75 on a 5-point-scale, range: 2-5), however lower than in the control group (4.20, range: 0-5, $t(119) = 2.074$, $p = 0.04$).
Conclusion: In view of the role of motor competence in ensuring a healthy lifestyle, actions are recommended to promote motor development and physical activity in children and adolescents with UVH.

Keywords: Univentricular heart, motor competence, physical activity, motivation

P-153
Psychomotor developmental characteristics of children with unrepaired congenital heart defects

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Background and Aim: Impairment of psychomotor development in patients with congenital heart defects (CHD) occurs frequently, both in cyanotic and non-cyanotic CHD (adequate cerebral tissue oxygenation). Literature considers as main extrinsic factors, traumatic events like heart surgery, especially extracorporeal circulation, and events during intensive care unit stay. The aim of this study was to evaluate children before neurological traumatic events took place in order to recommend preventive risk reduction strategies for further management of the patient.
Methods: The study included 28 children with unrepaired CHD evaluated with Denver Developmental Screening Test II (125 items covering main psychomotor developmental domains: personal-social skills, fine motor-adaptative skills, language skills, gross-motor skills) in order to determine and quantify deficits. Neuromarkers, neuron specific enolase (NSE) and protein S100 (pS100) were determined and correlated with psychomotor developmental scores. Statistical level of significance was considered $p<=0.05$, Pearson or Spearman correlation coefficient was calculated and data normality was assessed with Shapiro-Wilk test using Stata-version 13.
Results: Different levels of psychomotor development were assessed: all passed through level (the highest item below which the child did not fail any items), baseline level of competence (the highest of three or more consecutive items passed), highest item passed before consistent failure (until three items had been failed consecutively) and the upper limit of children's development (highest item passed beyond three consecutive items failed). Patients with CHD had more delays than normal children in a percent varying from 27.2-72.7 (table 1). Based on these, domain-specific and overall developmental functioning estimates were calculated (delayed in 27.2%) and a developmental quotient score was derived (lower than in normal children in 27.7%).
NSE was over the upper limit in 89.8% of cases and pS100 in 54.5% of cases. Correlation between psychomotor developmental delay and neuromarkers were found only with pS100 and included the following: personal social domain-all passed through level ($r = -0.69$,$p = 0.01$), personal social domain-baseline level of competence ($r = -0.68$,$p = 0.01$) and language domain-highest item passed before consistent failure ($r = 0.6$,$p = 0.04$).
Conclusion: Psychomotor developmental delay was found in majority of cases regardless pathophysiology: cyanotic or non-cyanotic CHD. The delay was corelated with higher than normal plasma levels of S100p.

Keywords: psychomotor development, congenital heart defect, neuromarkers, neuron specific enolase, protein S100

Table 1: percent of patients with developmental delay in each estimated level and psychomotor domain

Level / Domain	Personal/Social	Fine Motor	Language	Gross Motor
All passed through level	63.6%	36.3%	36.3%	54.5%
Baseline level of competence	72.7%	36.3%	36.3%	54.5%
Highest item passed before consistent failure	27.2%	27.2%	27.2%	45.4%
The upper limits of children's developmental	27.2%	27.2%	27.2%	4.5%
Developmental Functioning Estimates	45.4%	36.3%	27.2%	54.5%
Developmental Quotient Scores	45.4%	36.3%	27.2%	54.5%

P-154
Digital health nudging to increase physical activity in adolescents with congenital heart disease - preliminary results of a RCT

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Background and Aim: Digital health nudging has been shown to be an effective, age-appropriate, and modern e-health intervention to increase physical activity (PA) in healthy adolescents. However, such studies are lacking in adolescents with congenital heart disease (CHD), although activity promotion is particularly important in this vulnerable population. This study examines if digital health nudging via daily smartphone messages can increase PA in adolescents with CHD in a randomized-controlled trail (Clinical Trials Identifier: NCT04933786).
Methods: From June to November 2021, 41 patients (14.73 ± 2.13 years, 19 girls) with moderate or severe CHD were randomly allocated to an intervention group (IG; $n = 22$) or control group (CG; $n = 19$). Daily PA, measured in minutes of moderate-to-vigorous PA (MVPA) and daily step count, was objectively assessed by the wrist worn wearable "Garmin Vivofit jr.®". After baseline PA assessment for seven days, PA was assessed over the first four weeks of the overall planned 3-month trail period. The intervention group received daily, short and informative smartphone messages based on Bandura's social cognitive theory on the subject of PA, thus nudging them to be more active in their everyday lives over a period of four weeks. The control group only wore the wearable. Univariate analysis of covariance (ANCOVA) was conducted to determine the differences between IC and CG controlling for baseline PA.

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Results: There was no significant effect of the nudging intervention on MVPA (p = 0.117) and daily step count (p = 0.310) controlling for baseline PA. While the IG reach an average of 85.91 ± 30.03 minutes MVPA per day, the CG accumulated 90.28 ± 24.70 minutes MVPA per day. The IG conducted an average of 9,213 ± 3,730 daily steps, whilst the CG conducted 9,053 ± 3,245 daily steps. Baseline PA values did not significantly differ between IC and CG (MVPA: p = 0.208, steps: p = 0.204).
Conclusion: Preliminary results of the first four weeks of digital health nudging showed no increase of daily physical activity among adolescents with congenital heart disease. However, effects of a longer intervention period remain unclear. Since behavior change requires a certain amount of time, effects of the intervention can still be expected.

Keywords: e-health, digital health, adolescents, congenital heart disease, physical activity

P-155/Moderated Poster
Association between objectively measured physical activity and exercise capacity in children with congenital heart disease

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Background and Aim: Wearables are commonly used in medical and preventive fields. In this context, physical activity (PA) has been shown to correlate with exercise capacity in adults with congenital heart disease (CHD). However, both are independent domains and their relationship remains unclear in children with CHD. Therefore, this study investigated the relationship between objectively measured PA and exercise capacity (peakV'O2) in children and adolescents with CHD.
Methods: From September 2017 to September 2021, sixty-nine children (13.0 ± 2.9 years, 22 girls) with various CHD were included. Moderate-to-vigorous physical activity (MVPA) and daily steps were assessed with the "Garmin vivofit jr.®" for seven consecutive days. Participants also performed a cardiopulmonary exercise test (CPET) to quantify their peakV'O2. CHD severity according to ACC criteria was complex in 48%, moderate in 41% and simple in 11% of study participants. To examine the association between PA and exercise capacity, linear regression analysis was performed adjusting for age, sex, height, and weight.
Results: There was no association between MVPA and peakV'O2 (p = 0.293) as well as between steps and peakV'O2 (p = 0.242) in children with CHD after adjusting for anthropometrics. Overall, CHD children were remarkably active with a weekly average of 82.4 ± 36.8 minutes of MVPA per day and an average of 9,528 ± 3,939 steps per day. In total, 74% reached the World Health Organization recommendation of at least an average of 60 min MVPA per day. PeakV'O2 was 33.6 ± 7.7 mL/min/kg, representing a slightly diminished exercise capacity of 78.9 ± 17.5% of the age- and sex-related reference values.
Conclusion: There was no association between objectively measured PA and exercise capacity in children and adolescents with CHD. It is therefore compelling to assess both domains in the clinical setting - PA in the context of cardiovascular prevention and peakV'O2 as the strongest predictor for morbidity and mortality.

Keywords: physical activity, exercise capacity, wearables, congenital heart disease, children

P-156/Moderated Poster
Effect of physical activity on cardiovascular health in six-year-old children born to mothers with high cardiometabolic risk

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Background and Aim: Physical activity decreases cardiovascular (CV) risk and triggers heart remodelling in adulthood. We aimed to assess associations between objectively measured physical activity and CV health in early childhood.
Methods: Longitudinal observational follow-up study of 160 children (boys, N = 86) originating from Finnish Gestational Diabetes Prevention Study (RADIEL). Children born to mothers with increased cardiometabolic risk (BMI ≥ 30kg/m2, N = 93; gestational diabetes, N = 73) were examined at 6.05 ± 0.5 years (mean ± SD) with anthropometrics, body composition, blood pressure (BP), fasting glycaemia and lipids, energy intake, physical activity with accelerometery, pulse wave velocity (PVW), heart and vascular ultrasound.
Results: BMI Z-score, systolic and diastolic BP Z-scores were elevated in comparison to reference population (mean ± SD; 0.45 ± 0.92, 0.42 ± 0.70, and 0.45 ± 0.64, respectively). Boys had longer moderate-to-vigorous physical activity (MVPA) duration than girls (mean ± SD, 81 ± 22 min/day vs. 66 ± 19 min/day, P< 0.001), and lower body fatness (mean ± SD, 14.9 ± 4.8 % vs. 18.7 ± 5.0 %, P<0.0001). MVPA was weakly negatively associated with child body fatness, but did not significantly correlate with fasting glucose or lipids (Table 1). MVPA was inversely associated with diastolic blood pressure Z-score in boys (r = -0.26, P = 0.02). In multivariable linear regression models, MVPA was weakly, but independently associated with left ventricular mass and volumes, and resting heart rate (Table 1). Child carotid artery lumen diameter, intima-media thickness, distensibility coefficient, and central and peripheral PVW were not associated with physical activity.
Conclusion: In high-risk population of 6-year-old children, physical activity levels are higher among boys. At this stage, physical activity has a weak negative association with child adiposity and blood pressure. A physical activity-related increase in left ventricular mass and volumes, and a decrease in resting heart rate could represent a physiological adaptation during early childhood.

Keywords: cardiovascular risk, physical activity, obesity, child, echocardiography, ultrasonography

Table 1. Multivariable linear regression models assessing associations between physical activity and child cardiovascular health in early childhood.

	Model adjusted R^2	Predictors	Beta	P-value
Body fat [%]	0.137	MVPA [min/day]	-0.046	0.026
		Sex [male=1, female=0]	3.105	<0.001
		Energy intake [kcal]	0.001	0.297
Left ventricular mass [g]	0.390	MVPA [min/day]	0.086	0.011
		Lean body mass [kg]	2.924	<0.001
Left ventricular end-diastolic volume [ml]	0.368	MVPA [min/day]	0.073	0.030
		Lean body mass [kg]	2.790	<0.001
Left ventricular end-systolic volume [ml]	0.234	MVPA [min/day]	0.033	0.029
		Lean body mass [kg]	0.894	<0.001
Left atrial volume [ml]	0.298	MVPA [min/day]	0.032	0.070
		Lean body mass [kg]	1.091	<0.001
		Body fat [%]	0.292	0.005
Resting heart rate [beat/min]	0.120	MVPA [min/day]	-0.085	0.028
		Lean body mass [kg]	-0.351	0.373
		Age [y]	-5.815	0.003
		Sex [male=1, female=0]	1.167	0.497

MVPA - moderate-to-vigorous physical activity

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Young adults conceived through ivf show significantly increased systolic blood pressure but normal endothelial function and intima media thickness

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Background and Aim: Approximately 8 million children have been conceived through assisted reproductive technologies since their development in 1978. Recent studies showed that children and adolescents conceived through in vitro fertilization (IVF) demonstrate multiple cardiovascular alterations including endothelial dysfunction and increased arterial stiffness. To the best of our knowledge, limited data on vascular function in IVF adults is available. This study aimed to investigate whether IVF adults display vascular changes compared to naturally conceived peers.
Methods: In cooperation with the fertility center of our university hospital, IVF adults were invited for the present study. Naturally conceived peers were acquired by public calls. Blood pressure was measured in a reclined position after a ten-minute resting period with an oscillometric device. The average of three consecutive blood pressure measurements was calculated and analyzed. To assess endothelial function the reactive hyperemia index (RHI) was recorded by the EndoPAT®2000 device. Right carotid intima-media thickness (cIMT) was measured sonographically. The independent-samples t-test was utilized to compare continuous variables with normal distribution between both groups. Pearson correlation was applied for normally distributed variables.
Results: In total, 8 adult IVF (6 female, 2 male) and 15 naturally conceived subjects (10 female, 5 male) were included. No significant difference in age between the IVF and the control group was detected (20.88 ± 2.48 years vs. 21.93 ± 1.91 years; p-value = 0.266). IVF subjects demonstrated, compared to peers, a significantly increased systolic blood pressure (SBP) (130.83 ± 10.96 mmHg vs. 119.29 ± 7.59 mmHg; p-value = 0.007). RHI (2.07 ± 0.54 vs. 1.95 ± 0.51; p-value = 0.631) and cIMT (0.45 ± 0.03 mm vs. 0.46 ± 0.05, p-value = 0.493) did not significantly differ between the IVF and the control group. SBP correlated significantly with right cIMT (r = 0.840, p = 0.009) within the IVF cohort.
Conclusion: In this study, IVF adults demonstrated a significantly increased mean SBP, compared to naturally conceived peers, which in turn correlated significantly with an elevated cIMT. However, endothelial function did not show significant differences between both groups. The preliminary results of the study indicate that IVF individuals with increased blood pressure are at greater cardiovascular risk. Further studies are required to evaluate potential risk factors inducing vascular dysfunction in adults conceived through IVF.

Keywords: In Vitro Fertilization, Vascular Function, Assisted Reproductive Technologies

P-158

Impact of a program on sports participation for paediatric patients with hypertrophic cardiomyopathy

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Background and Aim: Hypertrophic cardiomyopathy (HCM) is one of the most common underlying causes of sudden cardiac death (SCD) in young athletes. High-intensity exercise is considered a trigger of major events in HCM, but light/moderate-intensity exercise may have benefits. Classically, international guidelines have recommended avoiding competitive sports in patients with HCM. However, sports recommendations should be established based on individual risk. The aim of this study is to describe a program on assessment and recommendations for sports participation in young patients with HCM and related gene mutation carriers.
Methods: 0-20 years-old patients with HCM and phenotype-negative mutation carriers accepted to be included. All of them were followed-up in a national reference centre for inherited cardiovascular diseases and underwent a sports program across the usual medical appointments during the years 2019-2020. Every patient was questioned about their physical activity and received sports recommendations based on the individualized estimated risk. Data from exercise stress test were collected when available.
Results: 85 patients with HCM and 71 healthy mutation carriers were included. Differences between groups were significant ($p < 0.05$). 6 (7%) patients with HCM did not practice any exercise, 46 (54%) practiced physical activity occasionally, 27 (32%) recreational sports, and 6 (7%) competitive sports. 38 (45%) did not received restrictions, 43 (51%) were recommended to avoid high-intensity sports, and 4 (4%) were totally restricted. 54 patients (64%) underwent a cardiopulmonary exercise test: 25 (47%) were normal, 7 (13%) presented a reduced functional capacity, 5 (9%) abnormal blood pressure response, and 8 (15%) reduced peak oxygen uptake. 8 (9%) HCM patients were considered at intermediate or high-risk of SCD: 4 (50%) presented an abnormal stress test. As for the phenotype-negative mutation carriers, 21 (30%) practiced physical activity occasionally, 36 (51%) recreational sports, and

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14 (19%) competitive sports. None got exercise restrictions. 14 (20%) underwent stress testing, normal in all cases.
Conclusion: Recommendations for sports participation in children and young patients with HCM may have a significant social impact and should be individualized based on risk stratification. Results of stress tests may add a prognostic value in some cases.
Exercise assessment should be included and generalized in HCM-related SCD prevention programs.

Keywords: Hypertrophic Cardiomyopathy, Sports Participation, Risk Stratification, SCD, Prevention.

P-159/Moderated Poster
COVID-19 vaccination from 6 to 17 years carries a similar risk for fatal or life-threatening myocarditis than being struck by lightning

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Background and Aim: There are some concerns about the cost/benefit ratio of Covid-19 vaccination in the pediatric age due to the greater response to immune stimulation in children and young adolescents. Myocarditis is a feared adverse event (AE).
Methods: On January 4, 2022 we searched the Pubmed and EMBASE datasets for population studies assessing myocarditis events in pediatric subjects, and the US nationwide data about COVID-19 vaccinations and AEs.
Results: The studies selected (n = 10) were highly heterogeneous, and did not include subjects <12 years. In a nationwide study the incidence of myocarditis was 1.34 over 100,000 in males and 7 over 1,000,000 in females aged 16-19 years after the first dose, and 1.51 over 10,000 in males and 4.6 over 1,000,000 in females after the second dose. All studies with available data reported a higher risk in males and after the second dose. Based on US data, the estimated incidence of myocarditis was 2.80 over 100,000 in fully vaccinated individuals (i.e., receiving two doses of mRNA vaccines or received one dose of a single-dose vaccine) aged 6-17 years. No events were reported in subjects <5 years (n = 9,985). The incidence of myocarditis was 1.05 over 100,000 in those aged 18-64 years, and 2.75 over 1,000,000 ≥65 years. The incidence of fatal or life-threatening myocarditis was 3.78 over 1,000,000 6-17 years, 1.78 over 1,000,000 18-64 years, and 8.54 over 10,000,000 ≥65 years, with the same order of magnitude than the odds of being struck by lightning (around 2 over 1,000,000). The incidence of COVID-19 myocarditis is 1.33 in 1000 <16 years, i.e., 47.5-fold higher than fully vaccinated individuals.
Conclusion: The reported incidence of myocarditis in subjects aged ≥12 years is slightly higher in males and after the second dose, but extremely low. No case of myocarditis <5 years were reported in the US. In fully vaccinated individuals aged 6-17 years, the estimated incidence of myocarditis was 2.80 over 100,000, and of fatal or life-threatening myocarditis 3.78 over 1,000,000, with the same order of magnitude than being struck by lightning, and far lower than the risk of COVID-19 myocarditis.

Keywords: COVID-19; vaccine; vaccination; myocarditis; children; adolescent; pediatric; risk; incidence; adverse event; population.

P-160
Impact of wearing a FFP2 mask on cardiopulmonary fitness in school-aged children

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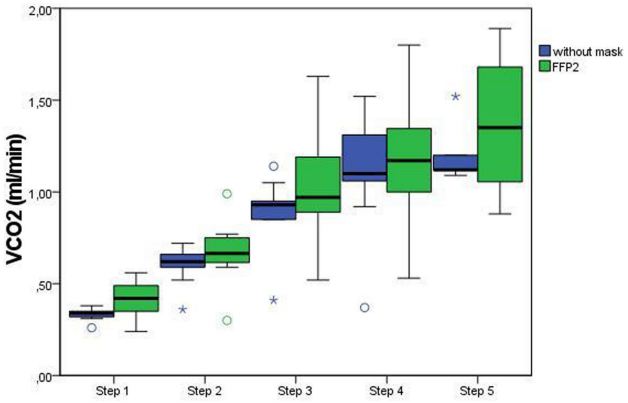
Background and Aim: Wearing face masks to detain the COVID 19 pandemic in schools has become an integral part of fighting the virus. The most effective mask is the FFP2 mask. There is a lot of public concern, especially regarding wearing a face mask at school and especially during school sports. It is therefore important to determine whether wearing a FFP2- mask during physical activity leads to changes measurable in cardiopulmonary exercise testing in children.
Methods: Cardiopulmonary exercise testing was performed two times by children aged 8-10 years as an incremental step test on a treadmill with and without a FFP2 within an interval of 2 weeks. A general questionnaire included medical history and sports participation since childhood.
Results: We included 10 children (mean age 8.4 ± 0.7 years, 6 males, 4 females). The mean parameters measured at peak exercise were comparable between both examinations (mean Peak VO2 = 39.3 ± 3.4 vs 45.6 ± 13.9 ml/min/kg; mean Peak HR 192/min ± 9 vs 188/min ±12, mean O2pulse 6 ±1.4 ml/min vs. 7 ± 1.8, mean VE 43.2 ±12.9 ml/min vs. 41.5 ± 12.7 ml/min). Neither did the respiratory gases (O2 and CO2) measured 1 min into each step differ significantly (s. figure). This study is currently ongoing.
Conclusion: Since there were no significant differences with respect to peak parameters as well as with respect to the respiratory parameters measured during each step, there is no indication to withhold physical activity even at peak capacity from children during a pandemic which makes wearing face masks mandatory.

Keywords: VO2 peak, cardiopulmonary exercise testing, sports, habitual exercise, FFP2 mask, covid 19 pandemic

VCO2 steps



Median, as well as interquartile range as well as Minimum and Maximum of VCO2 (l/min) per step in the examination with FFP2 mask in comparison with the examination without FFP2 mask

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The covid 19 pandemic and the challenge of growing up in children with congenital heart defects and adolescents

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Background and Aim: The WHO recommends physical activity (PA- 60 min/day) for children and adolescents, also having chronic diseases (CCD). Studies have shown that children and adolescents with congenital heart defects (CHD) exercise regularly and PA can be influenced by health programs (e.g. kidsTUMove). Children and adolescents with/without CCD, are willing to achieve the guidelines recommended by the WHO. Likewise, studies show that life quality (HrQl) in CHD has equally good values than healthy controls. However, the covid 19 pandemic has an impact on children's, adolescents and young adults lives. The aim is to summarize the impact and its measures on healthy children as well as in CHD/CDD and point out possible solutions improving the health of the affected subjects.

Methods: A systematic review regarding HrQl, mental health and PA in children with/without CHD/CDD was conducted. In addition, data was collected at different times of the pandemic using questionnaires (kindl, Kidsscreen, WHO 5) regarding HrQl in children with/ without CHD (age: 3-6, 8-16). A second survey was conducted in spring 2021 (age: 3-14) before and after a virtual sports camp (VIC). To have a glimps into adulthood HrQl data in student was evaluated in Dec 2020/Jan 2021.

Results: The review shows a decrease in PA of children in relation to the COVID-19 pandemic. Mental health (stress, anxiety and depression) was also affected as well as increasing concerns and fears. Result first survey: summer – fall 2020: Age 3-6: HrQLsum20 79% ±8.89 vs. HrQLfall20 78.98% ±11.21; $p > 0.05$; age 8-16 kidsscreen: HrQLsum20 51,63% ±7,64 vs. HrQLfall20 51,12% ±7,50. HrQl in healthy children remained constant, but HRQoLsum/fall20 in children with CHD was significant lower ($p<0.005$). HrQLspring21 after VIC increased (HrQLpre21 74.50% ± 11.34 vs HrQLpost21 80.13 %± 9.57; $p< 0.05$). Student WHO5 survey showed that 51.8% felt more burdened by second lockdown 2021. There was a significant worsening of study life balance since the onset of the pandemic ($p<0.001$).

Conclusion: It is important to provide suitable offers to increase PA, HRQoL, and mental health to minimize adverse health outcomes and prevent future health problems in adulthood in CHD/CDD.

Keywords: congenital heart disease, chronical disease, life quality, physical activity, pandemic

P-162

Neurodevelopmental disorder and congenital heart disease: initiating a systematic follow-up program

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Background and Aim: Neurodevelopmental disorders occur in 30 to 60% of patients requiring cardiac surgery for congenital heart disease. Surgery before the age of one year old is a major risk factor of neurodevelopmental disorder. In 2017, the AEPC Psychosocial Working Group produced a consensus document for a structured follow-up screening, but the specific modalities are depending on local resources and availability. The aim of our study was to report the systematic neurodevelopmental disorder follow-up recently developed in our institution.

Methods: In October 2020, our institution initiated a pediatric cardiac surgery program with a systematic neurodevelopmental follow-up program for children requiring cardiac surgery in their first year of life. This program includes: a clinical evaluation by a pediatric neurologist the day before surgery and before hospital discharge. At discharge, children are assigned to three follow up pathways, depending on risk factors for neurodevelopmental disorders (type of congenital heart disease, associated genetic syndrome, operative or postoperative complications, electroencephalogram or magnetic resonance imaging anomalies).

Results: Between October 2020 and October 2021, 34 children were included in this specific screening program. Thirteen patients were assigned to the Low-risk group pathway (named A) (general pediatrician follow-up with parental neurodevelopmental questionnaire at key ages). Nineteen children were assigned to the High-risk Group pathway (named C) (specific follow-up in medico-psycho-social centers for children with disability requiring specific treatments such as speech therapist, psychomotor therapist…). Two children were assigned to the Intermediate-risk group pathway (named B) (systematic neurologic reevaluation 3 months after surgery, before reassignment to pathway A or C). ASQ (ages and stages questionnaires) parental questionnaires for neurodevelopmental follow-up were collected at 6 months in 77% of cases and at 1 year in 83% of cases. Positive feedback was obtained from 94% of parents.

Conclusion: We started a recent neurodevelopmental disorder screening program for children requiring cardiac surgery before the age of one year old. Even though parents are often stressed when the neurological status of their child is discussed, this systematic screening for neurodevelopmental disorders has received positive feedback from parents. Improving the collection rate of ASQ questionnaires will optimize the completeness of follow-up.

Keywords: neurodevelopmental disorder, pediatric cardiac surgery, screening program, congenital heart disease, Ages & Stages Questionnaire

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The effect of ethnicity, maternal and early life anthropometry on cardiac structure and function in later childhood

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Background and Aim: The 'Developmental Origins of Adult Disease' hypothesis suggests that in utero and early life factors, including adiposity, impact on later cardiovascular health and disease risk. Ethnic background also effects cardiovascular disease (CVD) risk with adults of South Asian (SA) origin at increased risk. Adverse body fat patterns and increased truncal fat contribute to SA adult's increased risk, however less is known about the relationships of maternal and early life adiposity on later adiposity and CVD risk. The aim of this study was to investigate relationships between ethnic background and maternal and early childhood anthropometry with adiposity and cardiovascular status in mid-childhood.
Methods: The Manchester Heart and Growth Study is a mixed ethnicity mother-child cohort with pregnancy and birth data available. Anthropometric and echocardiographic measurements were made on 101 children, (56 White European and 45 South Asian) median age 9 years. Statistical analysis was carried out using SPSS to perform correlations.
Results: Maternal BMI was positively correlated with children's BMI at 9 years ($r = 0.28$, $p = 0.006$). Correlations between maternal BMI and truncal fat ($r = 0.35$, $p = 0.05$) and triceps SFT ($r = 0.3$, $p = 0.05$) in SA children were also significant. SA babies had lower birth weight, however there was no ethnic difference in BMI SDS at 9 years. SA children did however exhibit higher levels of body fat than WE children (whole body, right arm and truncal fat all $p<0.001$). SA children also exhibited greater changes in weight and height SDS but not BMI SDS from birth than WE children, indicating faster 'catch up growth'. Left ventricular (LV) indices were not different between SA and WE children, but measures of diastolic function (E/A and E'/A') were lower in SA compared to WE children. LV indices correlated positively to BMI SDS and body fat markers only in SA children
Conclusion: The findings of this study demonstrate that both ethnic background and maternal anthropometry during pregnancy impact on adiposity and cardiac structure and function in mid childhood. These findings support the developmental origins of adult disease hypothesis and are important when considering preventative public health strategies aiming to improve cardiovascular health and reduce future risk.

Keywords: cardiovascular risk, ethnicity, maternal, early life

P-165

Prevalence and outcomes of finnish MYH7 patients

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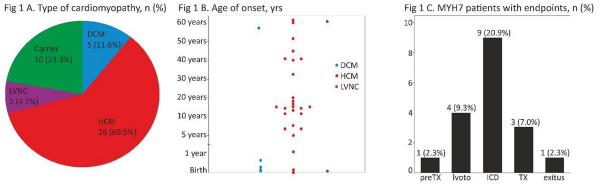
Background and Aim: Cardiomyopathies are a heterogeneous group of diseases affecting the heart muscle and are frequently genetic. The symptoms range from asymptomatic to dyspnea, arrhythmias, syncope, and sudden cardiac death. This study is particularly interested in MYH7 (beta-myosin heavy chain), as this gene is commonly mutated in cardiomyopathy patients. We analyzed the clinical presentation and natural history of patients with MYH7-related cardiomyopathy with special reference to patients under 12 years.
Methods: Our study included all identified MYH7 variants in the Helsinki University Hospital region beginning from 2002. Clinical, cardiac imaging, and echocardiography data from routine outpatient visits were collected. Genetic testing was performed as part of usual care. The primary study outcomes were all-cause mortality, cardiac transplantation, pre-transplantation examinations, LVOTO, or implantable cardiac defibrillator (ICD).
Results: We identified 43 patients with MYH7 variants, 18 of them (41.9%) index patients. The median age of disease onset was 12.0 years (15.0 years in HCM and four months in DCM). In patients with an age of onset >12 years, HCM was the most frequent diagnosis present in 17/19 (89%) patients. In patients with age of onset younger than 1-year 4/12 (33%) patients showed DCM, 2/12 (17%) showed HCM, 1/12 (8%) showed LVNC, and 5/12 (42%) patients were asymptomatic carriers. There were 18 (41.9%) primary outcome events (nine ICD implantations, five LVOTO, three cardiac transplantations, one pre-transplantation examination, and one exitus). Mean patient age at the primary outcome event was 25 years and the time from diagnosis to primary outcome event was 6.3 years. Patients who underwent ICD implantations were more likely over 12 years of age ($p = 0.045$). Otherwise, there were no statistically significant differences between primary outcomes stratified by age.
Conclusion: Our results characterize a clinically significant MYH7 cohort, emphasizing the high variability of the cardiomyopathy phenotype depending on age. We noticed that infants with MYH7 are a significant DCM phenotype-associated subgroup. We further demonstrate that patients under the age of 12 years have a similar symptom burden compared to older patients. Our study suggests that early clinical and genetic screening should be considered for younger MYH7 family members to identify patients needing closer monitoring and interventions.

Keywords: MYH7; Cardiomyopathy; Screening, age of onset

Type of cardiomyopathy, age of onset and clinical endpoints



A) Patients by cardiomyopathy (CMP) types. B) Age at diagnosis color-coded by CMP type. C) The number of patients with endpoints. PreTX = pre-transplantations examination, lvoto = left ventricular outflow tract obstruction, ICD = implantable cardiac defibrillator, TX = cardiac transplant.

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Vitamin D and calcium intake in children with fontan circulation in relationship to vitamin D status and bone mineral density

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Background and Aim: We have previously demonstrated a progressive deterioration of vitamin D status and bone mineral density (BMD) during childhood after Fontan-type palliation of congenital heart disease. Our aim was to investigate associations of vitamin D status and BMD with vitamin D and calcium intake.
Methods: From our outpatient clinic and hospital admission we recruited 55 children aged 5-18 years with Fontan circulation. For group comparisons (t-test, ANOVA), the study group was divided by age into 5-9 (group A), 10-15 (B) and 16-17 years (C). Dietary data was collected by a 256-item food frequency questionnaire, coded in the Norwegian food and nutrient calculation system KBS, and serum 25-hydroxy-vitamin D was analyzed. We assessed BMD by dual-energy X-ray absorptiometer (DXA). Reference group for dietary intake were age-matched Norwegian children from the 2015 national dietary survey UNGKOST 3.
Results: Fifty-five children were enrolled (group A n = 18, B n = 17 and C n = 20). Intakes of vitamin D and calcium in Fontan children were matching those of healthy Norwegian children. Multiple linear regression showed that 25OH-vitamin D was associated with both vitamin D intake (R = 0.34, p = 0.012) and time since Fontan operation (R = -0.36, p = 0.008) but not calcium intake. There was no relationship between DXA Z-scores and dietary intake of vitamin D or calcium.
Forty percent reported daily consumption of vitamin D supplements and in those, intakes of vitamin D were higher (18.5 vs. 9.7 µg/day, p<0.001), vitamin D status was better (77.3 vs. 52.8, p<0.001), but no difference BMD was observed. Supplements represented 38% of overall vitamin D intake.
In children with a low vs. normal BMD (DXA Z-score <-1.0 vs. ≥-1.0) intake of vitamin D was 24.5 vs. 11.8 µg/day (p = 0.07), and intake of calcium was 1334 vs. 1211 mg/day (p = 0.60).
Conclusion: Vitamin D status in children with Fontan circulation is positively associated with vitamin D intake. Children reporting daily consumption of supplements had a higher intake of vitamin D and better vitamin D status. Vitamin D supplements constitute large fractions of the total vitamin D intake. Supplements should therefore be advocated. We found no association between BMD and vitamin D or calcium intake.

Keywords: Fontan circulation, vitamin D, bone mineral density, calcium intake, nutrition, food frequency questionnaire

Result table

Results Mean ± standard deviation	Fontan					UNGKOST 3	
Group	A (n=18)	B (n=17)	C (n=20)	Total (n=55)		4th grade (n=636)	8th grade (n=687)
Vitamin D intake, µg/day (recommended: 10µ/day)	12.1± 6.2	18.8± 13.9	9.3± 6.1	13.0± 9.7		9.6± 5.4	8.8± 6.2
Calcium intake, mg/day (Recommended: 600-900 mg/day*)	1254± 430	1372± 649	1279± 685	1294± 601		1105± 326	1041± 392
Vitamin D supplement use, (%)	56	35	30	40	p=0.3[a] a; between age-groups		
Vitamin D intake µg/day, by supplement use yes / no				18.5 / 9.7	p<0.001		
Type of supplement	Cod liver oil, omega 3 capsules and multivitamin/mineral supplements						
Dietary sources of vitamin D	Supplements (38%) Fortified fats (21%) Fatty fish (20%) Milk/milk products (9%)						
Dietary sources of calcium	Milk/cream/ice cream (53%) Cheese (20%)						
Vitamin D status, 25(OH)D, nmol/L	76.3± 30.2 (n=18)	66.4± 29.5 (n=17)	47.0± 16.0 (n=20)	62.6± 28.2 (n=55)	p<0.001[b] p=0.016[c] b; group A vs. C c; group B vs. C		
	By vitamin D status (25(OH)D)						
	≥ 50 nmol/L (n=35)		< 50 nmol/L (n=19)				
Vitamin D intake, µg/day	15.0±10.3		9.8±7.7		p=0.06		
Calcium intake, mg/day	1308±611.5		1242±494.5		p=0.67		
Vitamin D status	**By supplement use**						
	Yes (n=22)		No (n=33)				
	77.32±27.7		52.76±24.3		p<0.001		
BMD status, DXA Z-score whole body	0.76± 0.8 (n=18)	-0.28± 0.9 (n=17)	-0.53± 1.2 (n=12)	0.06± 1.1 (n=46)			
	By BMD status (DXA Z-score)						
	≥ -1.0 (n=39)		< -1.0 (n=7)				
Vitamin D µg/day	11.8±8.0		24.5±15.2		p=0.07		
Calcium, mg/day	1211±551.3		1334±637.1		p=0.60		

Intakes are included vitamin D supplementation. *Calcium recommendation; 2-5 years: 600mg, 6-9 years: 700mg, 10-17 years: 900mg, >18 years: 800mg.
Dietary recommendations from "Nordic Council of Ministers. Nordic Nutrition Recommendations 2012: Integrating Nutrition and Physical Activity. Chapter 16: Vitamin D, and chapter 28: Calcium. 5 ed. Copenhagen: Nordic Council of Ministers 2014".

P-167

Carditis in acute rheumatic fever: 15-year single-center experience of a middle-income country in latin america

Diego Alejandro Lozano Espinosa[1], Kelly Christina Marquez Herrera[2], Victor Manuel Huertas Quinones[3], Adriana Soraya Diaz Maldonado[4], Roy Sanguino Lobo[5]
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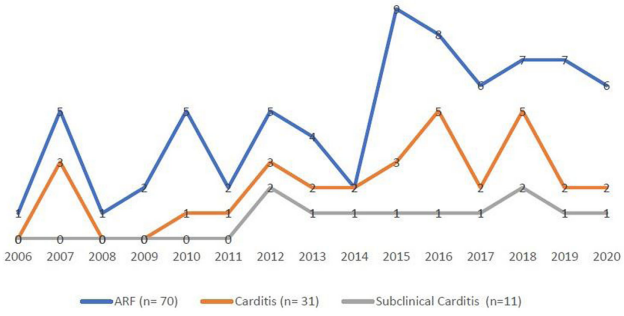
Background and Aim: Acute rheumatic fever (ARF) is the main cause of acquired heart disease in children from developing countries. Objective, to describe the clinical presentation, cardiac involvement and the frequency of carditis in children with ARF.
Methods: Retrospective study of children with ARF for 15 years (2006 to 2020) in a children's hospital in Colombia (middle-income country in Latin America).
Results: Seventy patients under the age of 18 with criteria for ARF were evaluated. The mean age was 10 years ± 2.9 years. Incidence was 30 cases of ARF per 100,000 children seen per year. There was a higher frequency of females (44/70; 63%). An average of five patients per year were diagnosed with ARF with the highest number of cases in 2015 and 2016 (nine and eight cases respectively). Chorea (73%) and arthritis (20%) were the most common reasons for hospital admission. Carditis was the second most frequent manifestation (31/70; 44%) with mild involvement more frequently (26/31; 84%). Subclinical carditis was present in 16% (11/70). Cases of ARF were proportionally more frequent from 2015 (43/70; 61% [2015-2020: six years] vs 27/70; 39% [2006-2014: nine years]), as did the frequency of carditis (19/31; 61%) and subclinical carditis (7/11; 64%). Mitral regurgitation was present in 29 of 31 patients with carditis (93%), followed by the aortic regurgitation in 17/31 (55%) and combined mitral and aortic valve involvement in 15/31 (48%). Eight percent had a PR prolongation. Twenty-eight percent had carditis and chorea (20/70) and 17% had carditis and arthritis (12/70). About cases with carditis, 48% (15/31) required anti-congestive therapy, 45% (14/31) glucocorticoids, 71% (22/31) acetylsalicylic acid and 29% (9/31) non-steroidal anti-inflammatory drugs.
Conclusion: The incidence in this study is high (moderate-risk) and greater than what has been documented in other countries in Latin America. Chorea was the most frequent initial manifestation, which reflects the late diagnosis. The case frequency of ARF, especially for carditis and subclinical carditis, increased considerably beginning in 2015, which could be related to a greater sensitivity of the criteria which were updated that year. Echocardiographic is trascendent because 35% of cases with carditis were subclinical.

Keywords: Acute Rheumatic Fever, carditis, subclinical carditis, chorea, arthritis

Figure: Distribution of ARF, carditis and subclinical carditis per year



An average of five patients per year were diagnosed with ARF with the highest number of cases in 2015 and 2016. Cases of ARF were proportionally more frequent from 2015, as did the frequency of carditis and subclinical carditis.

An average of five patients per year were diagnosed with ARF with the highest number of cases in 2015 and 2016. Cases of ARF were proportionally more frequent from 2015, as did the frequency of carditis and subclinical carditis.

P-168/Moderated Poster
Nutritional diagnosis in children with congenital heart disease and parental characteristics in a social program in a latin american country

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Background and Aim: Nutritional status is a topic of interest in children with congenital heart disease (CHD). The anthropometric characteristics of parents and family habits play an important role in the development of malnutrition. The objective is to establish the relationship between the nutritional status of children in the "Gift a Life" Social Program of the Fundación Cardioinfantil-Instituto de Cardiología and the characteristics of their parents.
Methods: Cross-sectional study with analytical component. Nutritional characteristics of children were established in relation to demographic and anthropometric characteristics of their parents (January 2019 - March 2020), using the classifications recommended by the Ministry of Health and Protection of Colombia - Resolution 00002465 of 2016.
Results: 375 children were evaluated (50.4% male, median age 10 years). In <5 years, weight-for-age (46.0%) and undernutrition (22.2%) prevailed; in children aged 5-17 years, weight-for-age (55.8%) and overweight (17.0%) were more prevalent. 106 children had a diagnosis of CHD, 60.2% non-cyanogenic. In the parental group (86.9% female, median age 41 years), there was a higher prevalence of overweight (42.7%) and obesity (36.0%). A relationship was documented between the nutritional status of parents and children aged 5-17 years ($p < 0.0001$). Weight and body mass index (BMI) of parents were associated with nutritional classification of children aged 5-17 years ($p < 0.02$). There was a weak correlation between parents' and children's BMI $R = 0.229$ ($p < 0.0001$).
Conclusion: During January 2019 and March 2020, demographic and anthropometric data were collected from 375 children and their accompanying parents, who attended the brigades of the social program "Give a Gift of Life" of the Fundación Cardioinfantil. The categorization of nutritional status in children showed a higher proportion of subjects with adequate weight in both age groups studied; in children under 5 years of age, deficit malnutrition appeared as the second most important category, while in children over 5 years of age the second highest prevalence was overweight. In accompanying adults, the most prevalent characteristic was malnutrition by weight excess.
In the study population, children's BMI correlated with that of their parents. The causal association needs to be explored by further studies.

Keywords: Nutritional diagnosis, congenital heart disease, social program.

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P-169/Moderated Poster

Adverse arterioventricular interaction in adolescents with chronic disease

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Background and Aim: Adolescents with chronic disease are frequently exposed to inflammatory, metabolic and hemodynamic risk factors, which can induce arterial changes. Indeed, the recent 'Cardiovascular Disease in Adolescents with Chronic Disease' (CDACD) study showed enhanced aortic pulse wave velocity (PWV), which reflects aortic stiffness, in adolescents with various chronic disorders. Enhanced aortic stiffness can increase left ventricular (LV) afterload and trigger a cascade of adverse arterioventricular interaction. This study investigates the relation between early aortic changes and cardiac function in the CDACD study participants.

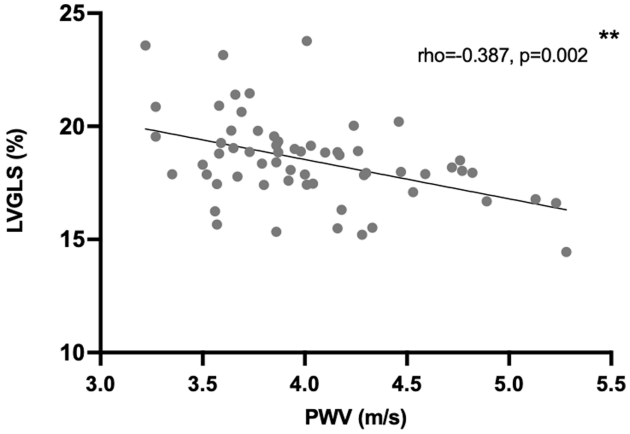
Methods: This cross-sectional single center study enrolled adolescents 12-18 years old with cystic fibrosis (CF, n = 24), corrected coarctation of the aorta (CoA, n = 25), juvenile idiopathic arthritis (JIA, n = 20), obesity (OB, n = 20), and healthy controls (n = 25) were studied with echocardiography and magnetic resonance imaging (MRI). Conventional markers of cardiac function and LV global longitudinal strain (LVGLS), which is an established (pre)clinical marker of LV dysfunction (LVD), were studied with echocardiography. Aortic PWV and aortic wall thickness were assessed with 3.0T MRI.

Results: Echocardiography showed normal global LV dimensions and preserved systolic and diastolic function in all adolescent disease groups, except for a decreased LVGLS in CoA, JIA, and obese adolescent compared to controls ($p<0.05$). Using multivariable regression analysis, decreased LVGLS was predicted by increased aortic PWV (standardized β -0.47, $p<0.001$).

Conclusion: The decreased LVGLS in several adolescents chronic disease groups reflects preclinical LVD. The association with increased aortic stiffness indicates adverse arterioventricular interaction. This impact of chronic disease on the cardiovascular system substantiates the emerging call for cardiovascular follow-up of adolescents with chronic disease, in order to improve their cardiovascular outcome.

Keywords: adolescents, chronic disease, PWV, GLS, adverse arterioventricular interaction

Figure - Correlation LVGLS and aortic PWV



Univariate correlation analysis between left ventricular global longitudinal strain (LVGLS) and aortic pulse wave velocity (PWV) showed a negative correlation with LVGLS (Spearman's rho = -0.387, p = 0.002) (n = 61), **p<0.01.

Interventional Cardiology

P-170

Percutaneous closure of subpulmonic ventricular septal defect with lifetech konar -multifunctional occluder

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Background and Aim: Ventricular septal defects (VSDs) comprise the most common congenital heart defect at birth. Subpulmonic location is an unlikely site for the VSD to close spontaneously and known to have complications such as aortic valve prolapse and regurgitation. Herein, we present a case of percutaneous device closure of a subpulmonic VSD using Multifunctional Occluder (MFO) device.

Methods: A 4-year-old girl was referred for further cardiac evaluation as she was found to have a pansystolic murmur and complaint of tired quickly. She weighed 14 kg. On cardiac examination, it was heard grade 4/6 pansystolic murmur best in the left lower sternal border. Electrocardiogram showed right ventricular dominance, with no strain. Echocardiogram showed a 4.2 mm subpulmonic VSD with left-to-right shunt, mild enlargement of the left heart. There were no pulmonary arterial hypertension, aortic valve prolapse or aortic regurgitation.

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After general anesthesia, sheats were inserted into the right femoral vein and artery using percutaneous technique. Left ventricle angiogram in the right anterior oblique (30°), cranial (20°) and lateral views showed 4,3 mm subpulmonic VSD. 8x6 mm MFO was deployed across the defect (Figure 1). Transthoracic echocardiogram and LV angiogram after closure of the VSD showed no LVOT protrusion and residual shunt.
Results: Although the device was in the proper position, it was understood that the VSD device was embolized into the left pulmonary artery within 24 hours after the procedure. The embolized device was removed with snare. Then the VSD was closed with the Lifetech KONAR MFO 9x7 mm.
Conclusion: Subpulmonic VSDs because of their proximity to the right aortic cusp have increased chances of aortic regurgitation, closure at early stages is advised. The MFO great versatility in the vascular access, allow the possibility of closing defects of larger sizes with lower profile sheaths that can be placed through both vascular approaches, anterograde and retrograde. However, the risk of embolization is high due to the softness of the device. Therefore, a device that is at least 2 mm larger than the measured defect size should be preferred to prevent device embolization.

Keywords: children, VSD, subpulmonic, multifunctional, occluder

P-171

Transcatheter closure of perimembranous ventricular septal defects guiding by transthoracic echocardiography in children

Irem Yenidogan, Ender Odemis; Koç University Faculty of Medicine

Background and Aim: Visualization of the ventricular septal defects andadjacent anatomical structures is crucial for the success of transcatheter VSDclosure. Although transesophageal echocardiography gives detailedanatomical information, it has also some disadvantages such as the requirement ofgeneral anesthesia and intubation and prolonged procedure time. The purpose ofthis study is to evaluate the safety and efficacy of transcatheter deviceclosure of perimembranous ventricular septal defects guided by transthoracicechocardiography in pediatric patients.
Methods: This study represents the results of one hundred fifty-three patients who had cardiac catheterization for the closure of perimembraneous ventricular septal defects. Patient records were reviewed for demographic data and clinical features. Patients' age, weight, pre and per-interventional transthoracic echocardiography and angiographic evaluation, procedure and fluoroscopy time, device types and complications were all recorded.
Results: A total of 153 patients (71 females and 82 boys) underwent transcatheter closure of the ventricular septal defect. Five patients had subpulmonic, 148 patients had perimembraneous defect sequentially. The procedure was attempted successfully in 97.38% of patients. The procedure processing time varies between 25 and 120 min (63.43 ± 23.26), the fluoroscopy time varies between 4.16 and 65 min (20.36 ± 12.81). The complications including embolization, vascular problems, hemolysis, valve insufficiencies were observed in 13 patients (8.5%) but no AV block and mortality were reported.
Conclusion: This study demonstrates that transcatheter closure of ventricular septal defect under the transthoracic echocardiography guidance in children is feasible, effective and safe method.

Keywords: perimembraneous ventricular septal defect muscular ventricular septal defect transcatheter VSD closure

P-172

Sequential percutaneous tricuspid and pulmonary valve implantation in a young child operated previously for ebstein anomaly

Irem Yenidogan, Ender Odemis; Koç University Faculty of Medicine

Background and Aim: We report a successful percutaneous tricuspid valve implantation (PTVI) followed by a percutaneous pulmonary valve implantation (PPVI) in a 11 kg weighted young child with Ebsteins anomaly of tricuspid valve and pulmonary stenosis who was previously treated surgically at one year of age with tricuspid ring annuloplasty and a transannular outflow patch. This article shows the feasibility of sequential percutaneous implantation of two valves in young patients with severe tricuspid and pulmonary valve insufficiency.
Methods: An infant diagnosed with Ebsteins anomaly of tricuspid valve with severe pulmonary stenosis underwent balloon pulmonary valvotomy at first month of life at another centre. A recurrence of severe outflow obstruction with tricuspid regurgitation warranted a surgical transannular patch repair of outflow with tricuspid ring annuloplasty using a 25mm ring at one year of age. When the child aged 2 years and 10 months, he had severe symptoms due to heart failure. Since he had medical history of previous surgery and unfavourable clinical evaluation for the open-heart surgery, we decided to implant tricuspid valve by transcatheter approach. In view of high surgical risks posed by severe biventricular dysfunction during a redo sternotomy, we decided for a transcatheter tricuspid valve-in-ring implantation.
Results: After the procedure, no complication occurred, and mild tricuspid valve regurgitation was detected by echocardiography. The patient was discharged on the second day of the procedure. At one year follow-up, the patient was asymptomatic and gained weight up to 14kg. The right ventricular end-systolic volume, end-diastolic volume and ejection fraction even improved comparing before PTVI on cardiac magnetic resonance imaging still right ventricular dilatation and dysfunction, were detected. Therefore, we decided to implant a valve into the pulmonary position by transcatheter approach. Patient has been followed for two years after PPVI and echocardiography shows still functional tricuspid valve and pulmonary valve without regurgitation. Patient reached 19 kg weight and he is doing well.
Conclusion: Sequential percutaneous tricuspid and pulmonary valve replacement can be performed successfully, even in small weight children. Given the risk of operative intervention, this strategy is a reasonable alternative to surgery for appropriately selected patients.

Keywords: percutaneous valve implantation tricuspid valve implantation Ebsteins Anomaly

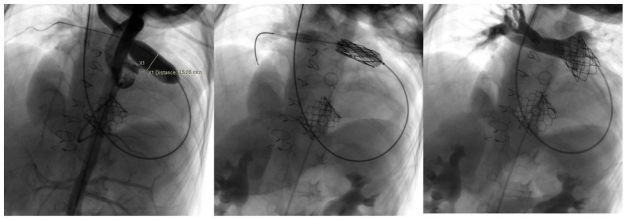

Figure 1. Percutaneously implanted valves in tricuspid and pulmonary position

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P-173

Bailout stenting for coarctation of the aorta in a preterm infant with hydrops fetalis

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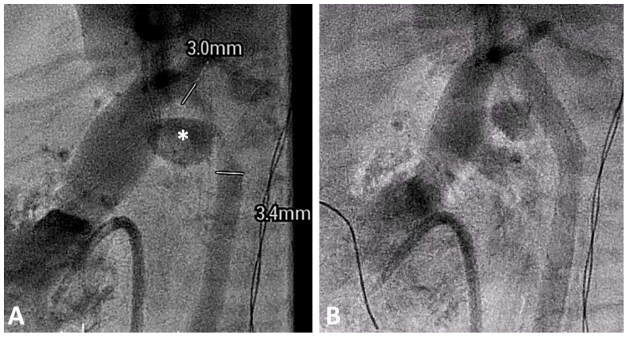
Background and Aim: Non-immune hydrops fetalis has multiple causes including cardiac malformations. It has a high mortality and morbidity rate.
Methods: We present the case of a preterm infant with hydrops fetalis caused by coarctation of the aorta and stenosis of the arterial duct.
Results: Hydrops fetalis with generalised oedema and ascites was discovered at 30 weeks of gestation. Ventricular function was impaired with bilateral AV-valve regurgitation. The child was delivered by urgent c-section. Birth weight was 2.2 kg. Postnatal management required immediate mechanical ventilation and inotropic support. Paracentesis was performed to improve ventilation. The child was anuric and developed worsening oedema and effusions. Echocardiography revealed normal intracardiac anatomy but showed severe aortic coarctation and a narrowed duct. Prostaglandin infusion was started, but repeat echocardiography confirmed prostaglandin-refractory duct stenosis. Surgery was not an option due to profound generalised oedema. The decision was made to perform balloon-angioplasty or, in case of an unsatisfactory result, coarctation stenting. Access was gained from the right femoral vein and artery with a 4F and 3.3F sheath, respectively. Angiography confirmed a tight coarctation and long segment distal arch hypoplasia. Peak-to-peak pressure gradient was 40 mmHg, with a non-pulsatile pressure trace in the descending aorta. Balloon angioplasty with a 4 mm balloon resulted in only minimal improvement. A 3.5 mm x 15 mm bare metal coronary stent was implanted reaching from the origin of the left subclavian artery into the descending aorta. Inotropic support could be weaned off over the next 3 days. Renal function recovered. The child lost about 1 kg of fluid over the first 3 days and was extubated 9 days after intervention. The stent was removed surgically at 10 weeks of age and an extended end-to-side anastomosis of the aorta was performed. Balloon angioplasty due to severe re-coarctation was performed 7 weeks after surgery.
Conclusion: Premature closure or stenosis of the ductus arteriosus can be a cause of non-immune hydrops fetalis due to right heart failure. In case of concomitant critical coarctation of the aorta, delivery of the affected foetus does not result in haemodynamic improvement. Bail-out coarctation stenting is a good option if surgery cannot be safely performed.

Keywords: Coarctation stenting, hydrops fetalis

Angiography pre- and post stenting



Left ventricular angiography via a transvenous route revealed distal arch hypoplasia and a tight coarctation with duct stenosis. The pulmonary end of the ductus arteriosus (*) showed aneurysmal dilatation (A). Stent implantation resulted in complete relief of the obstruction at the aortic isthmus (B).

P-174

Covered stent implantation for treatment of systemic venous baffle leak and obstruction after atrial switch operation

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[1]Department of Congenital Heart Disease and Pediatric Cardiology, University Hospital Schleswig-Holstein, Kiel, Germany; DZHK (German Center for Cardiovascular Research), Partner Site Hamburg/Kiel/Lübeck, Germany; [2]Department of Paediatric Cardiology, Alderhey Children's NHS Foundation Trust, Liverpool, UK; School of Biomedical Engineering and Imaging Sciences, King's College London, UK; [3]Department of Congenital Heart Disease and Pediatric Cardiology, University Hospital Schleswig-Holstein, Kiel, Germany

Background and Aim: Baffle-related complications such as stenosis and/or leak are not uncommon after the atrial switch operation. Percutaneous treatment options are often sought to avoid the morbidity and mortality associated with re-operation. The anatomy of the leaks may be complex and therefore cross-sectional imaging and 3D reconstruction can be helpful for procedural planning.
Methods: We report the case of a 44-year-old patient with transposition of the great arteries presenting with long standing cyanosis and exertional limitation after atrial switch operation.
Results: Atrial switch operation was performed at 3 years of age. Worsening cyanosis and limited exercise tolerance were noted during follow-up. Oxygen saturation dropped from 90% to 75% on an exercise test. Haemoglobin was elevated with 19.9 g/dl suggestive of chronic cyanosis. Echocardiographic assessment was limited by poor acoustic windows. Clinical suspicion for a baffle leak was high.

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Cardiac catheterisation was performed which demonstrated a large communication between the inferior systemic venous baffle and the pulmonary venous atrium. An attempt of device occlusion with an atrial septal occluder resulted in inferior baffle obstruction. Cross-sectional imaging and 3D reconstruction clearly visualized the anatomy. Implantation of a covered stent seemed to be possible on a virtual model without interfering with surrounding structures, especially the mouth of the coronary sinus which drained in the anticipated cranial stent landing zone.
Repeat catheterization was performed. Due to bilateral femoral vein thrombosis, vascular access was obtained from the right jugular vein. After balloon interrogation, a 6 cm long covered stent was implanted mounted on 26 mm diameter balloon-in-balloon catheter, and resulted in baffle leak exclusion and complete relief of obstruction.
Follow-up evaluation with MRI demonstrated unobstructed flow in the inferior baffle and no residual shunt. Saturation at rest and during exercise remained above 95%.
Conclusion: Besides the intended use for coarctation of the aorta and right ventricular outflow tract stenting, balloon-expandable covered stents can be used to treat baffle-related complications such as stenosis and/or leak after atrial switch operation. Cross-sectional imaging and 3D reconstruction including creation of a 3D virtual model is helpful tool in individualised pre-procedural planning for percutaneous congenital heart disease interventions.

Keywords: Covered stent implantation

3D reconstruction and virtual model

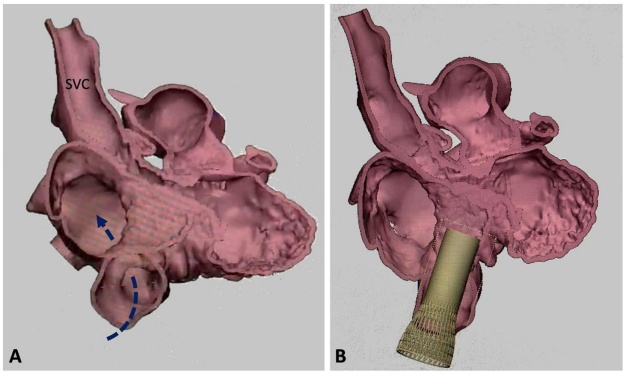

Figure 1 Cross-sectional imaging with and 3D reconstruction demonstrated a large communication (blue arrow) between the inferior systemic venous baffle and the pulmonary venous atrium (A). Stent placement appeared to be possible in a virtual 3D model without interfering with surrounding structures (B).

P-175
Stenting for coarctation of the aorta (COA) in small patients (< 5 years; < 15 kg) - short-, mid- to long-term results

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Background and Aim: Nowadays, small patients with native CoA are managed surgically. However, surgery for re-CoA is associated with increased morbidity and even mortality. There are patients with native CoA who present strong contraindications for surgery. CoA-stenting may be an alternative management option in these children. We present the short-, mid- to long-term results after CoA-stenting in small children.
Methods: Between Mai 2013 and November 2021, 77 small patients (age < 5 years; weight 15 kg) with CoA were managed with percutaneous stent implantation (male: 48, median age 5,0 [min. 0; max. 52] months; median weight 5,9 [min. 0,9; max. 14,7] kg).
Diagnoses: re-CoA post-surgery n = 71 (Norwood n = 31, CoA-end-to-end anastomosis n = 18, complex arch reconstruction n = 22), native CoA n = 6 with relative contraindications for surgery, for example ALL on chemotherapy, critical state or low birth weight of a patient. Eighty stents were implanted in 77 patients (Coronary stent n = 2, Osypka baby stent n = 4, Cook formula stent n = 75, Max LD n = 1). Unsuccessful balloon angioplasty preceded stent implantation in 15 patients.
Results: All procedures were successful. The median systolic gradient declined from 35 [min. 0; max. 118] mmHg to 0 [min. 0; max. 32] mmHg (p<0,001). The stenosed aortic diameter increased form a median value of 3 [min. 1; max. 6] mm to 7 [min. 3,5; max. 11] mm (p<0,001). In 19 patients the left subclavian artery was covered by the stent (re-opening by balloon angioplasty n = 4). There were no serious complications. The median follow-up time was 3,9 [min. 0; max. 103] months, during this time 19 patients required re-dilatation and 8 re-stenting with a larger stent. Median time of re-intervention was 7,4 [min. 0; max. 70] months.
Conclusion: Percutaneous stent implantation for Re-CoA and in selected patients with native CoA can be performed successfully in very young patients. However, repeated stent angioplasties and further on interventional "opening" of the stent with a larger stent or alternatively surgery shall be necessary to augment the aorta to adult size.

Keywords: Coarctation of aorta, Stenting, paediatric cardiology, hypoplastic left heart syndrome

P-177
Pulmonary artery stenosis and valve insufficiency in tetralogy of fallot – a flow analysis pre and post treatment

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Background and Aim: Tetralogy of Fallot (ToF) is the most common form of cyanotic congenital heart disease. Long-term complications include pulmonary regirgitation and pulmonary artery stenosis. Whether, when and how to treat these is still topic for debate. The aim of this study was to numerically explore the combination of pulmonary valve regurgitation and pulmonary artery stenosis in patients after ToF repair.
Methods: Two cases were selected, representing the most common patterns of pulmonary artery stenosis after ToF repair: one with left pulmonary artery stenosis and one case with bifurcation stenosis. A 3D model of the patient-specific anatomy was reconstructed. Mass flow was applied at the inlet and a 3-element windkessel at the outlets. For each case 4 scenarios were simulated: with- and without valve, with- and without stenosis.
Results: In stenotic arteries high flow velocities are found with pressure gradients and post-stenotic recirculation zones. High wall shear stress is seen in stenotic areas. Stent placement reduces velocity, pressure gradients and wall shear stress. Recirculation zones move towards the branch ostia. Regurgitation increases post-stent placement. Valve placement solves regurgitation and increases

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diastolic pressures. This results in a decrease in pulse pressure and flow velocities.
Conclusion: Stent placement results in an increase in regurgitation in the branches. The effect of stent implantation depends on the diameter, branching angle and origin of the stented artery. Implantation of a valve, reduces pulse pressure thereby decreasing flow velocity and wall sheart stress. This study emphasizes the importance of individual decision making in patients with the combination of pulmonary regurgitation and pulmonary artery stenosis.

Keywords: Hemodynamics, pulmonary valve regurgitation, pulmonary artery stenosis, valve replacement, stent placement.

P-178

10-year single center experience in percutaneous treatment of pulmonary artery stenosis in congenital heart disease: initial and long-term outcomes

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[1]Department of Pediatric Cardiology, Wilhelmina Children's Hospital, The Netherlands; [2]University Medical Center Utrecht, Department Cardiology, The Netherlands.

Background and Aim: Pulmonary artery stenosis (PAS) is seen in 2-3% of all congenital heart disease (CHD) patients. In the recent era, most stenosis are treated by stent implantation or balloon angioplasty. Large reports on long-term outcomes of these procedures are limited. The aim of this study was to describe our 10-year single center experience in interventional treatment of PAS patients with an emphasis on re-interventions.
Methods: This study has a retrospective, descriptive design. All pulmonary artery interventions were included. Data on procedural details, complications, re-interventions and survival was collected and analyzed. A logistic regression including 33 variables was performed to identify risk factors for re-intervention.
Results: 473 procedures were included. A total of 499 stents were implanted, 312 balloon angioplasties and 84 PPVI's were performed. Major adverse events occurred in 5.1% of the procedures. 25.6% of the procedures was followed by a re-intervention (mean time 29.3 (24.2) months). Reason for re-intervention was somatic growth in 50% and intima proliferation in 4% of the cases. Low age and weight at intervention, an underlying syndrome were independent risk factors for re-intervention.
Conclusion: PAS is a highly heterogenic substrate. Comprehensive interventional treatment is often necessary and the majority of the patients will need one or more re-interventions. Especially in the young CHD population, timing of interventional treatment needs to be carefully considered in the face of somatic growth. Under treatment needs to be avoided to guarantee lung development.

Keywords: Stent, balloon angioplasty, percutaneous treatment, pulmonary artery stenosis

P-179

To treat or not to treat pulmonary arteries: epinephrine provocation to unmask right ventricular load

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Background and Aim: Pulmonary artery stenosis (PAS) is common in congenital heart disease (CHD). The indication for treatment of PAS is primarily based on invasively measured pressure gradients. Anesthetics used during cardiac catheterization cause systemic and pulmonary hypotension. We hypothesize that this leads to underestimation of right ventricular (RV) pressure load and under treatment of PAS. The aim of this study is to describe the use of epinephrine to unmask RV pressure load in patients with PAS.
Methods: All cardiac catheterizations in which epinephrine was administered to evaluate RV load were included. There was an indication for treatment in case of a right to left ventricular (RV:LV) pressure ratio >0.6. The indication for treatment before and after epinephrine was evaluated to determine its role in clinical decision making.
Results: A total of 74 procedures were included. In all procedures the invasively measured LV pressure was below the awake blood pressure. At baseline, 33 patients had a RV:LV ratio >0.6. In 41 patients the baseline RV:LV ratio was <0.6. After epinephrine bolus, the LV pressure was raised up to the awake blood pressure. In 19 of the 41 patients without baseline indication, this resulted in a RV:LV ratio >0.6 thereby revealing the indication for treatment. No epinephrine related complications were registered.
Conclusion: The hypotensive properties of anesthetics during cardiac catheterization may lead to underestimation of RV pressure load. Invasive pressure measurements should be performed under conditions similar to awake conditions. Epinephrine provocation prevented under treatment in 25% of our patients.

Keywords: Hemodynamics, pulmonary artery stenosis, treatment indication, percutaneous treatment

P-180

Effectiveness and longevity of repeated percutaneous pulmonary valve-in-valve implantation: a two-centre experience

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Background and Aim: Transcatheter pulmonary valve implantation (TPVI) represents an important treatment option for right ventricular outflow tract (RVOT) dysfunction. In some patients reintervention is required for degeneration of the previously implanted valve. The placement of a second transcatheter valve, the so called "valve-in-valve (ViV) procedure", has been reported as a reasonable option in such cases, although limited data are reported about the longer-term results and whether they are associated with a different adverse event profile than first-time TPVI. This study sought to report safety and efficacy of the ViV procedure performed in two experienced German centers. In addition, longevity data of the second valve is a main objective
Methods: Data from two centers were entered retrospectively. They included all patients who provided written informed consent and underwent transcatheter reintervention on the RVOT after initial TPVI with either Melody or Sapien valves. Indication for the procedure was RVOT dysfunction due to stenosis and/or regurgitation. Patients with associated conditions requiring cardiothoracic surgery and active infection were excluded from the study.
Results: From December 2006 until December 2021 a total of 670 patients were treated with TPVI in the two centers. Of them, 34 (5,4%) received a ViV procedure after a median time of 58,4 months (range: 4,6-141 months). Mean weight and height of the ViV population were respectively 66 + 14,6 kg and 164 + 15,7 cm with a mean age of 30,6 + 10,6 years. Pre-stenting of the landing zone was done in 88% of patients during the first

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TPVI and in 65% at ViV procedure. Patients were followed-up for a median of 44,4 months (range: 0-104 months): one (2,9%) death was reported after 24 days (due to gut bleeding in a severely compromised patient), two patients (5,8%) underwent balloon dilatation without implantation of a third valve, while a ViViV procedure was performed in one (2,9%) patient. Four (12%) patients underwent surgical explantation during follow-up mainly due to restenosis of the RVOT

Conclusion: A percutaneous ViV procedure is technically feasible and effective for treatment of repeated RVOT dysfunction. In selected patients it is a safe option for expanding the time period free from surgical reintervention

Keywords: transcatheter pulmonary valve implantation (TPVI), Valve-in-Valve (ViV), right ventricular outflow tract (RVOT) dysfunction.

P-181/Moderated Poster

How is the melody™ tpv doing 15 years after implantation ?

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Background and Aim: Data on medium and long-term performance of the Melody™ valve are lacking. Analysis of up to 15 years follow-up.

Methods: Single center non-randomized prospective observational study from all implanted Melody™ valves in pulmonary position between 2006-2021. The following data were analyzed: mortality, valve failure, re-interventions, endocarditis and valve function.

Results: 234 Melody™ valves were implanted at mean age of 20.8y (3.5–81.6) with male/female ratio of 2:1. The indication was pulmonary stenosis (PS) in 110/234 (47.2%), regurgitation in 73/234 (30.9%) and mixed in 51/235 (21.9%) of the patients. The implant zone was native or patched RVOT in 79/234 (33.5%), homograft in 123/234 (52.8%) and bioprosthesis (e.g. Contegra™) in 32/234.The original implant zone diameter was mean 20mm (10–25). Patient survival was 97.8% at 15 years follow-up with 5 deaths at a mean age of 43.7y (20.7-74.0), none were valve related.

Valve failure was observed in 21/234 cases (8.9%), mainly due to endocarditis 11/21 (52.4%). Surgical removal of the valve was done due to endocarditis (n = 8) and to sub-PS (n = 3). Repeat TPV due to PS in 4 cases, endocarditis in 3 and stent fracture in 1. Balloon dilation in 15 cases (6.4%) due to somatic growth/PS.

The incidence of endocarditis was 13.4 per 1,000 patient years. Endocarditis occurred in 23/234 patients (9.8%) mean 2.6y (0.3–9.0) after implantation at 23.2y (8.1-49.5). The annual incidence peaked in 2015 at 3.5%, but with general measures decreased since 2018 to<1%.

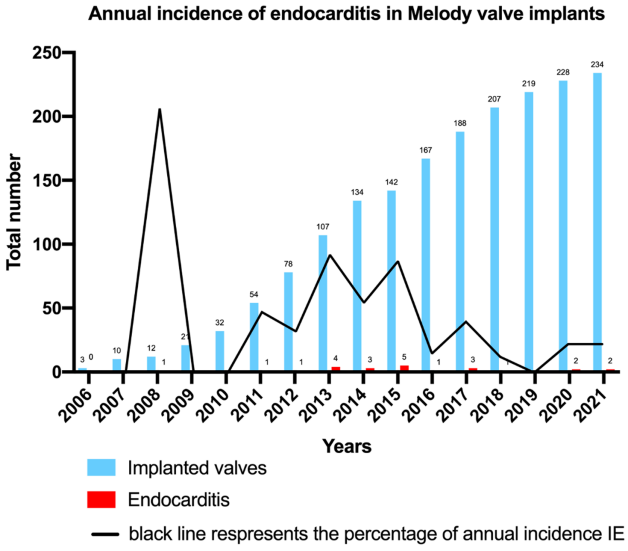
The valve function showed a mean RVOT gradient of 25.3mmHg (10-60) at 10 y and 27.2 mmHg (10-46) at 15y FU (p 0.455). The mean grade of pulmonary regurgitation was 1/4 (0-2) at 10y and remains 1/4 (0-3) at 15y follow-up (p 0.392).

The mean tricuspid regurgitation is mean 1/4 (0-3) at 10y follow-up and 1/4 (0-2) at 15y follow-up with an estimated right ventricular pressure of 32.8mmHg (10-60) at 10y follow-up and 37.0mmHg (19-60) at 15y follow-up (p 0.116).

Conclusion: Up to 15 years follow-up the Melody™ valve showed a low risk for valve failure with overall well preserved valve function. The main reason for valve failure was endocarditis. The Melody™ valve confirms to be competitive with other conduits.

Keywords: Melody valve, TPV, endocarditis, valve failure, valve function

Incidence endocarditis



visual presentation of annual incidence of endocarditis

P-182

Novel echocardiographic score to predict duct-dependency after percutaneous relief of critical pulmonary valve stenosis/atresia

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Background and Aim: This study aimed to identify clinical, hemodynamic or echocardiographic predictive features of persistent duct-dependency of pulmonary circulation (PDDPC) after effective percutaneous relief of pulmonary atresia with the intact ventricular septum (PA-IVS) or critical pulmonary stenosis (CPS).

Methods: From 2010 to 2021, 55 neonates with PA-IVS or CPS underwent percutaneous right ventricle (RV) decompression at our Institution. After successfully relief of critical obstruction, 27 patients (Group I) showed PDDPC, whereas RV was able to support the pulmonary circulation in the remaining 28 patients (Group II). Clinical, hemodynamic, and echocardiographic features of these two groups were compared.

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Results: No significant difference in clinical and hemodynamic data was found between the groups. However, tricuspid valve (TV) diameter <8.8 mm, TV z-score <-2.12, tricuspid/mitral valve annular ratio <0.78, pulmonary valve diameter <6.7 mm, pulmonary valve z-score <-1.17, end-diastolic RV area <1.35 cm2, end-systolic right atrium area >2.45 cm2, percentage amount of interatrial right-to-left shunt >69.5%, moderate/severe tricuspid regurgitation, RV systolic pressure >42.5 mmHg, tricuspid E/E' ratio >6.6 showed each significant predictive value of PDDPC. These parameters were used to build a composite echocardiographic score, assigning one point each above the respective cut-off value. An echocardiographic score ≥4.00 showed high sensitivity (100%) and specificity (86%) in predicting PDDPC.
Conclusion: Clinical and hemodynamic features do not predict the short-term fate of the pulmonary circulation after successful treatment of PA-IVS/CPS. However, a simple, composite echocardiographic score is useful to predict PDDPC and could be crucial in the management of this frail subset of patients.

Keywords: Pulmonary atresia; Pulmonary stenosis; Arterial duct stenting; Shunt; Echocardiography.

P-183

Spontaneous closure of the arterial duct after transcatheter closure attempt in preterm infants

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Background and Aim: Transcatheter closure of patent arterial duct (TCPDA) in preterm infants is an emerging procedure. Patent arterial duct (PDA) spontaneous closure after failed TCPDA attempts is seen but reasons and outcomes are not reported.
Methods: We retrospectively included all premature infants <2 kg with abandoned TCPDA procedures from our institutional database between September 2017 and August 2021. Patients data and outcomes were reviewed.
Results: Procedure was aborted in 14/130 patients referred for TCPDA. Two patients had spasmed PDA upon arrival in the catheterization laboratory and had no intervention. One patient had ductal spasm after guidewire cross. Four patients had unsuitable PDA size/shape for closure. In seven patients, device closure was not possible without causing obstruction on adjacent vessels. Among the 12 patients with attempted TCPDA, five had surgery in median delay of 3 days after TCPDA and seven had a spontaneous PDA closure within a median delay of 3 days after the procedure. Only the shape of the PDA differed between surgical ligation group (short and conical) and spontaneous closure group (F-type).
Conclusion: In case of TCPDA failure, mechanically induced spontaneous closure may occur early after the procedure. Surgical ligation should be postponed when clinically tolerated.

Keywords: arterial duct, ductus arteriosus, closure, premature, transcatheter

P-184

Endovascular exclusion of a threatening thoracic aortic aneurysm in a young girl with severe aortitis

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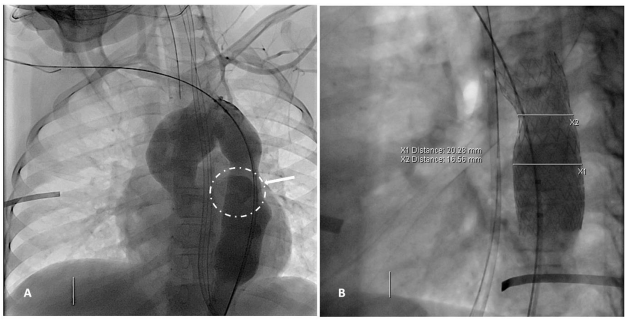
Background and Aim: Endovascular treatment is becoming an interesting option in the management of complex aortic lesions, for which surgery was previously the gold standard even in children thanks to new device, small delivery sheath and alternative vascular access.
Methods: We report the case of an 11-old girl weighing 36 kg, hospitalized for life-threatening hemoptysis occurring in the setting of severe aortitis, with compression of the left main bronchus by a large aneurysm of the descending thoracic aorta. This severe aortitis was most certainly of infectious origin, tuberculosis being the main hypothesis.
Results: A multidisciplinary team opted for a two staged strategy with first line hybrid management to exclude the aneurysm, and in a second time a surgical reconstruction of aorta and its branches. Because of the small size of the femoral vessels on angioCT scan (5mm diameter), vascular access was achieved through surgical denudation of the left common femoral artery and a 14 Fr sheath was gently inserted over a superstiff guidewire. The aneurysm was excluded with a 57 mm covered OptimusTM XXL (AndraTec, GmbH, Koblenz, Germany) mounted on a 24x40 mm BIB ballon (NuMED, Inc., Hopkinton, NY, USA). The angiographic result was satisfactory, no endoleak was noted with full exclusion of the aneurysm. The stent was stable without disruption of the aorta. Femoral artery was reconstructed with an end-to-end anastomosis and the patient received an anticoagulation with unfractionated heparin. A Doppler ultrasound performed 24 hours after the procedure showed femoral permeability.
Conclusion: Endovascular treatment is an interesting alternative option in staged treatment of threatening thoracic aorta aneurysm, allowing the exclusion of the aneurysm in critically ill patients not eligible for surgery.

Keywords: thoracic aortic aneurysm, aortitis, endovascular treatment



A: Severe aortitis with a dilated Aorta of irregular caliber; saccular aneurysm of the descending thoracic Aorta compressing the left stem bronchus (arrow).
B: OPTIMUS covered stent excluding the aneurysm.

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P-185

Begraft aortic stents: a european multi-centre experience reporting acute safety and efficacy outcomes in congenital heart diseases

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Background and Aim: : Stent implantation for the treatment of vessel stenosis in congenital heart diseases has become the preferred method of treatment. Availability of covered stents may decrease complications and may have an important role in the management of patients with complex anatomy. Aim of this study is to evaluate the feasibility and safety of the pre-mounted cobalt-chromium stent-graft covered ePTFE Aortic BeGraft in a broad spectrum of vascular lesions.
Methods: This is a multicenter retrospective study of 60 implanted Be Graft Aortic stents between 2016 and 2021 in four different european centers.
Results: 57 patients aged 12 years (range 4-59 years), with body weight of 59.5 Kg (range 14-103 Kg) underwent Be Graft stent implantation. Fourty-seven patients had aortic coartation, 6 patients had stenosis of pulmonary conduit, 2 patients had conduits stenosis in a total cavopulmonary connection (TCPC) and one patient was treated for a wide portocaval fistula. All the stents were implanted successfully. Median stent diameter was 16 mm (range 12-24 mm) and median length was 39 mm (range 19-49 mm). Three patients received 2 stents. Mean final long sheath size was 12 +/- 3 Fr. In all cases was possible to treat the underlying lesion (for aortic coarctation peak velocity dropped from a mean of 3,5 m/s to 2,1 m/s). Three major complications occurred. During a pulmonary conduit stenting procedure, the stent balloon ruptured with some difficulties in retrieval, 1 patient experienced a cerebral air embolization without neurological consequences and 1 patient had a femoral artery occlusion requiring vascular surgery for reperfusion. Median follow up was 11 months. No late complications occurred and only 1 patient needed aortic coarctation stent re-dilatation due to somatic growth
Conclusion: Be- graft stent can be used safely and effectively in a wide spectrum of congenital heart diseases. Whether these good results will be stable in the long term follow up still needs to be investigated given its recent introduction into clinical practice.

Keywords: Aortic Begraft stents, vascular lesions, complex congenital heart disease

P-186

Rescue and bridge to recovery by returning to fetal circulation: hybrid approach in a newborn with acute left heart failure

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Background and Aim: The hybrid technique consisting of ductal stenting and bilateral pulmonary banding is one of the treatment options in high-risk patients to avoid neonatal cardiopulmonary bypass.
Methods: Here, we report on a newborn with acute left heart failure due to acute viral myocarditis treated with Hybrid procedure.
Results: A 8 day-old newborn, weighing 2700 g, was referred to us because of fever of 39.2 °C. He was admitted to NICU with a possible diagnosis of neonatal early sepsis. Physical examination showed tachypnea (55 /min), tachycardia (180 beat/min), blood pressure of 55/38 mmHg, preductal and postductal oxygen saturation of 80% and 70% consequently. Laboratory testing revealed C-reactive protein 4.3 mg/dl, NT-proBNP >70.000 pg/ml. Enterovirus IgG was positive by ELISA. Echocardiography showed impaired left ventricular function, moderate mitral and tricuspid insufficiency, widely patent ductal arch with right to left shunt, suprasystemic right ventricle systolic pressure of 60 mmHg and enlarged left ventricle. We started furosemide, spironolactone, hydrochlorothiazide and lisinopril as well as beta blocker and intravenously prostaglandin (5 ng/kg/min). At the second day of admission, the patient progressed to cardiovascular decompensation. He was intubated and initiated milrinon (0.5 μg/kg/min), suprarenin (0.6 μg/kg/min), noradrenalin (0.1 μg/kg/min) and nitric oxid (20 ppm) treatment. However, he deteriorated despite of intensive medication. We decided to perform an interventional cardiac catheterization. Coronary angiography revealed no ischemia-related disease. We carried out stenting of ductus as well as balloon dilatation of patent foramen ovale for stabilization of the systemic circulation and surgically bilateral pulmonary banding for restriction of pulmonary blood flow. After balloon dilatation of PFO, the gradient between left and right atrium was decreased from 16 mmHg to 4 mmHg. After stenting of ductus, oxygen saturation was 85% as postductal and 75% as preductal. One day after the intervention, the patient was underwent surgically bilateral pulmonary banding. After three weeks, he was discharged with anticongestive therapy. On follow-up of three years, echocardiography showed mild mitral insufficiency, enlargement of left atrium and ventricle as a sequel of enteroviral myocarditis.
Conclusion: Hybrid procedure can be an alternative palliative approach for rescue and bridge to recovery in a newborn with acute left heart failure

Keywords: newborn, acute heart failure, hybrid, transcatheter,

P-187

A feasible alternative approach for central venous line: percutaneous transhepatic placement of a hickman catheter in a young adult

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Background and Aim: Central venous accesses are especially important for patients with disease requiring total parenteral nutrition and intravenous medication. For this reason, the jugular, subclavian, femoral or intercostal veins are used commonly in clinical practice.
Methods: Here, we present the case of an 25-year-old girl with short-bowel syndrome indicating parenteral nutrition, who underwent interventional subhepatic placement of a Hickman catheter into the right atrium as an alternative option for central venous line.
Results: A 21-year-old female with short-bowel syndrome and a central venous catheter for necessity of parenteral nutrition had been referred to us because of thorax pain, serous discharge from the catheter insertion site and fever of 39.1 °C. A Hickman catheter had been placed via anterolateral thoracotomy in 2015 because of thrombosis of the great veins. Laboratory testing had showed CRP 66.3 mg/dl. We had started ampicillin-sulbactam and linezolid therapy and removed the catheter in consequence of catheter infection as well as laid down a new Hickman catheter by transhepatic approach in 2017. After two years, the lumens of

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catheter malfunctioned and did not re-open despite of alteplase treatment. We decided to set a new Hickman catheter via subhepatic approach. We extracted the previous catheter easily without problem and closed up the vein with a coil 5x5 mm with no complication. Then we placed the the Hickman catheter over the 0,014" wire via the hepatic vein into the right atrium without complication (Figure 1).

Conclusion: This case demonstrated that the placement of central catheter by subhepatic approach could be an alternative choose for obtaining a central venous line with feasible intervention and simple replacement if it is needed.

Keywords: central venous line, Hickman catheter, transcatheter

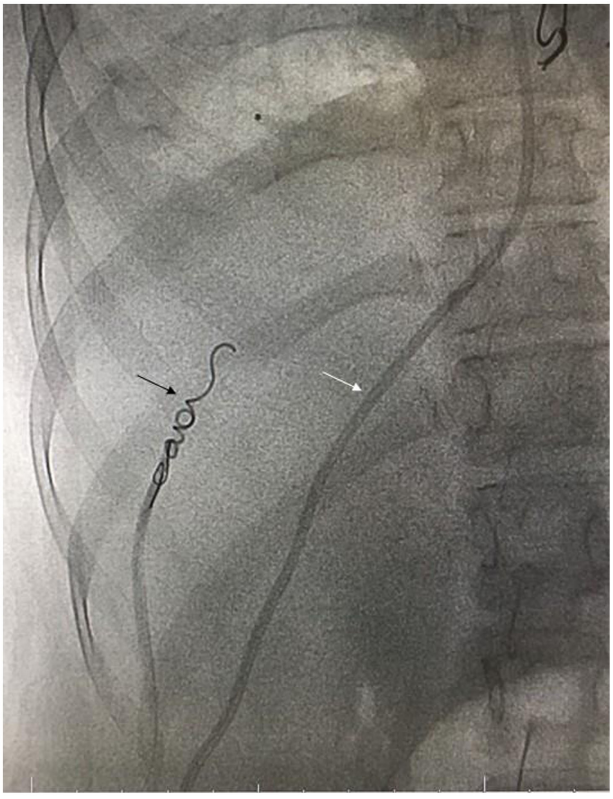

Figure.1 In this picture, we can see the occlusion of previous central venous line with the coil (black arrow) due to malfunction of the catheter and the new central venous line (white arrow) via the hepatic vein to right atrium.

P-188

Management of Aorta to left ventricular tunnel over two decades in a national paediatric cardiology centre

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Background and Aim: Aorto-ventricular tunnel is a rare congenital cardiac defect with an incidence of less than 0.1% of all children born with congenital cardiac disease. The tunnel originates in the ascending aorta and bypasses the aortic valve creating a direct connection between the ascending aorta and either the right or left ventricular cavity. It was originally reported by Levy et al. who coined the phrase aorto-left ventricular tunnel (ALVT). The majority of cases originate above the right coronary cusp and present in the first year of life.

Methods: This is a retrospective case series of all patients who were diagnosed with an ALVT from 2001 to 2021 at the national paediatric cardiology centre at Children's Health Ireland, Crumlin, Dublin.

Results: Four patients presented during the study period with ALVT at a median age of 8 months (range 0.1-10 months). Two patients (50%) had associated complex cardiac anomalies. Patients with ALVT will often demonstrate "to and fro" murmurs associated with thrills and bounding pulses. The classic clinical examination findings complements the imaging modalities. Echocardiogram is the primary imaging modality and in the majority of cases is sufficient in making the diagnosis. The use of contrast-enhanced CT can be helpful in outlining the tunnel further and clarifying its relationship to the coronary arteries. The mainstay of treatment is procedural with older reports focus primarily on surgical closure, but in more recent times, transcatheter closure has been describe as a safe and feasible alternative. In this patient cohort, one patient with associated non-compaction cardiomyopathy and dysplastic aortic and pulmonary valves underwent surgical closure while the subsequent three patients were treated by transcatheter closure via right femoral artery access with successful occlusion and excellent outcomes.

Conclusion: Aorto-left ventricular tunnel is a rare anomaly which previously necessitated surgical repair. This case series highlights not only the importance of the clinical examination in its detection but also the usefulness of multi-modal imaging prior to definitive closure. Furthermore, we demonstrate the safety and feasibility of a transcatheter closure approach in appropriately selected patients among this heterogenous patient cohort.

Keywords: Aorto-left ventricular tunnel; Interventional Cardiology

Echocardiographic image of ALVT

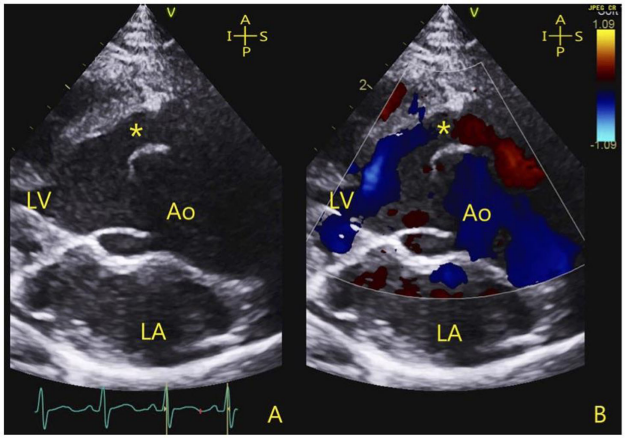

Figure 1. Colour doppler imaging of the ALVT(*) in a parasternal long axis view. Ao: aorta, LV: left ventricle, LA: left atrium.

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Non-surgical stage I to avoid neonatal surgeries in newborns with HLHS and HLHC

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Background and Aim: Therapy for Hypoplastic Left Heart Syndrome (HLHS) and -Complex (HLHC) is still associated with significant morbidity and mortality. We aim to improve the outcome of these vulnerable patient groups by minimizing neonatal surgical trauma and postponing surgeries to later infancy. We describe four patients who underwent a compassionate stage 1 procedure (S1P) using a novel, fully transcatheter approach.
Methods: The non-surgical S1P was performed in three newborns with HLHS and one with HLHC with associated aortic atresia and inlet ventricular septal defect.
Non-surgical S1P was performed in 4 sedated, spontaneously breathing newborns with a median weight of 3345 g and a median age of 5 days. First, an endoluminal pulmonary artery (PA) banding of the right and left PA was performed. For this purpose, Micro Vascular Plug (MVP) devices were manually modified to pulmonary flow restrictors (PFR) by removing one of the ten PTFE-covered end-cells of the MVP with a scalpel. Following bilateral PFR's placement, the arterial duct was stented with a self-expandable Sinus-Superflex-DS. At discharge, all patients received bisoprolol and clopidogrel.
Results: Non-surgical S1P were performed without mortality and complications. Taking the learning curve into account, the median length of hospital stay was 16 days. During the interstage, three reinterventions were necessary. It included stenting of the atrial septum in one, dilatation of left-sided pulmonary vein stenosis in another, and a partial dilatation of the right PFR followed by placement of a coronary stent within a slowly developing aortic coarctation in the third patient with HLHC. Two patients received a surgical comprehensive stage II at four months. The patient with the HLHC received a Yasui operation at the age of three months. The removal of the PFR´s went smoothly with only a slight injury of the pulmonary intimal tissue. The fourth patient died in the interstage period.
Conclusion: Non-surgical S1P was successful in all four newborns. One patient died in the interstage period. A close follow-up during interstage is mandatory as the initial morphologies, and functional status can suddenly change. A prospective, multicenter study will be initiated to compare the transcatheter with the hybrid approach.

Keywords: Hypoplastic left heart, pulmonary flow restrictor, transcatheter technique, arterial ductus stenting

P-190

Successful transcatheter closure of a right coronary artery to right ventricle fistula in a newborn

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Background and Aim: BACKGROUND: Coronary artery fistula (CAF) complicated by coronary artery ectasia is a rare congenital malformation with an estimated incidence between 0.2-0.6%. Indications for intervention and treatment strategies vary depending on patient size and unique anatomic features and frequently pose a significant challenge in youngest patients
Methods: We present a case of successful percutaneous closure of the fistula performed on five-day-old child weighed 3 kgs. An asymptomatic one-day old term neonate was referred for investigating a pathologic heart murmur. Vital signs, oxygen saturation and electrocardiogram were normal. The echocardiogram demonstrated a large fistulous communication between dilated proximal right coronary artery (RCA) and the enlarged right ventricle (RV) with reduced systolic function. Patent foramen ovale and arterial duct were present. A computed tomography (CT) coronary angiogram confirmed the CAF anatomy: course of ectatic RCA and RV entry point. Based on apparent haemodynamic significance of the CAF and potential risk of developing heart failure, pulmonary hypertension, myocardial ischaemia, arrhythmia, thrombosis or rupture if left untreated, early transcatheter closure of CAF was uneventfully performed. Coronary angiography delineated small coronary branches arising distally to the fistula (Figure 1A). A 6mm-Amplatzer Vascular Plug (AVP) Type II was deployed within the distal segment of the fistula via 4Fr catheter in left femoral artery using retrograde arterial approach. Complete occlusion of fistula was achieved while the perfusion of the distal coronary artery branches was preserved (Figure 1B). Post-procedure prophylactic intravenous administration of Heparin was initiated (20units/kg/hr) and this was eventually converted to oral Aspirin (15 mg).
Results: Transient non-specific repolarization ECG changes persisted for 48 hours post-procedure. At 6-month follow-up patient was well and thriving. Echocardiogram demonstrated normal ventricular function.
Conclusion: Indications for surgical or percutaneous closure of CAF in children are still controversial. Feasibility and effectiveness of transcatheter closure in older children has been demonstrated but is more challenging in younger and smaller patients. Further studies are required to define patient selection criteria and interventional technique, outcomes and long-term prognosis in this group of patients.

Keywords: coronary artery-ventricular fistula, transcatheter closure

Device closure of coronary artery fistula

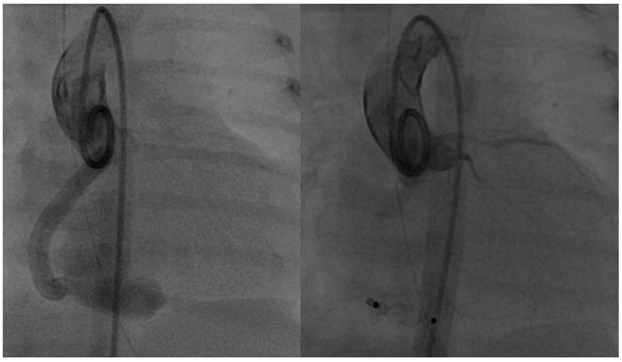

Figure: 1A Injection in ascending aorta, RCA is grossly dilated and forms a fistulous connection with the RV at an apparent small pouch. 1B Insertion of 6mm-AVP, the fistula is completely occluded with no residual flow seen
(Abbreviations: RCA: right coronary artery, RV: right ventricle, AVP: Amplatzer Vascular Plug

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Percutaneous retrieval of embolized catheter fragments from preterm newborn to adult: a single center experience for 10 years

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Background and Aim: We aimed to present our experiences in 15 of 16 patients with embolized catheter which were successfully retrieved by percutaneous intervention
Methods: During 10 years, sixteen patients with embolized umbilical vein catheters, port catheters, central venous catheters, catheter fragments and guide wires were examined. Demographic characteristics of the patients, catheter indications, embolized catheter types, localizations and lengths of catheter, durations of flora, entry points during retrieval of embolized catheters, snare's features used, grasping location of the embolized catheter, and additional procedures were examined retrospectively
Results: Of the 16 patients, 7 were girls, their ages were between 11 days and 39 years. 14 of the patients were children. Their weights were between 1.3 kg and 65 kg. The umbilical vein catheter in five patients, port catheters in seven patients, a double-lumen central venous catheter in one patient, the distal part of the fragmented sheath in one patient, and the guide wire in 2 patients were embolized
Conclusion: The retrieval procedures of umbilical vein, central vein and port catheters are safe in experienced hands. Rarely, those catheters may break and embolize. As soon as it is diagnosed, embolized catheters should be removed to prevent complications. Since the retrieval of embolized catheters by percutaneous transcatheter route is safe and successful, it should be used as the first choice

Keywords: children, embolized catheter, transcatheter retrieval, umbilical vein catheterization

P-192/Moderated Poster

Piccolo in interventional pda closure; multicenter study from premature to adolescent

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Background and Aim: The closure of PDA with the transcatheter method has taken its place as the first choice in treatment with the development of new devices and techniques. In this multicenter study, we discussed the mid-long-term results of the efficacy and safety of the The Amplatzer Piccolo Occluder previously called ADO II AS device in PDA closure in premature, infant and children.
Methods: Between January 2016 and October 2021, a total of 645 patients, 152 of whom were less than 1 months old, underwent PDA closure with the Amplatzer Piccolo Occluder device from the 5 different centers in Turkey. The median age of the patients was 2.2 years (3 days-17 years). There were 62 patients ≤1.5 kg, 90 patients ≤1.5-3 kg, and mean follow-up was 20.4 months, range (1-71) months. 372 (54.5%) were females, 396 of all patients closed by retrograde route. The mean PDA diameter was 1.8 mm at its narrowest point. Ductal anatomy most frequently was Type A in 285, Type E in 92 and Type C in 90 patients. Duration of the fluoroscopy were 6.2 mins. The procedure successful rate 99.1% in all cases. Device embolization occurred in 6 patients (0.9%), 5 of them were removed with snare and one patient underwent surgery. Cardiac perforation developed in one premature baby, other than that, no major complication or death was observed. Mild-moderate stenosis was observed in the left pulmonary artery in 4 patients (0.6%) and in the descending aorta in 2 patients (0.3%). Hemodynamically insignificant residual shunting was observed in only one patient (0.1%) at the latest follow-up.
Results: This study has shown that the Piccolo Occluder device is safe and effective in closing PDAs in all age groups from premature to adolescent. In addition, Piccolo device has some an advantageous such as low profile to use in premature and newborn babies, and minimal embolization risk, low residual shunt rate after closing.
Conclusion: The Piccolo Occluder device can be considered as an ideal occluder. It can be better positioned and stable in small PDA's. The lower profile, smaller delivery catheter size and symmetry of this device allows for venous or arterial approach.

Keywords: The Amplatzer Piccolo Occluder, Patent Ductus Arteriosus closure, children, premature baby, pediatric interventions

P-193

Cardiac MRI and catheterization combined – a pragmatic and cost effective setup

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Background and Aim: Combined cardiac MRI-catheterization (MRI-cath) has the potential to produce a comprehensive hemodynamic work up and to mitigate the pitfalls of conventional catheterization, especially in patients with complex congenital heart defects requiring pulmonary vascular resistance (PVR) assessment. One important drawback of MRI-cath is the need for an advanced and costly hybrid suite which is not available in many centers worldwide. We present herein a pragmatic workflow utilizing existing resources at the Skåne University Hospital Lund, Sweden.
Methods: From July 2017 to December 2021, 49 combined cardiac MRI-catheterizations were performed. Since no MRI-cath suite was available, patients were hemodynamically assessed in the anesthesia room adjacent to the MRI suite using a mobile X-ray C-arm or transported between the MRI and standard catheterization suite under general anesthesia. The selected patients displayed a variety of highly complex cardiac anomalies, 30 with univentricular (UV) and 10 with biventricular physiology. In 7 patients within the UV group, MRI-cath was repeated ≥2 times. Patients underwent MRI-cath to A) assess operability B) guide transcatheter interventions based on MRI data C) assess interventional outcome using

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MRI D) primarily assess the PVR for nonsurgical reasons. Assessment of PVR was of key importance in the majority of cases.
Results: In group A, 14 of 28 patients were deemed operable and displayed intraoperative pulmonary pressures in line with PVR obtained via MRI-cath; 13 patients had an uneventful postoperative course and 1 patient died shortly postoperatively due to pulmonary bleeding. An additional 12 patients were considered inoperable, 10 of whom were late presenters. In group B and C, 10 interventions were performed and the planned interventional outcome was achieved in all. In group D, ischemic stroke occurred in 1 case, 3 days after MRI cath. The patient fully recovered following standard care.
Conclusion: This study shows that combined MRI and cardiac catheterization using available resources is feasible and safe, thus enabling access to important hemodynamic information and clinical decision-making for a more patients with complex CHD.

Keywords: Combined MRI catheterization, cMRI, Magnetic Resonance Imaging, PVR, Hemodynamics, Complex CHD

P-194
Clinical hypnosis associated with local anesthesia for cardiac catheterization in paediatric population

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Background and Aim: Hypnosis is a promising non-pharmacologic adjunct treatment for improving pain and interventional anxiety management in paediatric interventions. Its efficiency and impacts in paediatric cardiac catheterization (CC) are unknown.
Methods: In a prospective monocentric study, all patients aged <18 years who had CC under hypnosis with local anesthesia from January to December 2021 were included. Pain and anxiety were evaluated by the ANI (Analgesia Nociception Index) measured during the intervention, and the Visual Analogue Scale (VAS) assessed by the patients after the intervention.
Results: Sixteen patients were included. Median age was 10.5 years old |range 4-16 years], Median weight was 37 kg [range 15-79 Kg]. Catheterizations were interventional in 10 patients (62.5%) and for diagnostic purposes in 6 patients (37.5%) (table1). Hypnosis indications were general anesthesia (GA) contraindication in 4 patients (25.0%), the need of accurate pressures measurements without GA interference in 2 patients (12.5%) and interventionist/patients preferences in 10 patients (62.5%). In one patient of 4 years old, the hypnotic status was not achieved and the procedure was aborted before the installation. Procedures were accomplished successfully in 15 patients (93.7%) without any additional sedation even in challenging cases; large atrial septal defect (ASD) closed with 39 mm Occlutech device in 16 years old patient, and catheterization in 14 years old patient with Fontan failure, elevated pulmonary vascular resistances, cyanosis (Hemodynamic Vulnerability Score = 5) with GA contraindication. Median procedures and fluoroscopy times were 67.5 min and 5.3 min respectively. In one case, pulmonary artery pressures normalized comparing to previous catheterization under local anesthesia alone leading to the cancellation of cardiac surgery on mitral stenosis. VAS score was under 5/10 in all patients. The ANI remained above 50 (non painful zone) in all but one patient. No significative decrease of the ANI was found during the intervention comparing to baseline ($p = 0.62$). One patient with language difficulties had short and transient decrease of the ANI (49) during one minute after the puncture. No complications were recorded.
Conclusion: Paediatric CC are feasible under hypnosis even in complicated cases. Hypnosis was efficient to manage pain and stress in all cases, it ensures more reliable pressures measurement.

Keywords: Cardiac Catheterization, Hypnosis, paediatric population, congenital heart disease, Analgesia Nociception index

Clinical Hypnosis associated with local anesthesia for cardiac Catheterization in Paediatric population

Patients	Age (years)	Gender	Weight (kg)	Procedure indication	Hypnosis Indication	Anxiety previous procedure	Clinical Hypnosis success	ANI score (mean)
1	16	F	48	ASD closure	Family choice	Low	Yes	49
2	8	M	21	diagnostic catheterization/ Pulmonary hypertension	Reliable hemodynamic assessement	Moderate	Yes	-
3	7	F	18	ASD closure	Family choice	Importante	Yes	86
4	11	M	48	diagnostic catheterization/ Pulmonary hypertension	GACI	-	Yes	75
5	16	F	74	ASD closure	Family choice	Importante	Yes	52
6	4	M	20	PDA closure	Family choice	Low	Yes	70
7	16	F	50	Coronarography	GACI	Moderate	Yes	69
8	7	F	21	ASD closure	Family choice	Low	Yes	92
9	12	F	43	diagnostic catheterization/ aortic coarctation	Reliable hemodynamic assessement	Low	Yes	67
10	16	M	79	electrostimulating procedure	Family choice	Low	Yes	-
11	4	M	15	PDA closure	Family choice	Moderate	No	/
12	8	F	46	Diagnostic catheterization/ anomalous pulmonary venous return / aortic coarctation	Reliable hemodynamic assessement/GACI	Low	Yes	79
13	14	F	25	diagnostic catheterization/ Fontan	Reliable hemodynamic assessement/GACI	-	Yes	III
14	12	F	47	PDA closure	Family choice	-	Yes	64
15	6	M	19	ASD closure	Family choice	Low	Yes	81
16	11	M	30	ASD closure	Family choice	Low	Yes	97

Characteristics of patient who undergone cardiac catheterization under clinical hypnosis
ASD: Atrial septal defect, PDA: Persistent Ductus Arteriosus, GACI: General anesthesia contraindication

P-196
A new solution for stenting large right ventricular outflow tracts before transcatheter pulmonary valve replacement

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Background and Aim: Pre-stenting right ventricular outflow tracts (RVOTs) before transcatheter pulmonary valve replacement (TPVR) is essential. Optimus-XXL is a new extra-large, balloon-expandable, cobalt-chrome stent with promising technologies. We assess our preliminary experience with this new stent and focus on its performance and outcomes.
Methods: From June 2020 to November 2020, 15 patients with congenital heart diseases, dysfunctional RVOTs, and target TPVR diameter ≥23 mm received Optimus-XXL stents before proceeding to TPVR using the Edwards SAPIENTM valve. Standard safety and outcomes were prospectively assessed.
Results: Patients' median age and weight were 25.8 years (range 10.5-63.1 years) and 58 kg (range 43.8-101 kg), respectively. Underlying diagnosis was Tetralogy of Fallot (66.7%) and RVOTs were patched (80%). 15 bare-metal stents were implanted using femoral (n = 14) and jugular approaches (n = 1). One conduit rupture was immediately controlled with a covered Optimus-XXL. Median stent length was 43 mm (range, 33-57 mm) and median target expansion diameter was 28 mm (range, 23-30 mm). Two procedural incidents occurred during stent delivery and were percutaneously treated. Stent stability was documented during TPVRs immediately performed in 14 patients. The median percentage of stent expansion was 95.9% (range, 83.2-100%), regardless of balloon size. The median stent recoil was 4.1% (range, 0-16.8%). The median percentage of stent shortening was 13.7% (range, 7.2–19.5%) at a median diameter of 28mm (range, 24-30

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mm). There was no stent fracture on the short-term follow-up (median, 4.5 months).
Conclusion: We report the first 15 successful human implantations of Optimus-XXL stents in dysfunctional RVOTs with excellent preliminary results. Safety and efficacy were proven across a wide spectrum of patient sizes and anatomical variations. Optimus-XXL should be considered as a valuable adjunct in the armamentarium for routine and complex TPVR procedures.

Keywords: Right ventricular outflow tracts; Optimus-XXL; stent implantation; transcatheter pulmonary valve replacement; new device.

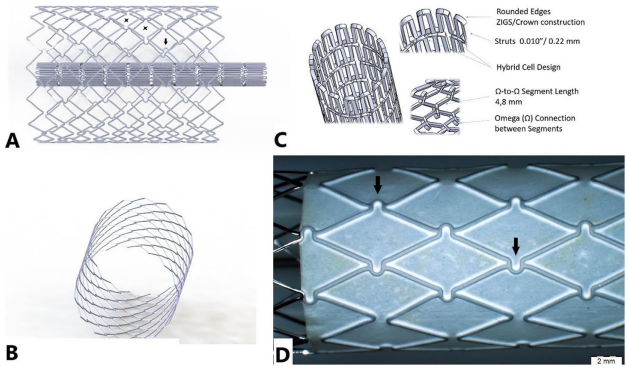

Figure 1 Bare metal Optimus-XXL stent (57 mm) before and after expansion (A, B). Note the special hybrid design (black stars) with omega-flex-connectors (black arrows). Detailed schematic presentation of the Optimus-XXL special construction features (C). Covered Optimus-XXL stent (D). Note how inner and outer Nano-PTFE stretchable layers are thermally bonded to the stent giving the impression that the covering has been pressed onto the metal struts while both ends remain bare segments. Optimus is trademark of AndraTec, GmbH (Koblenz, Germany). Reproduced with permission of AndraTec, GmbH© 2021. All rights reserved.

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Requirement and management of redo-interventions after tetralogy of fallot repair – a retrospective study

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Background and Aim: Re-interventions after initial repair of TOF are frequently required depending on the underlying anatomic features and the subsequent technique of surgical repair. Data on follow-up and postoperative outcome are essential to evaluate a good timing and choice of treatment.
Methods: 183 patients corrected for TOF between 2003 and 2019 at UZ Leuven, Belgium, were evaluated for their need of re-interventions retrospectively. Mean follow-up was 8.9 y (0.38 to 16.38 y). Data on anatomic findings, surgical and interventional procedures and their timing were analyzed.
Results: Initial TOF repair was performed by placing a transannular patch in 70/183 (38%) patients (group 1), an infundibular patch alone or in combination with a Hegar dilation or a supravalvular patch in 97 (52%) children (group 2). In 15 (8%) patients just a VSD closure was performed (group 3) and 1 patient received a RVOT conduit for repair.
In group 1, 27/70 (39%) needed a re-intervention and 3/70 (4%) patients a second re-intervention. A percutaneous valve replacement was performed in 19/27 (70%) and surgical valve replacement in 5/27 (18%) patients. An infundibular resection was done in 2 patients and a revision of the transannular patch in 1. The main reason for re-intervention was severe pulmonary insufficiency (≥ 3/4, 75%).
In group 2, 14/97 (24%) patients required a re-intervention which was significantly lower compared to group 1 ($p < 0.05$). Indication was RVOT obstruction (PIG > 60 mmHg). Surgery was performed by valve replacement in 2/14 (14%) patients, an infundibular resection in 6/14 (42%), a supra- or infundibular patch in 5/14 (35%) patients and a transannular patch in 1/14 (7%).
Patients in group 3 (VSD repair) had no re-intervention.
Conclusion: Despite following a valve sparing management, 38% of TOF patients were corrected by placing a transannular patch associated with a significant higher risk of re-interventions. These were performed preferentially by percutaneous valve implantation in our center. Although less frequent, surgical repair was required due to the anatomic position of the stenosis in patients with valve sparing repair. The best timing of re-intervention, considering both, the course of earlier re-interventions and possible downsides of postponing, needs prolonged follow-up periods.

Keywords: TOF, follow up, re-interventions, surgery, percutaneous valve implantation

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Serial dilatation and intentional fracturing of stents to adapt to patient growth – bench side tests and initial clinical results

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Background and Aim: Controversy exists in stent implantation in small children because these patients outgrow the stents. We developed a treatment strategy on the bench side including intentional stent fracturing and describe our initial experience with the first patients.
Methods: A series of bench side tests was made. We dilated small stents which we routinely use in children above the rated diameters until ultimately fracturing them. The diameters and pressures needed to fracture these stents were noted. The concept of intentional stent fracturing was used to treat a pilot series of patients. Clinical and interventional details are presented.
Results: Bench side testing of coronary stents (Coroflex blue, Onyx, Bentley coronary) and of the different sized Cook Formula stents confirmed that they all can be fractured. Useful strategy to prevent the development of a "napkin ring" was to implant a second larger stent before using dilatation with ultra high-pressure balloons. This was implemented in the treatment strategy of the first patients.
In seven patients, previously implanted stents were dilated serially and ultimately fractured. The fractured stents were Coroflex blue (n = 3), Bentley coronary (n = 2) and Cook Formula (n = 2). The stents were implanted in branch pulmonary arteries (n = 5), in the right ventricular outflow tract (n = 1) and in the aortic arch (n = 1). After dilation up to the fracturing diameter known from the bench

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side tests, a second larger stent was implanted and the initial stent was fractured with ultra high-pressure dilatation up to 40 bar. Fracturing of the stent was possible in all patients. In one patient a transient dissection of the left pulmonary artery without clinical consequences occurred. No other complications were noted.
Conclusion: Serial dilatation and intentional fracturing of stents in growing children is possible in the clinical setting. In our initial patient series no serious complications occurred. Serial dilatation and intentional fracturing may play an important role in the management of growing children with congenital heart defects.

Keywords: stent fracturing, stent implanation, children

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Interventional catheterization for congenital heart disease: outcomes in a developing country

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Background and Aim: Interventional catheterization (IC) for congenital heart disease (CHD) requires extensive technical resources which can be challenging in low- income countries. We report the results of IC for CHD in Sudan.
Methods: Retrospective study including patients who underwent IC at 2 Sudanese Cardiac Centers by one operator from 2004 to 2021. Procedures performed by visiting teams were excluded. Procedure type, immediate outcomes and long-term outcomes are described.
Results: in the study period, 1500 cardiac catheterization procedures were performed including 670 IC, in the last 5 years the rate of IC increased from 44 to 55%. Of these 470 were performed by the author. The age ranged from 1 day to 65 years, adults constituted 12%. IC procedures included ductus arteriosus (DA) closure (188 patients, 40%), pulmonary valve (PV) balloon dilatation (141 patients, 30%), balloon atrial septostomy (BAS) (33 patients, 7 %) balloon dilatation of aortic valve (AV) (22 patients, 5%) and atrial septal defect (ASD) closure (20 patients, 4%). Procedure's success rates were 99% for DA closure, 90% for PV dilatation, 83% for ASD closure, 82% for AV dilatation and 90% for BAS. Other procedures include balloon dilatation of re coarctation, right ventricle outflow tract and PDA stenting and temporary pacing.
Procedure related complications included one mortality a 3 months old patient with PV stenosis, device embolization in 4 patients with PDA (3 retrieved and one needed surgery), and one with ASD who needed surgery. Limitations included shortage of catheter supplies, the high procedure cost and limitations of intensive care and surgical facilities. Follow up of patients who underwent therapeutic procedures showed that 95% are asymptomatic with no residual lesions. Of those who underwent palliative procedures, 30% did not have the definitive surgery because of financial or technical limitations.
Conclusion: IC can be established in resource-limited centers by collaboration with regional centers and gradual scaling of case complexity. Innovative approaches to overcome shortage of supplies are needed. Surgical services in this setting still needs to be promoted.

Keywords: Interventional, catheterization, Sudan

P-200 / Moderated Poster

Importance of RVOT gradient in the diagnosis of transcatheter pulmonary valve endocarditis

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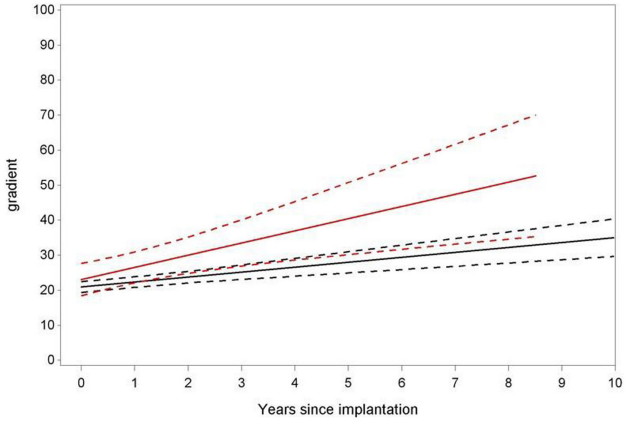
Background and Aim: Infective endocarditis (IE) is a threat for the transcatheter pulmonary valve (TPV) and risk factor for valve failure. Increased RVOT gradient is often observed at the time of IE. The aim was to study if an increased RVOT gradient can play a role in the diagnosis of TPV IE.
Methods: Single center retrospective analysis of patients with TPV with the Melody™ valve implantation between 2006 and 2019. Serial transthoracic echocardiographic data were analysed in all patients after TPV. The evolution of the gradient was compared between patients with and without endocarditis using linear mixed models.
Results: A total of 201 Melody™ valves were implanted in 197 patients at a mean age of 20.5 years (3.6 - 81.6). IE occurred in 22 patients (11.2%) after a mean period of 3.2 (0.7 – 10.3) years. Freedom of IE was 86.4% after 5 years. Cox regression analysis with the gradient as a time-varying predictor for endocarditis showed a HR 1.060 (CI 1.030;1.090) $p < 0.001$. Younger age at TPV replacement showed an increased tendency to develop IE in the study group HR 0.987 (CI 0.949;1.026) p 0.500.
Conclusion: An increased RVOT gradient might play an important role in the diagnosis of TPV IE, in addition to the modified Duke criteria.

Keywords: Melody valve, infective endocarditis, RVOT gradient

gradient as predictor



Slope for patients with endocarditis is indicated by a red line, patients without endocarditis with a black line. Dashed lines refer to the pointwise 95% confidence interval

visual presentation of evolution of gradient comparison between IE and non IE

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P-201

The conduit in a fontan circuit frequently becomes restrictive during growth but can safely be expanded beyond nominal size

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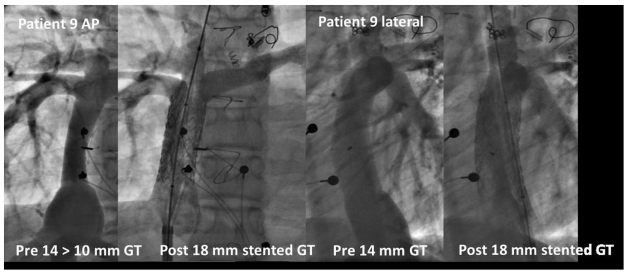
Background and Aim: When connecting the ICV to the pulmonary artery in a TCPC, the largest tube reasonable for that age will be inserted. With time this conduit may become restrictive due to compression, shrinkage, kinking, protrusion of fenestration closure device, peal and somatic growth; this restriction will increase venous congestion and decrease output both at rest and during exercise.
Methods: Fontan patients were assessed by catheterization towards the end of growth or when symptomatic. The conduit was expanded with a balloon mounted stent when the conduit was below nominal value, a residual fenestration needed to be closed, or when the IVC-conduit was disproportional; absence of a gradient was no contra-indication. A stent was delivered at low pressure for reexpansion, but pressures up to 20-28 atmospheres were used to reach a final diameter of 18-20 mm.
Results: Fourty-four patients were treated. Age at Fontan 4.5 ± 1.5y (range 2.1-15.7; 1 reconversion at 35.0y). Conduits: 2 homografts (14-16 mm), 42 GoreTex Stretch vascular graft (17.6 ± 1.7mm, range 14-22, median 18mm). After 12.0 ± 3.2y (3.6-27.1, median 11.8y) the conduit had narrowed to 13.5 ± 1.8mm (9.0-18.0, median 14mm); the conduit was expanded to 19.2 ± 1.2mm (16-22mm, median 20mm) with a single (39) or double stent (5) (Covered CP n = 23, Andrastent XXL n = 25, Optimus XXL n = 2). We aimed to cover from the pulmonary anastomosis till just beyond the caudal ICV anastomosis, avoiding the possibility of late stent compression by the liver. No contrast extravasation was observed; no complications occurred except in 1 patient where the SCV was inadvertently dilated (transient sinus arrest-AVB). The cross sectional surface had increased 217 ± 57% (range 112-484, median 204%). In 6 patients the left pulmonary artery was also stented. All patients remained on their usual anti-aggregation (ASA 1-2 mg/kg/d). 32 patients reported an improved exercise tolerance (no formal testing). With a follow-up of 4.2 ± 2.5y (0.3-14.9, median 3.3 years) no complications occurred.
Conclusion: A Fontan conduit is frequently more restrictive than intended. The conduit can be reexpanded to nominal value, but also dilated up to 18-22 mm without complications. Further follow-up is required to determine if this will result in prolonged enhanced exercise tolerance, decreased congestion and/or slowing of ventricular stiffening.

Keywords: Fontan, Interventional Cardiology, stent

Stent expansion of Fontan conduit.



Röntgen views of stent expansion of a 14 mm GoreTex graft to 18mm (AP & lateral, before & after).

P-202

Transseptal puncture in children under 20 kg

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Background and Aim: Transseptal puncture(TP) is a widely utilized technique for left-sided electrophysiological studies, interventions in left heart lesions or creating interatrial shunts in congenital heart diseases. Although TP is a risky and challenging procedure in young children, it can be performed safely in experienced centers. This study aims to evaluate the outcome of TP in children under 20 kg.
Methods: Patients under 20 kg who underwent TP in our center between December 2015-December 2021 were included in the study retrospectively. The clinical and demographic characteristics of the patients, TP procedure, procedure and fluoroscopy times, and complications were recorded. 3D mapping (EnSite Precision, St. Jude Medical, St.Paul, MN, USA) was routinely used in patients undergoing TP for catheter ablation. TPs were performed with a biplane angiography system in the catheter angiography laboratory. A Brockenbrough needle (BRK Transseptal Needle; Abbott/St.Jude Medical, Inc) was used for TP. In patients whose Brockenbrough needle could not achieve sufficient patency, TP was performed by applying cautery energy over the Brockenbrough needle.
Results: A total of 21 patients were included in this study. Eight of them were girls. The mean age of the patients was 3.4 years (0 days-8.4 years) .9/21 of these patients had a bodyweight of less than 10 kg, and the remaining 12/21 patients were between 10-20 kg (mean 11 kg (2.7-20kg)). The median procedure and fluoroscopy times were 140 min and 3 min, respectively. TP was performed in 7 patients during an electrophysiological study due to SVT.TP was performed in 7 patients with hypoplastic left heart syndrome to

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create an interatrial shunt. In 7 patients, TP was performed to create a right-left shunt due to pulmonary hypertension.TP was not successful in 4 patients. The mean weight of the patients who failed TP was 4.5 kg. The only major complication was pericardial tamponade developed in a patient diagnosed with complex congenital heart disease(CHD).
Conclusion: In conclusion, transseptal puncture can be performed safely in suitable patients. It may be riskier in complex CHD patients with abnormal cardiac anatomy than patients with normal cardiac anatomy performed for the electrophysiological study.

Keywords: Transseptal puncture, children, congenital heart disease, electrophysiological study

P-203
Rescrewing the embolized duct occluder for device retrieval

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Background and Aim: Transcatheter device occlusion became the standard treatment of choice in patent ductus arteriosus (PDA). Rarely, device disposition may lead to embolization. Catheter retrieval of foreign bodies is a well-established procedure. We reported device retrieval by rescrewing of embolized duct occluder (DO).
Methods: A 10-year and eight-month-old girl with Down syndrome was diagnosed with a large PDA with a left to right shunting, secundum type atrial septal defect and pulmonary hypertension. Catheter angiography showed a type A PDA, measured 8 mm at its narrowest point. After test occlusion for pulmonary hypertension, PDA was embolized using Amplatzer Duct Occluder I 12/10 mm in a standard manner. Control angiogram showed an optimally positioned device in the ampulla with no residual shunt. Transthoracic echocardiography on the following day showed patent arterial duct and embolized device. Fluoroscopy confirmed the embolized device position in the descending aorta. The retention screw was facing caudally, enabling us to re-screw quickly. After rescrewing, the device was withdrawn to the 7F guiding catheter. Subsequently, PDA was embolized by Amplatzer Duct Occluder I 16/14 mm.
Results: Device embolization rarely occurs after transcatheter PDA closure. Snares and bioptomes are the most common materials for device retrieval. Rescrewing might be an alternative method in some cases.
Conclusion: Embolized device retrieval by rescrewing method might be a safe and rapid alternative in cases with favorable device position.

Keywords: patent ductus arteriosus, duct occluder, device embolization, device retrieval.

P-204
Interstage interventions after the norwood procedure for hypoplastic left heart syndrome and variants

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Background and Aim: Surgical mortality for the Norwood procedure remains high. In addition there is a significant inter-stage mortality prior to the bidirectional Glenn procedure (BDG).In terms of resource utilization, the Norwood procedure is associated with the highest costs. We analysed the requirement for (unplanned) interstage catheter interventions in a single institutional cohort undergoing close clinical follow-up.
Methods: We analysed the follow-up data of 94 consecutive patients surviving the Norwood/ Sano procedure, between January 2003 and December 2019. The chief diagnosis was hypoplastic left heart syndrome. Other univentricular variants with systemic outflow obstruction were also included.
Results: Sixty two patients (66%) required an interstage reintervention. These consisted of treatment for recoarctation (n = 44; balloon dilation or stent implantation), branch pulmonary artery (PA) stenoses (n = 17; balloon angioplasty), or Sano shunt stenosis (n = 14; stent implantation). Intervention at more than one site was required in a subset of patients. In addition, reintervention at the same site (repeat dilation or stent implantion) was also required in some patients. Minimum aortic arch diameter (pre- versus post treatment) increased from median 3.1 (2.3 to 3.3) mm to 5.1 (4.2 to 6.2) mm ($p<0.001$). Correspondingly the pull-back gradient decreased from 40 (36 to 46) to 9 (5 to 10) mm Hg ($p<0.001$), and the echo gradient from 54 (45 to 64) to 12 (10 to 16) mm Hg ($p<0.001$). The branch PA diameters increased from 2.4 (2.1 to 3.0) to 4.7 (4.2 to 5.1) mm Hg ($p<0.001$). Minimum Sano shunt diameters increased from 2.0 (1.5 to 2.1) to 5.9 (5.8 to 6.0) mm ($p<0.001$); this was associated with an improvement in systemic saturation from 63 (60 to 65) to 80 (79 to 82) %; ($p<0.001$). Unexpected interstage death at home occurred in two patients; no autopsy data were available. The remainder achieved a BDG palliation.
Conclusion: Close follow-up, including home saturation monitoring and routine two weekly outpatient visits identify the majority of infants requiring interstage catheter interventions. The proportion of patients requiring (unplanned) interventions to successfully achieve a BDG remains high.

Keywords: Norwood, catheter interventions, outcomes

Arrhythmia / Electrophysiology

P-205
Timothy syndrome and anaesthesia: a case of extreme QT prolongation

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Background and Aim: Timothy syndrome is an extremely rare multisystem disorder. It is caused by a de novo mutation in exon 8A (type 1) and exon 8 (type 2) of the CACNA1C gene. Clinical features include long QT, syndactyly and congenital heart defects. Patients are at high risk of malignant arrhythmias during anaesthesia. We present the case of a type 1 Timothy Syndrome (TS1) patient who developed extreme QT prolongation with 2:1 atrioventricular block (AVB) during anaesthesia.
Methods: Case Report
A five-year-old, 17 kg, patient with TS1 required revision of a chest wound. TS1 had been diagnosed in the neonatal period. At 8 weeks old she was resuscitated from a ventricular fibrillation arrest following anaesthesia with inhalational agents. An epicardial pacemaker was placed at 3 months of age. She was managed medically with Beta-blockers and Mexiletine for the first 4 years of life, when implantation of a hybrid implantable cardioverter-defibrillator system with a transvenous sensing/pacing lead and a subcutaneous coil, seemed appropriate.

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On this occasion general anaesthesia was required again: two Paediatric Cardiac Anaesthetists attended and an external defibrillator was ready. An inhalational induction was performed with Sevoflurane and Nitrous Oxide, followed by Isoflurane for maintenance of anaesthesia. Intra-operatively extreme QT prolongation became evident, resulting in 2:1 AVB and ventricular rates as low as 40 beat per minute (BPM). In the setting of a maximal QTc measured at 780ms and very high voltage T waves, the internal pacemaker started to over-sense the T waves. Reducing the sensitivity as low as 1.2mV and maximization of the refractory period window of the pacemaker did not troubleshoot the situation. Finally the ventricular rate was reduced to 50 BPM to avoid pacing on the T wave. Resulting hypotension was managed with Metaraminol. As the anaesthetic agents cleared, the QT shortened and satisfactory pacing was achieved. The patient underwent an otherwise uneventful wound revision and recovered from anaesthesia without suffering from ventricular arrhythmias.
Results:
Conclusion: The Timothy population is at high risk of significant cardiac events in the perioperative period. Careful multidisciplinary pre-operative planning can help reduce the risk of perioperative ventricular arrhythmias.

Keywords: Timothy syndrome, Long QT, Anaesthesia

P-206
The effects of percutaneous pulmonary valve implantation on electrocardiographic changes

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Background and Aim: Percutaneous pulmonary valve implantation (PPVI) has become a safe and feasible alternative treatment option for some congenital heart disease patients with right ventricular outflow tract dysfunction. The effects of PPVI on electrocardiographic changes remain unknown. This study aims to evaluate the pre-post procedural ECG changes and arrhythmia burden.
Methods: Between January 2015 and March 2020, PPVI procedures were performed on 129 patients. The clinical and demographic characteristics of the patients, procedure details, 12-lead ECG, Holter ECG, and follow-up data were recorded retrospectively using the hospital database. Because of missing follow-up data, 127 of them were included in the study.
Results: A total of 127 patients, 55 female (43%) and 72 male (%57), were included in this study. The mean age was 18,5 years (5,8-63 years). The primary underlying diagnosis was Tetralogy of Fallot (n = 100/127). %59,8 of the patients had pulmonary regurgitation, %7,9 had pulmonary stenosis, and %32,3 had mixed lesions. The mean pre-PPVI QRS interval was 141,8ms (74-220ms), and the post-PPVI QRS interval was 140,6ms (80-200ms). The mean corrected QT interval was 429,1ms (352-480ms). There was no difference between pre-PPVI and post-PPVI results of these parameters. The mean pre-PPVI QT dispersion was 30,5 (10-130), and post-PPVI immediately it was 27,4 (4-83), and at last control, QT dispersion was 24,7 (5-100). The QT dispersion was shortened post-PPVI immediately and shortened significantly at follow-up (P = 0,04; P = 0,001 respectively).
Conclusion: PPVI shortens the QT dispersion, which indicates the inhomogeneity of ventricular repolarization and may be helpful to prevent malignant arrhythmias in this way.

Keywords: Percutaneous pulmonary valve implantation, arrhythmia, electrocardiography, QT dispersion

P-207 / Moderated Poster
Two broken pacemaker CANS in one patient

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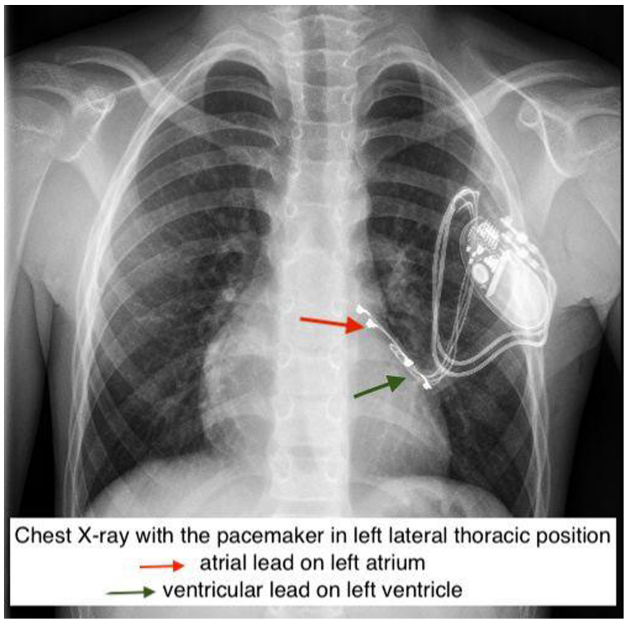
Background and Aim: Hardware problems of pacemaker generators are very rare, occurring in about one in 1000 person/years. However, in pacemaker dependent patients they could be life threatening.
We report on a boy experiencing pacemaker fractures resulting in pacemaker dysfunctions.
Methods: After elective surgery for VSD-closure at the age of six months a complete atrioventricular block persisted and a pacemaker was implanted via left lateral thoracotomy with epicardial leads (left atrium and left ventricle) at postoperative day nine.
At the age of seven years the pacemaker was exchanged due to battery depletion with an AssurityTM MRI (Abbott), placed at the same position and attached to the rib with few stitches. Postoperative course and subsequent outpatient visits were uneventful and the patient asymptomatic.
On regular outpatient visit 18 months later ECG showed no pacemaker activity but junctional escape rhythm at 45bpm. No contact with the pacemaker could be established with the programmer; chest X-ray revealed no visible damage. On revision, no damage of the pacemaker and leads could be identified; measurements for sensing, capture thresholds and impedances were excellent. Postoperative course and outpatient visits again were uneventful. But 17 months later, the second pacemaker (also AssurityTM MRI, Abbott) was again undetectable with telemetry. Another pacemaker replacement was performed successfully; the leads were intact again.
Results: The cross-section of the explanted devices revealed a small cracked area in the titanium case directly under the header displacing the case inward. This crack allowed fluid ingress resulting in circuit damage. Scanning electron microscope confirmed a main and multiple small cracks, clearly illustrating the brittle nature.
Conclusion: According to our knowledge this is the first report of pacemaker dysfunction due to cracks in the can. We hypothesize that the rupture was caused by a combination of: (1) embrittlement of the titanium case in the region of the crack and (2) excessive mechanical stress at the implantation site on the lateral chest wall in a very active child.
To improve safety it is mandatory (1) to improve the material properties of the pacemaker can and (2) to optimize the surgical technique for positioning and fixation, especially in epicardial pacemaker systems in children.

Keywords: pediatric, pacemaker, epicardial, dysfunction,

Chest x-ray



P-208

Arrhythmia in a teenager with a congenital heart defect

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Background and Aim: Cardiac arrhythmias in children most often occur in patients with congenital heart diseases who have undergone surgery. Supraventricular arrhythmias are the most common in the paediatric population. They can be very difficult to treat.
Methods: A case study of a 13-year-old patient with chronic heart failure has been carried out.
Results: The patient underwent a closure of atrial septal defect surgery by occlusion device (Amplatzer) in 2011. Since the surgery she has had multiple attacks of supraventricular and ventricular tachycardia. There is a family history of dilated cardiomyopathy. In 2018 she had a cardiac arrest and implantation of the endocavitary cardioverter defibrillator system (ICD) in 2018. Then, in secondary prevention, left-sided sympathicotomy Th2-Th5 was performed. In 2020, during electrophysiology studies, atrial fibrillation isolated within the left atrium was registered. One year after ICD implantation, arrhythmias clearly intensified, attacks of tachycardia interrupted by adequate ICD discharges persisted, symptoms of palpitations and fatigue were not responding to pharmacological treatment. Currently, the anti-arrhythmic treatment has been modified. That treatment improved the contraction of the heart muscle and decreased the number of tachycardia. Unfortunately, the patient developed anxiety and depression disorders. Currently, the patient is monitored 24 hours a day by Event-Holter telemetry device.
Conclusion: Although most of arrhythmias in children are mild, it is always important to remember about the presence of life-threatening arrhythmias that are not amenable to pharmacological and interventional treatment. There are cases of patients in which the only therapeutic option is implantation of an ICD-defibrillator cardioverter in conjunction with pharmacological treatment.
Solutions such as cardiac telemonitoring with the use of devices intended for home use include the following benefits: better health and greater safety of patients, fewer scheduled specialist visits, savings on procedures and additional funds in the budget of the public payer. Patients with arrhythmia might require multidisciplinary care, including a psychologist.

Keywords: arrhythmias, atrial septal defect, cardioverter defibrillator

P-209

Cold water face immersion and the heart rate response in children and adolescent with type 1 long QT syndrome

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Background and Aim: Patients with type 1 Long QT syndrome (LQTS) have a greater risk of developing arrhythmias during physical activity, particularly swimming and diving. The diving reflex is activated when submersing the face in cold water. This powerful autonomic reflex leads to temporary breathing cessation, bradycardia and an increased peripheral vasoconstriction. The objective of this study was to evaluate the cardiac autonomic response during face immersion in children with congenital type 1 LQTS compared to healthy age- and sex-matched controls.
Methods: 18 subjects were included (age range 7-19 years, mean 11 years): 9 children with genotype-verified type 1 LQTS, and 9 one-to-one age- and sex-matched healthy controls. All LQTS patients were on betablocker medication. After 3 minutes semi-supine resting, the participants performed face immersion, to the level of the ears, in 10 degree Celcius (10°C) water for as long as they were able or maximum 25 seconds. All included participants performed face immersion for more than 10 seconds. Heart rate (HR) data from a single channel ECG recorder (Actiwave Cardio, CamNtech Ltd, UK) was resampled at 4 Hz after cubic spline interpolation. Data was presented as group means and standard error of the mean (SEM). The change in heart rate after face immersion was analyzed by repeated measures analysis of variance. Group comparisons t-tests were performed at specific timepoints.
Results: Figure 1 shows the heart rate response during the first 10 seconds of face immersion. Initial heart rate was higher in controls than in LQTS patients (118 bpm vs 104 bpm, $p = 0.005$), but no significant difference was found after 10 seconds (64 bpm vs 62 bpm, $p = 0.78$). A significant interaction was found between group and time ($p<0.001$) revealing that heart rate decrease was slower in LQTS patients (-2.37 bpm/s) compared to controls (-3.72 bpm/s).
Conclusion: Our study showed an aberrant heart rate response during face immersion in cold water (10°C) for these LQTS patients. The slower heart rate decline indicates weaker parasympathetic activation which may be the result of an autonomic imbalance.

Keywords: Long QT syndrome, Heart rate, Autonomic, Arrhythmia, Diving reflex

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Face immersion

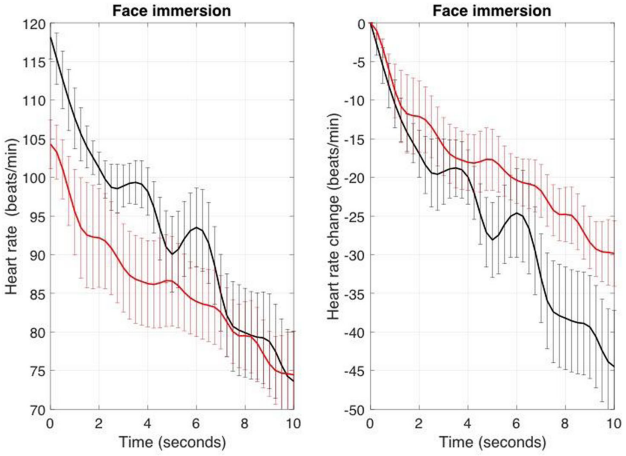

Figure 1. Heart rate changes in LQTS patients (red) and controls (black) in the first 10 s during face immersion. Data are mean ± SEM.

P-210

Risk prediction in pediatric HCM with the ECG risk-score - an inter-observer reproducibility study

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Background and Aim: Hypertrophic cardiomyopathy (HCM) is the leading cause of sudden cardiac death (SCD) in children and young adults. Available risk prediction algorithms for childhood-HCM do not include ECG-parameters. An ECG-risk-score >5p has been shown to detect pediatric HCM patients with subsequent cardiac arrest with high sensitivity, and our goal was to study the inter-observer reproducibility of the ECG risk-score in pediatric HCM.
Methods: A sub-set of 100 different ECGs from pediatric HCM-patients were coded and sent for blind analysis to an experienced pediatric cardiologist who described the method and to a fellow in pediatric cardiology who had never used the method previously. The ECGs were analysed according to the ECG risk-score assessing QRS-electrical axis, voltage-amplitude, duration, ST/T-wave abnormalities and QTc duration. The results were compared with a Bland-Altman plot.
Results: There was excellent agreement in limb-lead voltage-measurements, with only 1% mean difference between observers, 8.06 mV [IQR 5.86-9.80 mV] versus 8.03 mV [5.83-9.86]. Inter-observer variability of total ECG risk-scores ranging between 0-14 points was also very low as shown by Figure 1. There was no significant bias of either observer scoring higher than the other, and median difference of ECG risk-score was 0. There was no inter-observer discrepancy whatsoever in ECG risk-score when analyzing digital ECGs. In old non-digital ECGs there was exact agreement in number of points scored in 82%, with maximum difference of one point in the remainder, which in no case alter patient assignation from low to high-risk group or vice versa. The discrepancies arose mainly in two situations with non-digital ECGs: Firstly in ECGs with large voltage amplitudes overlapping each other over several leads making the calculation of exact limb-lead amplitude sum difficult, with resulting sum from one observer just below, and from the other just above, a scoring cut-off. Secondly if a flatly inverted T-wave reached -1mV or not, giving 1 or 0 points.
Conclusion: The very low inter-observer variability, zero in digital ECGs, makes the ECG-risk-score an easily available tool for initial risk-stratification of pediatric HCM-patients.

Keywords: ECG-risk-score, inter-observer variability, risk stratification HCM

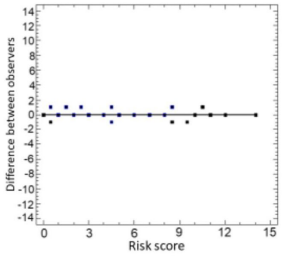

Figure 1 Bland-Altman plot of inter-observer variability in ECG risk-scores.

P-211

Younger postnatal age is associated with a lower heart rate on holter monitoring in healthy newborns

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Background and Aim: Ambulatory 24-hour Holter monitoring is the standard test in arrhythmia screening in all age groups. In newborns, the normal values for Holter parameters are still based on studies executed in 1980's, and the normal limits for heart rates varies between these studies. In the present study, our aim was to set the basis for new reference values for Holter parameters in newborns.
Methods: Healthy term newborns (≥ 37 0/7 weeks of gestation) born to healthy mothers were prospectively recruited from maternity wards of Tampere University Hospital between December 2018 and April 2021. In total, 70 newborns and their Holter recordings included in the analysis. The association of heart rate with age and sex was assessed by linear regression analysis.
Results: The mean (SD) age at the start of the monitoring was 6 (2) days (range: 2.7–9.5 days). The means (SD) for mean, minimum and maximum heart rates were 141 (10), 88 (12) and 208 (13) beats per minute (bpm), respectively. In the linear regression analysis, the mean and minimum heart rate were positively associated with age in days ($p < .001$). Each consecutive day of age raised the mean and minimum heart rate by 4 bpm (95% CI: 2.9, 5.2) and 4 bpm (95%

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CI: 2.4, 5.2), respectively. The effect of age on mean and minimum heart rate was strongest in children aged 3–6 days: one day raised the mean heart rate by 8 bpm (95% CI: 3.7, 12.1) and minimum heart rate by 8 bpm (95% CI: 2.4, 13.5). The minimum – but not mean – heart rate was positively associated with female sex ($p = .047$): the effect size of sex on minimum heart rate was 5 bpm (95% CI: 0.1, 9.4).

Conclusion: The present study shows lower heart rates in the youngest newborns. Larger sample sizes are needed to calculate the age-specific reference values for heart rate in newborns.

Keywords: heart rate, Holter, newborn

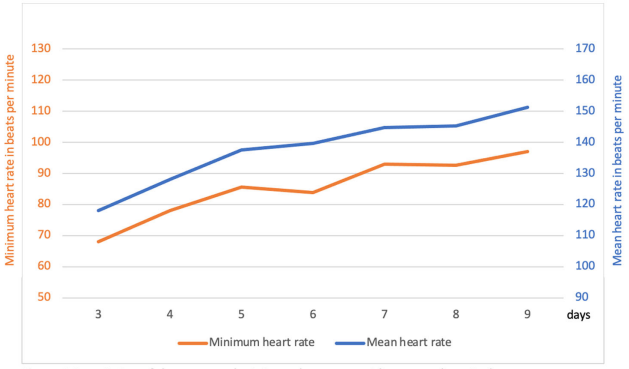

Figure 1 Association of the mean and minimum heart rate with postnatal age in days

P-212

Open-window mapping helps to visualize the location of accessory pathways in children

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Background and Aim: Accessory pathway mapping is currently based on point-by-point mapping, which is a time-consuming and operator-dependent process. With Open-window mapping (OWM), an automated high-density mapping system using a 16-electrode mapping catheter and an automated detection algorithm, atrial and ventricular signals are simultaneously recorded. This may lead to better and more precise visualization of accessory pathways, which may be helpful, especially in cases of broad or complex accessory pathways. Up to now, data especially in children is very limited.

Methods: We studied three consecutive subjects with symptomatic Wolff-Parkinson-White syndrome (WPW). Patients (3f) were 14, 13 and eleven years of age, with the diagnosis of one right-anterior and two right-posteroseptal accessory pathways. After completion of OWM as an additional diagnostic tool, cooled ablation was performed with standard ablation catheter. The successful site of ablation was determined by the loss of pathway function.

Results: OWM was 100% effective at predicting the site of ablation. All pathways were promptly and persistently eliminated at the location site, which have been visualized by OWM.

Conclusion: OWM, using objectively achieved data, is capable of correct direct visualization of accessory pathways. This non-operator dependent technique may be helpful to rapidly map and ablate accessory pathways also in children.

Keywords: arrhythmia, electrophysiology, ablation, accessory pathways, Wolff-Parkinson-White syndrome, mapping

P-213

Utilisation of home monitoring in children with cardiac devices in the COVID-19 ERA

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Background and Aim: The covid-19 pandemic created huge challenges for healthcare worldwide, in the UK reiterating the populations need for an efficient national health system. With a device implantation rate of 1700/million population (UK), regular monitoring is a significant undertaking. We focus on a single tertiary cardiac centre for children, serving an 8 million population, and their ability to deliver such a service at one of the most challenging times in the history of the NHS.

Methods: We retrospectively gathered data for the period April 2020 - April 2021 for all children under our service with; permanent pacing system (PPM), implantable defibrillator (ICD), loop recorder (LINQ). Data was gathered from our secure cardiac database including demographics, device particulars, settings and longevity. First hand accounts from families were gathered by one clinician using a standardized structured set of questions. The responses were tabulated to reveal common themes and key words.

Results: Pre-pandemic 122 patients had devices, with 42 (34%) having monitoring capability at home. As of April 2021 136 patients had devices in situ, with 87 (64%) on home monitoring. See table for data.

Conclusion: Early in the pandemic parents of children with cardiac conditions were justifiably anxious, unclear as to whether their child was at an increased risk given their chronic condition. More complex children were asked to shield initially which meant their medical appointments became impractical. At a time when new ways of working became a necessity as opposed to a novelty our centre utilised the technology available to keep children and their families out of hospital. Home monitoring providing autonomy for our patients and families, and a link to those caring for them at a time of ongoing uncertainty. However there were limitations, 6% of our PPM systems don't have a compatible home monitoring system and those with poor network signal in the home aren't eligible. Furthermore demand for home monitoring has rocketed, which device companies have struggled to meet meaning we must prioritise our most complex and high risk patients.

Keywords: paediatric, cardiology, pacemaker, defibrillator, device, monitoring

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99	19.5	Machine automated	
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Requiring clinician attention = 82	16%		

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P-215

The effect of anaesthetic drugs on the inducibility of avnrt and eat in paediatric patients during electrophysiological studies

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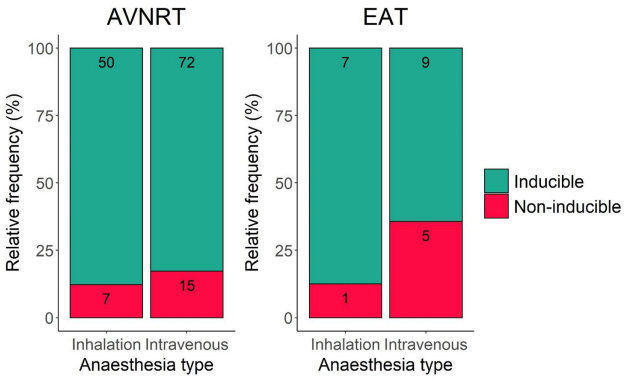
Background and Aim: In paediatric patients, invasive electrophysiological studies (EPS) and radiofrequency catheter ablations (RFA) of supraventricular tachycardia (SVT) are often performed under general anaesthesia. In patients with atrioventricular nodal reentrant tachycardia (AVNRT) and ectopic atrial tachycardia (EAT), reliable diagnosis and subsequent therapy is not possible in sinus rhythm, and tachycardia must be inducible during EPS. This study aims to assess the problem of non-inducible AVNRT and EAT during EPS under general anaesthesia.
Methods: In a monocentric retrospective study, anaesthesia protocols of 166 patients undergoing EPS were analysed in terms of substances applied before and during the first attempt to induce tachycardia.
Results: In 122 patients, an AVNRT was found during EPS. They were compared to 22 whose tachycardia was not inducible but most probably due to an AVNRT mechanism. Another 16 patients with inducible EAT were compared to 6 whose EAT appeared on surface ECG but not during EPS. Demographic characteristics were similar among all groups. Inducibility did not differ ($p = 0.42$) between AVNRT patients with inhalational anaesthesia (sevoflurane and/or nitrous oxide) and patients with intravenous anaesthesia (propofol with/without remifentanil). Under intravenous anaesthesia, AVNRT inducibility tends to be increased when propofol is used alone (88%) compared to its combination with remifentanil (76 %, $p = 0.16$). Lidocaine, applied to reduce pain during initiation of propofol infusion, was administered with similar frequency in inducible and non-inducible AVNRT patients ($p = 0.41$). The EAT group exhibited lower inducibility under intravenous anaesthesia (64%) than under inhalational (88%), however without significance ($p = 0.35$). EAT patients under intravenous anaesthesia exhibit a trend towards better inducibility when the propofol infusion rate is reduced by combining it with remifentanil (75%) than when propofol is used alone (50%, $p = 0.58$). All six non-inducible EAT patients were exposed to lidocaine, while seven of the 16 inducible cases were not ($p = 0.10$).
Conclusion: Tachycardia induction succeeds with similar frequency under both inhalational and intravenous general anaesthesia in children with AVNRT. In children with EAT, inhalational anaesthesia is associated with a trend towards better inducibility.

Keywords: Atrioventricular nodal reentrant tachycardia (AVNRT), Ectopic atrial tachycardia (EAT), Anaesthesia, Children, Electrophysiological study

Inducibility rates in AVNRT and EAT patients under inhalational or intravenous anaesthesia



P-216

A retrospective case review of exercise tolerance testing data for children diagnosed with inherited or suspected inherited and acquired arrhythmias

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Background and Aim: Exercise tolerance testing (ETT) is used routinely within paediatric cardiology services. However, there is little evidence pertaining to normal ranges and safety when advising exercise following ETT for children with inherited, suspected inherited and acquired arrhythmias, and particularly for those unable to complete a full ETT. Development of fatigue or symptom onset, resulting in variable lengths of testing or early termination can be a limiting factor. This service evaluation project aims to evaluate current activity and define research needs to support the clinical service by exploring children's physiological responses to controlled ETT using the paediatric arrhythmia protocol – figure 1.
Methods: 108 paediatric patients (male 56), aged 4 – 16 years who completed ETTs between 2015 - 2020 were included in the analysis. A total of 175 ETTs were recorded with 99 ETTs representing repeat testing for 32 children. Children undergoing ETTs were diagnosed with Long QT Syndrome (LQTS), Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT), Supraventricular Tachycardia, Wolff Parkinson White Syndrome or were being investigated for LQTS, CPVT, familial Sudden Arrhythmic Death Syndrome or out of hospital cardiac arrest.
Results: The range in ETT time frame completion was 3.13 – 15 minutes (mean 8.95). 10% of ETTs were terminated in <6 mins.

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Fatigue was the most frequently reported reason for termination 48%; tired/sore legs 15.4%; speed of the treadmill 5%; dizziness 4%; short of breath 4%; ECG leads disconnected 2.9%; reached target heart rate 2.9%; completed the protocol 1.7%. Antiarrhythmic medications including beta-blockers and sodium channel blockers were being taken during 63 (36%) ETT's. Ventricular ectopy occurred at a frequency of 18.3%, mostly in the CPVT cohort. SVT occurred in the recovery period during 2 ETTs (1.1%).
Conclusion: Childhood fatigue and leg discomfort are recognised complaints when running up-hill during exercise testing resulting in early termination of ETT. 161 (92%) ETTs were terminated before protocol completion, with fatigue and tired legs being the greatest cause of ETT termination. There were no adverse events; however, ETTs were terminated for a small number due to equipment failure. Further analysis is required to correlate medications, ETT completion and to further analyse heart rate and rhythm responses.

Keywords: Exercise tolerance testing, children, inherited arrhythmia, acquired arrhythmia

P-217 / Moderated Poster
AVNRT cryoablation in children < 26 kg: efficacy and safety of electrophysiologically guided low voltage bridge strategy

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Background and Aim: Recent studies have reported that the low voltage bridge (LVB) strategy is very effective in atrioventricular nodal reentrant tachycardia (AVNRT) ablation.
The purpose of our study was to evaluate the efficacy and safety of AVNRT cryoablation in pediatric patients < 26 Kg using the electrophysiologically guided LVB strategy both in the acute phase and in a mid-term follow up.
Methods: Thirteen pediatric patients (69% males, mean age 7 ± 1.65 years, mean weight 24.8 ± 1.77 Kg), with previous history of AVNRT confirmed by the transesophageal atrial stimulation, underwent slow pathway cryoablation using the electrophysiologically guided LVB strategy.
Results: In 12 patients a typical AVNRT was induced, while 1 patient showed both a typical and an atypical form. An AV "jump" occurred in 46% of patients. One small LVB was found in 7 patients (54%) at the M1-M2 level, in 4 (31%) at P2 or P2-M1 level, in 1(7.5%) at A2 and another 1 (7.5%) at M1 level. Acute success was 100% with an average of 5 cryoablations per patient. No peri-procedural complications occurred. All procedures were performed with near-zero fluoroscopy. No recurrence occurred during the follow up (10.58 ± 10.23 months).
Conclusion: AVNRT cryoablation using the electrophysiologically guided LVB strategy seems to be an effective and very safe procedure in small children who are the highest risk patients for this type of ablation.

Keywords: Cryoablation, AVNRT, low voltage bridge, pediatric

P-218
Catheter ablation in patients with congenital heart disease: a single centre experience

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Background and Aim: Cardiac arrhythmias are a major concern in patients with congenital heart disease. Catheter ablation is growing as therapeutic option for these patients and can be rather challenging in this group of patients due to the anatomical complexity of the underlying lesion particularly in those who have undergone corrective or palliative surgery. The purpose of this study is to present a single centre experience in catheter ablation arrhythmias in patients with congenital heart disease.
Methods: Patients with congenital heart disease, operated or not, who were submitted to radiofrequency ablation from 01/01/2016 to 31/12/2021 in a tertiary referral centre.
Results: A total of 44 catheter ablations were performed in 36 patients (mean age 43 ±3 years) with congenital heart disease. The vast majority (82%) had been previously submitted to cardiac surgery. Median cycle length of the 43 tachyarrythmias mapped was 330ms (interquartile range 290-400), consisting in 22 cavotricuspid isthmus atrial flutter, 10 atrial re-entrant tachycardia, eight focal atrial tachycardia and three atrioventricular re-entrant tachycardia. In three other cases the ablation was guided by substrate, while in seven cases an atrial fibrillation ablation was performed. No adverse events were recorded. During a median follow up time of 37 months (IQR 12-51), with a 1.3 procedures per patient, freedom from arrhythmia was achieved in 91% of the patients (30% of them maintained antiarrhythmic drugs.
Conclusion: Catheter ablation plays a major role in the treatment of arrhythmias in patients with CHD. The procedure is safe and our results show good outcomes.

Keywords: Catheter ablation, interventional electrophysiology, congenital heart disease

P-219
Impact assessment of premature ventricular contractions on children's cardiac output

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Background and Aim: Premature ventricular contractions (PVCs) are one of the most frequent children's heart rhythm disturbances. However, hemodynamics changes during arrhythmia haven't been studied thoroughly. This work aims to assess stroke volume

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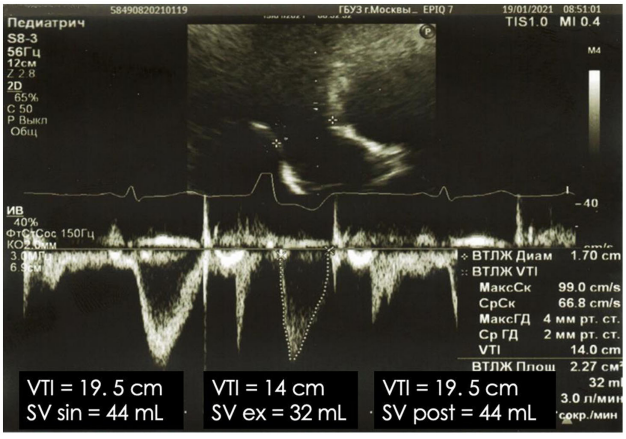
(SV) dynamics and daily cardiac output (CO) among children with PVCs.
Methods: Left ventricular SV was measured by the Doppler VTI method on sinus contraction (SVsin), extrasystole (SVex) and post-extrasystolic contraction (SVpost). Stroke volume deficit (ΔSV) was calculated in each cycle (SVsin+SVex+SVpost) concerning assumed sinus rhythm by following formulas. In a single PVC with compensatory contraction presence was considered: ΔSV1 = 2SVsin – SVex - SVpost; in interpolated PVC compensatory contraction complete absence was considered: ΔSV2 = SVsin – SVex; in PVCs couplets one of compensatory contraction absences was considered: ΔSV3 = 3SVsin – 2SVex – SVpost. Daily cardiac output deficit (ΔCO) for each contractions' variant was calculated as ΔCOn = ΔSVn × (such cycles number). Extrasystoles numbers and variants were determined by 24-hour Holter ECG monitoring.
Results: 54 children (21 female, 33 male) from 8 to 17 y.o. with daily PVC from 2666 to 18238 (PVC burden 2.4 -15.8%) were examined. Other cardiac pathology was excluded, nobody had low LV ejection fraction beyond the arrhythmia. Single PVCs were registered among all 54 patients (100%), couplets – 12 patients (22.2%), interpolated cases – 2 patients (3.7%). 46% have right-sided PVCs, 54% have left-sided PVCs. Average ΔSV1 = 13.1 ± 2 mL, ΔSV2 = 19.7 ± 3 mL, ΔSV3 = 30.1 ± 6 mL. Estimated daily losses: ΔCOmin = 26.6 L, ΔCOmax = 218.8 L. Correlation ΔCO with patient's gender, age and PCV localization are absent.
Conclusion: PVC is a complex problem involving decreased left ventricular filling (diastole defect) with subsequent cardiac output deficiency. The CO deficiency increases with rise in the number of PVC, the absence of compensatory contractions (ΔCO2/ΔCO1 = 1,5; ΔCO3/ΔCO1 = 2,3) or their inefficiency. Last changes may reflect hidden pumping dysfunction of the left ventricle, which can lead to clinical symptoms' appearance and decrease of patients' life quality.

Keywords: premature ventricular contractions, cardiac output

Example of stroke volume measuring during a three contractions cycle in a single PVC.



Patient 14 y.o, single unifocal PVCs arising from the left sinus of Valsalva with 18238 frequency per day (PVC burden 15,8%). SVsin = 44 mL, SVex = 32 mL, SVpost = 44 mL. ΔSV1 = 12 mL; ΔCO1 = 218,8 L.

P-220

Arrhythmia ablation in congenital heart disease patients – a single centre retrospective study

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Background and Aim: Arrhythmias are an important cause of morbidity and mortality in congenital heart disease (CHD) patients. They can be treated by ablation during electrophysiological studies, but are usually complex due to the anatomical variation, presence of structural disease and scar tissue.
Methods: Retrospective review of ablation cases in CHD patients in a single centre from 2001 to 2020. Data on demographics, CHD group, arrhythmia mechanism, treatment outcomes (immediate and long term), and complications was collected.
Results: 53 patients were identified (51% male), who had a total of 77 procedures. Median age at first procedure was 29.8 years (min 0.7, max 65.8). The most prevalent CHD group was right obstructions (25 pts, 47.2%), followed by septal defects (13 pts, 24.5%), left obstructions (4 pts, 7.5%), tricuspid valve malformations (4 pts, 7.5%), functionally univentricular heart post-Fontan operation (4 pts, 7.5%), transposition of the great arteries post-Senning operation (2 pts, 3.8%) and Truncus arteriosus (1 pt, 1.9%). The median number of surgeries was 1 (min 0, max 4), and 12 pts (22.6%) underwent at least one palliative surgery. On the first procedure arrhythmias found were right atrial macroreentrant taquicardia (23 pts, 43.4%), right atrial focal tachycardia (4 pts, 7.5%), left atrial focal tachycardia (1 pt, 1.9%), atrial fibrillation (4 pts, 7.5%), accessory pathway-mediated atrioventricular (AV) reentrant tachycardia (7 pts, 13.2%), AV nodal reentrant tachycardia (4 pts, 7.5%), right ventricular focal tachycardia (3 pts, 5.7%), right ventricular macroreentrant tachycardia (4 pts, 7.5%), presence of substrate for right ventricular macroreentrant tachycardia without clinical manifestations (1 pt, 1.9%), and non-specified atrial tachycardia (2 pts, 3.8%). On first procedure complete success was achieved in 81.1%, partial success occurred in 3.8% and empirical treatment was performed in 1.9% (1 pt); palliative treatment (AV node ablation) was performed in 3.8%; procedure failure occurred in 7.5%. Recurrence of arrhythmia occurred in 39.6%, but of these 28.6% had a different mechanism. 18 patients had at least one repeat procedure, and on the first repeat procedure success was achieved in 72.2%.
Conclusion: CHD patients present a challenging population, but on our series it was possible to treat their arrhythmias, even tough multiple procedures
were frequently necessary.

Keywords: CHD, arrhythmia, ablation

P-221

Atrial fibrillation in pediatric patients: clinical features and treatment strategy

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Background and Aim: To evaluate the clinical features, comorbidities and the treatment strategy of atrial fibrillation (AF) in pediatric patients (pts).
Methods: Children (n = 39) aged 3-17 years (21 females) with idiopathic AF were included into the study. All pts received treatment in one institution. Demographics, clinical presentation, comorbidities and treatment were analyzed.
Results: The AF was paroxysmal in 24 pts (61.5%). Clinical manifestations have been found in 79.5% (n = 31). AF was associated with atrial flutter (AF) in 13 (33.3%) cases, Atrial premature beats (APB) have been found in 23.1% (n = 9). Antiarrhythmic drug therapy (AAD) was administrated in 76.9% (n = 30). Catheter ablation was performed in 6 pts (15.4%) (after failed or intolerant AAD): cryoballon pulmonary vein isolation (PVI) – 5.1% (n = 2), radiofrequency (RF) PVI – 5.1% (n = 2), cryoballon PVI with left atrial roof linear RF ablation – 2.6% (n = 1), AF trigger RF catheter ablation – 2.6% (n = 1).
Conclusion: Pediatric AF involves treatment challenges and high recurrent rate. AF clinical manifestation may have atypical symptoms as well absence of symptoms. AF and APB are common comorbidities and may be potential triggers. Previously potential AF trigger elimination can be considered as optimal approach in pediatrics pts.

Keywords: atrial fibrillation

P-222
Heart rate variability analysis in pediatric patients with duchenne muscular dystrophy

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Background and Aim: Duchenne muscular dystrophy (DMD) is a rare genetic disease leading cardiomyopathy and heart failure due to dystrophin deficiency. Heart rate variability (HRV) abnormalities were previously reported but this has not been extensively studied so far in DMD.
Methods: A prospective, longitudinal, observational study approved by the local ethics committee was conducted. The inclusion criteria were the genetically confirmed diagnosis of DMD. Controls were matched from internal database provided the individual had no significant hemodynamic abnormalities and the specific study was qualified normal by expert reader. Philips-Zymed system was used for recording and analysis. Statistical calculations were done using Wizard 2.0.10 using non-parametric tests considering data distribution. Multiple studies in the same patient were considered separate data points. Heart rate (HR) is given in bpm. HRV parameters were corrected for HR using appropriate exponential regression. Data is given as median (range), $p<0.05$ was considered significant.
Results: Ninety-three DMD patients having 156 Holter ECG studies and 551 controls with 648 studies were included. Patients were 9.6 (0–18) and controls 11.3 (0–18) years old, $p = 0.700$. Patients had faster 24h-average HR 101.5 (76–125) vs 86 (51–152) $p<0.001$, faster minimum HR 63 (41–90) vs 51 (34–112) $p<0.001$, and lower maximum HR 158 (118–211) vs 164 (92–231) $p = 0.005$ thus making the min-max HR span narrower 96 (57–151) vs 108 (46–167), $p<0.001$.
All raw HRV parameters differed significantly between the groups, ASDNN5 50.2 (26.4–232.1) vs 65.3 (11.5–137.1) $p<0.001$, RMSSD 34.3 (14.2–1179.5) vs 52.1 (7.2–325.8) $p<0.001$, SDANN5 78.0 (24.8–644.5) vs 117 (19.9–275.6) $p<0.001$, SDNN 95.2 (37.7–925.9) vs 138.3 (25.3–347.7) $p<0.001$.
After correcting HRV variables for the heart rate the differences were insignificant for ASDNN5c 102.3 (58.2–308.8) vs 109.8 (19.6–206.8) $p = 0.113$ and RMSSDc 79.0 (34.3–420.4) vs 86.9 (14.5–419.0) $p = 0.084$, lesser for SDANN5c 73.6 (31.9–390.3) vs 82.6 (13.8–409.8) $p = 0.033$ and significant only for SDNNc 204.6 (82.2–240.5) vs 235.9 (95.1–442.9) $p<0.001$.
Conclusion: It is of most importance to correct for heart rate when analyzing HRV indices in patients with DMD as they commonly present with sinus tachycardia. After correction the magnitude of HRV abnormalities in DMD is less prominent than previously reported. The clinical consequences of this phenomenon need further investigation.

Keywords: Heart rate variability, sinus tachycardia, Duchenne muscular dystrophy, Holter ECG, cardiomyopathy

P-223
Atrial fibrillation in the pediatric ward: rarely a benign condition

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Background and Aim: Atrial fibrillation (AF) is uncommon in children in absence of CHD. Epidemiological data are lacking. The presence of AF with structurally normal heart, requires careful etiological work-up.
Methods: A 15-years-old girl with Hay-Wells syndrome, was hospitalized for a recent diagnosis of diffuse large B-cell lymphoma. She presented suddenly with thoracic oppression plaints. Monitoring showed a heart rate varying between 200 and 220bpm. On ECG, AF was diagnosed. Previously, she underwent transthoracic echocardiography (TTE) that showed a small atrial septum defect type secundum, further normal. Previous ECG was normal, particularly no pre-excitation or early repolarisation. Blood gas excluded ionic perturbations. Given the hemodynamic tolerance, she was charged with Amiodarone 800mg/m^2 orally. After 6h, heart rate decreased to 180bpm but still no sinus rhythm. She developed pallor, hepatomegaly and blood pressure dropped to 79/43mmHg. TTE showed normal function, completed with a transesophageal echocardiography in order to rule out intracardiac thrombus. We proceeded to electrical conversion, under propofol and ventilatory support. Cardioversion was obtained after one shock of 1J/kg. Cardiac MRI showed tumoral invasion in the lateral wall of the left atrium (LA).
Results: In contrast to adults, with LA dilation and myocardial fibrosis causing LA dysfunction and electromechanical conduction delay, the substrate for AF in children is often different. AF can be an early manifestation of a cardiomyopathy or unrecognised channelopathy, or be associated with any of the genetic and extracardiac causes of AF, as in our case. In absence of other underlying extra-cardiac trigger, such as hypertension, hyperthyroidism, pulmonary embolism, viral infection, sepsis or drug overdose, we considered neoplastic invasion of the LA wall as etiological mechanism of AF.

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Although pharmacological conversion has been observed, we experienced that synchronized electrical cardioversion was the most straightforward procedure to rapidly establish sinus rhythm. There was no recurrence in our case, the oncological condition being treated.

Conclusion: In patients without cardiac condition before onset of AF, its etiology should always be investigated. Oncological conditions are part of the differential diagnosis as we showed in this case report. Cardiac MRI can be a helpful tool. Electrical cardioversion is a rapid and very effective treatment.

Keywords: atrial fibrillation, child, lymphoma

ECG and MRI findings

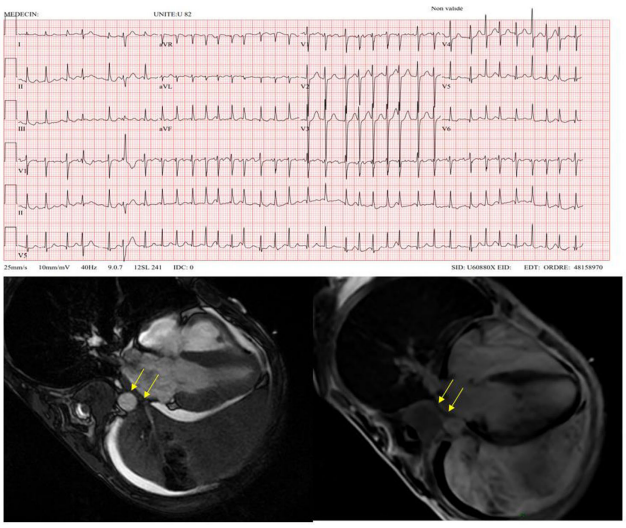

Image 1: Electrocardiogram at onset: atrial fibrillation with rapid ventricular response. Incomplete right bundle branch block secondary to fast heart rate. Image 2: Cardiac MRI Steady-state free precession (SFFP) 4-chambers view showing the extensive tumoral process in the left pulmonary area coming into close contact with the left atrium, with paraneoplasic pleural and pericardial effusions (A); and likely tumoral invasion of the left atrial wall on the LGE acquisition (B).

P-224

Use of a noninvasive continuous monitoring device the nuubo system in the management of selected paediatric group of patients: a pilot study

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Background and Aim: Twelve-lead electrocardiography (ECG) remains the gold standard for the diagnosis of arrhythmias. However, intermittent arrhythmia may not be detected using standard ECG. Standard 24-hour to 14-day Holter monitoring increases the chance of arrhythmia diagnosis, but they are related to the patient's discomfort and his limited motor activity during registration. We would like to present our first experience with Nuubo system in pediatric patients.

Methods: Children the age of 4-17 years with recurrent episodes of palpitations, syncope, with Wolf-Parkinson-White pattern, family history of sudden cardiac arrest or ventricular arrhythmia in athletes were qualified for the study. The registration period, using Nuubo nECG platforms, lasted up to 30 days or until the occurrence of palpitations. All patients before long-term cardiac monitoring had 12-lead ECG and Holter EKG performed.

Results: Twenty-nine patients were enrolled and included in the final analysis. The median age for patients enrolled was 14 years (range, 4-17 years), and 48% of patients were male. Eight of patients (28%) reported at least one episod of symptoms (pallpitations, presyncope or syncope) during the study. Arrhytmia related to the symptoms was revieled in 2 (7%) of patients. Eight of patients (28%) was diagnosed with significant arrhythmia – 5 (17%) were qualified for electrophysiological study, 1 (4%) had modified pharmacotherapy and 2 (7%) were qualified for genetical testing. All the patients found the Nuubo monitoring system comfortable to wear as opposed to 0% for the Holter monitor.

Conclusion: Nuubo nECG platform is useful, non-invasive diagnostic tool of palpitations in children which can replace conventional 12-lead ECG and Holter monitoring.

Keywords: ecg, holter ecg, syncope, pallpitatins

P-225 / Moderated Poster

Catheter ablation of accessory pathways in children and patients with congenital heart disease: predictors of outcome

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Background and Aim: Outcomes of catheter ablation of accessory pathways (AP) in pediatric patients have improved significantly over time. There is still however occasional procedural failure and recurrence of AP conduction. We aimed to review our experience with these procedures and identify predictors of outcome.

Methods: We reviewed our database of pediatric and congenital catheter ablation procedures for demographic, clinical, procedural and outcome data. Procedures were performed from 01/07/1994 to 07/11/2017. Statistical analysis was performed using SPSS for Windows. Pearson's Chi-Square test was used for categorical data, and Mann-Whitney U-test for continuous data.

Results: There were 405 patients (61% male), ages 10,9 ± 11 yrs (0.3-35 years). At baseline, 116 patients had a normal ECG, 276 had preexcitation and 13 had incessant tachycardia. Moderate or complex CHD was present in 26 patients (6.4%). General anesthesia was used in 383 patients (94.5%). Radiofrequency current was used in 380 pts (94%) and cryoablation in the remaining. 3-D mapping ± fluoroscopy was used in 330 pts (81.5%) and fluoroscopy only in the remaining. The AP location was right-free wall (RFW) in 73 (18%), septal (S) in 164 (40.5%), left free wall (LFW) in 146 (36%) and multiple in 22 (5.5%). Mean procedure time was 192.7 ± 82.1 min and fluoroscopy time 11.2 ± 17.6 min. Successful ablation was achieved in 392 procedures (96.8%). Recurrence of AP conduction occurred in 46 patients (11.6%). The procedure and fluoroscopy times were significantly shorter with use of 3D mapping than without (182.3 ± 180 vs 241 ± 220 and 6.3 ± 3.5 vs 32.7 ± 22.5 min respectively, $p < 0.05$). Procedure failure was higher for RFW vs LFW ($p < 0.027$) and for patients with CHD ($p < 0.01$). Recurrence was higher for both RFW vs LFW ($p < 0.015$) and S vs LFW ($p < 0.002$). There was no difference in procedure failure or recurrence between radiofrequency and cryoablation.

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Conclusion: Accessory pathway ablation is highly successful in pediatric and congenital patients. The success and recurrence rates in our experience are consistent with data published in large registries and a prospective study. Location of AP and presence of significant CHD are related to results. Use of 3-D mapping has resulted in significant shortening of the procedure and fluoroscopy times.

Keywords: Accessory pathways, catheter ablation, pediatric, congenital heart disease

P-226

Feasibility and diagnostic value of recording smartwatch ECGs in neonates and children

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Background and Aim: The possibility and diagnostic value of recording smartwatch ECGs independently or with the active participation of an adult in younger children remains to be demonstrated. We sought to evaluate the agreement of smartwatch-derived single-lead ECG recordings with 12-lead ECGs for diagnosing various electrocardiographic abnormalities in neonates and older children.

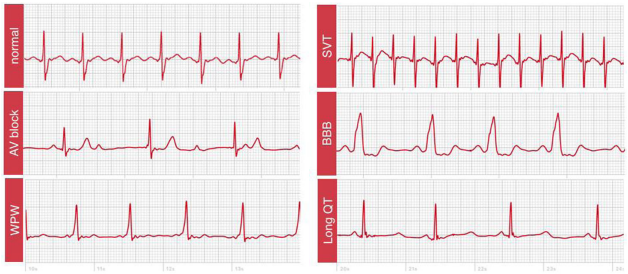
Methods: A 12-lead ECG and an ECG using Apple Watch 4 were obtained in 88 children (from 1 week to 16 years of age) with normal (n = 56) or abnormal (n = 32) 12-leads ECG (AV block n = 5, supra-ventricular tachycardia (SVT) n = 5, bundle branch block n = 12, pre-excitation n = 6, long QT n = 4). In children younger than 6 years old, the ECG-recording was performed with the active participation of an adult who applied the neonate or children's finger to the crown of the watch. In children older than six, recording was performed after brief teaching without adult guidance. All 12-lead ECGs were independently evaluated by two blinded cardiologists. Apple Watch ECGs were independently evaluated by another blinded cardiologist.

Results: In all 88 children, the smartwatch tracing was of sufficient quality for evaluation. All 56 normal tracings were correctly identified. Of the 32 children with characteristic abnormalities identified on 12-lead ECG, 5 (16%) were missed, mostly because of baseline wander and artifacts, resulting in incorrect diagnosis of "normal tracing". Rhythm disorders (AV block or SVT) and bundle branch blocks were correctly detected in most cases (9/10 and 11/12 respectively). The accuracy was less for the diagnosis of pre-excitation or long-QT (5/6 and 2/4 respectively).

Conclusion: ECG smartwatches have the potential to detect clinically relevant conditions in children, with the parental assistance until the age of six.

Keywords: Smartwatches electrocardiograms. Neonates.

Smartwarch ECGs



P-227

Cardiac resynchronization therapy for heart failure associated with left ventricular apical pacing

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Background and Aim: Left ventricular apical pacing (LVAP) has been proven to preserve LV function in children with complete atrioventricular block and structurally normal heart. We aim to present cases of severe LV dyssynchronopathy associated with LVAP and leading to CRT.

Methods: Three pediatric patients (aged 5.3, 11.6 and 17.9 years) with complex structural heart disease, morphologically left systemic ventricle, complete AV block and initially normal LV function developed significant LV dysfunction and heart failure symptoms during epicardial dual-chamber LVAP lasting for preceding 4.9, 3.2 and 7.0 years, resp. They were evaluated clinically and echocardiographically including speckle tracking imaging and upgraded to CRT by implanting additional epicardial leads to left ventricular posterolateral basal free wall and anteroseptal epicardial surface to resynchronize the LV.

Results: The patients presented with a major apical to basal LV electromechanical delay along with signs of classic pattern dyssynchrony (Figure). After CRT upgrade QRS duration and apical to basal LV mechanical delay decreased in all 3 pts. and classic pattern dyssynchrony resolved completely in 2 pts. LV function improved within a period of 3.9 – 5.3 months in 2 of 3 (Pat. No 1 and 2, Table). Pat. No 3 did not improve despite successfull electrical resynchronization.

Conclusion: In a subset of pediatric patients with complex structural heart disease LVAP may lead to LV dyssynchronopathy due to a major apical to basal LV electromechanical delay. Predispositions to such development are poorly understood. CRT aimed at apico-basal LV resynchronization led to major improvement in LV function in 2/3 described pts.

Keywords: heart failure, pacing, cardiac resynchronization therapy

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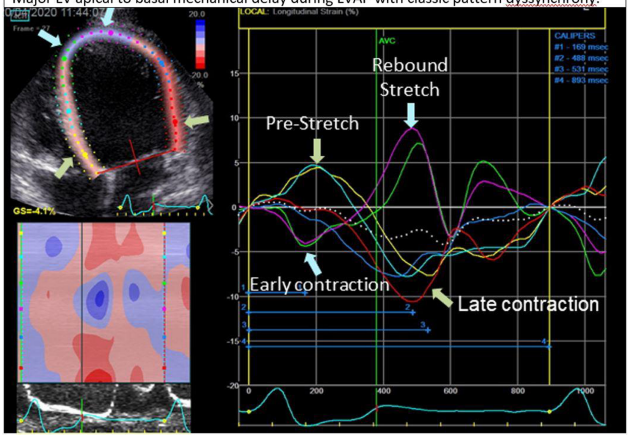
Table and Figure

	Patient No 1		Patient No 2		Patient No 3	
Age at CRT [years]	5.6		11.6		17.9	
CRT response	Before	After	Before	After	Before	After
NYHA class	2	1	2	2	2	2
NTproBNP [ng/L]	610	383	-	191	2906	9179
QRS duration [ms]	120	100	160	140	200	120
LV EF [%]	29	50	44	55	24	26
LV apical to basal mechanical delay [ms]	235	53	123	79	352	174
Major LV apical to basal mechanical delay during LVAP with classic pattern dyssynchrony:						



CRT response and typical signs of apical to basal LV mechanical dyssynchrony

P-228

Syncope in the young elite athletes

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Background and Aim: Background. Syncope in the young athletes is unknown problem in terms risk of sudden cardiac death. Aim of study was to determine the prevalence of syncope among young elite athletes.
Methods: Methods. In the course of the study "EPidemiology of the SyncOpe in chilDren and adolscents in Elite Sport (EPISODE-S)" 1687 young elite athletes 16.3 ± 1.5 years old, members of the young Russian National teams from 52 sports discipline were surveyed. Control group was 1732 nonathletes school children the same age and gender.
Results: Results. 113 (6.7%) athletes had syncope (girls – 73.5%). According to the results of questioning in schools, syncope was noted in 4.2% of school children ($P < 0.05$ vs athletes). More often, syncope registered in basketball (10.5%), judo (10.3%), rhythmic gymnastics (9.1%), figure skating (8.0%), volleyball (7.4%). In all cases among athletes syncope had neurally mediated (reflex) nature by the results of the additional examination. In four out of 1687 athletes (0.24%) were firstly revealed the long QT syndrome, but they haven't got syncope.
Conclusion: Conclusions.
1. In the young elite athletes prevalence of syncope is 6.7% (4.2% for non- athletes) and significantly more common in girls (11.6% vs. 3.4%).
2. More often, syncope occurs in sports where tall athletes are selected (basketball, volleyball) or strict weight control is required (rhythmic gymnastics, figure skating, judo).
3. The majority of the young athletes has benign neurally mediated nature of syncope and doesn't need restrict for the sport, but the first of all they require the exclusion of diseases with a high risk of sudden cardiac death.
4. Long QT syndrome was revealed in 0.24% of young elite athletes.

Keywords: young elite athletes, syncope, sudden cardiac death, long QT syndrome, epidemiology

P-229

Cardiorespiratory fitness in children with long qt syndrome: a controlled cross-sectional study

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Background and Aim: Congenital long QT syndrome (LQTS) is the most frequent inherited arrhythmia, characterized by a prolonged QT interval, and an increased risk of syncope and sudden cardiac death. Because of sports and exercise restrictions, children with LQTS are at risk of physical deconditioning. Guidelines on sports participation in cardiovascular disease have become less restrictive over time, however, the cardiorespiratory fitness has not been well evaluated in the paediatric LQTS population. We aimed to evaluate, in children with LQTS, the cardiorespiratory fitness, the muscular strength and architecture, and the level of physical activity, in comparison with healthy controls.
Methods: This controlled bicentric cross-sectional study enrolled 20 children with LQTS children aged 6 to 17 years old compared to 20 healthy matched subjects. They underwent a complete cardiac check-up with cardiopulmonary exercise test and muscular ultrasound, performed muscle strength by functional tests, and wore an activity monitor for 14 days (Actigraph GT3X).
Results: In children with LQTS, peak oxygen uptake (VO2peak) and ventilatory anaerobic threshold were moderately impaired but significantly lower than in healthy controls (33.9 ± 6.2 vs. 40.1 ± 6.6 mL/Kg/min, $P = 0.004$, $d = -0.96$; 23.8 ± 5.1, 28.8 ± 5.5 mL/Kg/min, $P = 0.005$, $d = -0.95$, respectively). All children with LQTS had betablockers and a lower maximum heart rate during exercise. LQTS children had lower leg strength (119.5 ± 33.2 vs. 147.3 ± 36.1 cm, $P = 0.015$, $d = -0.8$) and muscle pennation angle (12.2 ± 2.4 vs. 14.3 ± 2.8°, $P = 0.01$, $d = -0.8$).
There was no difference on moderate-to-vigorous physical activity level between groups (36.67 ± 12.65 min/day vs. 41.92 ± 18.29 min/day, $P = 0.30$). Overall, 50 % had patterns of physical deconditioning.
Conclusion: Physical capacity in children with LQTS is moderately altered, from multifactorial limitation. Exercise rehabilitation could be of interest in LQTS children with significant physical limitation.

Keywords: Paediatrics, long QT syndrome, cardiorespiratory fitness, muscular deconditioning, exercise.

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The prevalence of 12-lead resting ECG abnormalities in duchenne muscular dystrophy patients

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Background and Aim: Electrocardiographic abnormalities are one of the leading manifestations of cardiac involvement in Duchenne muscular dystrophy (DMD), a chromosome X-linked dystrophinopathy. Despite, in most cases, asymptomatic, they are observed already in young patients with preserved left ventricle systolic function. We sought to determine prevalence of ECG findings in different age groups in DMD patients.
Methods: This is a single-centre observational study based on analysis of resting ECG recordings acquired between 2018 and 2020 in patients with genetically confirmed DMD diagnosis. All recordings were evaluated following the protocol recommended by Polish Cardiac Society. This included assessment of PQ, QRS, QT duration, R and S amplitudes in all leads, QRS morphology and average heart rate (HR). Reference values by Rijnbeek et al. were used. Right (RVH) and left (LVH) ventricle hypertrophy definition was based on Polish Cardiac Society guidelines. Statistical analysis was performed using Prism 9 software (GraphPad Software, USA).
Results: Sixty-two male patients aged 9 (2-17) were included, and sixty-four 12-lead resting ECG recordings were analyzed. Sinus tachycardia (HR > 98th percentile) was found in 23 (35.9%) recordings. Shortened PQ interval was present in 28 (43.7%) instances, most frequently in leads: I, aVL, V4, V5. Intraventricular conduction disturbance observed as QRS fragmentation with narrow QRS complexes was found in 39 (60.9%) recordings, most commonly in leads: aVF, III, V1, II. Abnormally high R waves (observed only in right side precordial leads – V1 and V2) were detected in 16 (25.0%) cases in V1 and 30 (46.9%) cases in V2. In 28 (43.5%) recordings RVH ECG criteria, defined as R wave amplitude>98th percentile or increased R/S ratio in V1 and V2, were fulfilled. There were n = 34 (54.8%) younger (<10 y.o.) and n = 28 (45.2%) older (≥10 y.o.) patients. No significant differences between the groups were found.
Conclusion: Electrocardiographic abnormalities are prevalent and appear early in the course of the disease. Their long-term prognostic value, utility for risk stratification and usefulness in tailoring cardiac prophylaxis and treatment in this patient group is unknown. Considering scarce and conflicting literature in this subject matter long-term observational study is warranted.

Keywords: Duchenne Muscular Dystrophy, Resting ECG,

P-231

Factors associated with propranolol monotherapy failure in infants with supraventricular tachycardia

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Background and Aim: Infants diagnosed with supraventricular tachycardia (SVT) are treated with prophylactic antiarrhythmic medication (AM) to avoid recurrence of SVT and associated morbidity. Propranolol is commonly used as initial AM. We studied the potential risk factors for propranolol monotherapy failure.
Methods: This descriptive retrospective cohort study included infants less than 12 months of age diagnosed with AVRT in the five tertiary care hospitals between 2005 and 2017. Infants were separated into two groups: those with successful propranolol monotherapy and those needing additional AM separately or in combination with propranolol.
Results: A total of 278 infants met inclusion criteria. Of those, 229 (82 %) had propranolol as initial AM. Propranolol monotherapy was successful in 144 (63 %) infants and unsuccessful in 85 (37 %). Antenatal arrhythmia (OR 2.15, 95 % confidence interval (CI) 1.17-3.95), admission to ICU (1.78, 1.02-3.13) and need for cardioversion (3.17, 1.79-5.62) were associated with propranolol monotherapy failure in univariate analysis. Median initial dose of propranolol in successful monotherapy was 2.8 mg/kg/day (interquartile range (IQR) 2-3) and the highest dose 3.2 mg/kg/day (IQR 3-3.6). In patients who had antenatal arrhythmia, propranolol was effective only in 29 out of 57 infants (51 %) and in those who needed cardioversion in 62 out of 122 infants (51 %). Antenatal arrhythmia (OR 2.13, 95 % CI 1.05-4.32) and need for cardioversion (3.68, 1.96-6.89) remain statistically significant independent risk factors for propranolol monotherapy failure when age, sex, prematurity, and admission to ICU were included in multivariate logistic regression analysis.
Conclusion: Antenatal arrhythmia and need for cardioversion are associated with propranolol monotherapy failure and early breakthrough of arrhythmias.

Keywords: Supraventricular tachycardia, infants, antiarrhythmic medication, prophylaxis, propranolol

P-232

Catheter ablation of right-sided accessory pathways with and without fluoroscopy in children: no difference in success, recurrence and complication

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Background and Aim: Catheter ablation with limited fluoroscopy via 3D electro-anatomical technology is commonly performed in children with atrial or ventricular tachycardia. The aim of this study was to evaluate the efficacy and safety of catheter ablation of right-sided accessory pathways (APs) performed with limited and without fluoroscopy in children.
Methods: A total of 51 children treated with right-sided accessory pathway ablation between July 2016 and September 2021 were evaluated retrospectively.
Results: The mean age was 12.2 ± 0.5 years and 30 (58.8%) patients were male. A history of previous ablation was positive in 12/51 patients. Thirty-seven (72.5%) patients presented with manifest APs. A total of 58 substrates of the right APs in 51 patients were ablated during this time period. 7 patients had ≥2 substrates. Two patients had a left and right AP. The location of APs were anteroseptal (parahisian) in 14 patients, anterior in 5, anterolateral in 3, lateral in 8, posterolateral in 7, posterior in 5, posteroseptal in 8 and midseptal in 8. Conduction property was antegrade-only in 6/58 substrates, retrograde-only 18/58 and both in 34/58. We performed ablation in 1 patient under the tricuspid valve in the right ventricle. Radiofrequency (RF) ablation was applied

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to 38 substrates. Cryoablation (Cryo) was used in 18 patients and both RF and Cryo in 2 because of proximity of His bundle. The mean procedure time was 216.3 ± 9.6 min. No X-ray was used in 17 (33.3%) patients. Limited fluoroscopy was used in 34 (66.6%) patients and median fluoroscopy time was 3.4 ± 0.6 min. The mean total radiation dose per body surface area was 731.8 ± 161.1 mGycm2/m2. The acute success and recurrence rate was 96.5% (56/58) and 10.3% (6/56) at a mean follow-up of 30.4 ± 2.8 months. No ablation-related complications occurred. Comparison of 18 patients ablated with no fluoroscopy to the 33 patients ablated with limited fluoroscopy showed no statistically difference in terms of acute success and recurrence rate ($p>0.05$). The procedure time was significantly lower in the patients with no fluoroscopy ($p = 0.002$).

Conclusion: Catheter ablation of right-sided APs with limited/zero fluoroscopy in children seems to be fast, safe and effective with high success rate and relatively low recurrence rate.

Keywords: right-sided accessory pathways, children, electroanatomic mapping, arrhythmia,

P-233

A rare origin of atrial flutter in a girl with congenital heart disease: 3D-electromapping guided radiofrequency ablation of the left atrial ceiling

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Background and Aim: Atrial flutter is a uncommon arrhytmia in children and associated with significant morbidity and mortality in children with congenital heart disease. Here, we present a 4-year-old girl with aort and mitral valve replacement, sinus node disfunction, epicardial pacemaker and atrial flutter originating from left superior atrium ablated successfully.

Methods: Here, we present a 4-year-old girl with aort and mitral valve replacement, sinus node disfunction, epicardial pacemaker and atrial flutter originating from left superior atrium ablated successfully.

Results: A 4 1/2-year-old girl, weighed 15.1 kg, was administered to the hospital because of being fatique during the bicycling for one year. She had history of transcatheter patent ductus arteriosus disclosure (January, 2010), aort and mitral valve replacement (February, 2010), and epicardial pacemaker implantation due to sinus node disfunction after the operation (March, 2011). During the follow-up, atrial flutter was detected at the age of 4-year-old, then propofenon (50 mg three times in a day) was started and a cardiac electrophysiologic study and a possible ablation were planned. During the endocardial electrophysiologic study, we recorded a counterclockwise macroreentry atrial tachycardia with 2:1 atrioventricular block at the cycle length 290 ms charactherized by negative flutter waves in II, III and aVF (Figure 1). The tachycardia cycle length in the right atrium was 90 msn, therefore wir performed transseptal puncture for the left atrial mapping. The ablation was performed a lineer ablation in the ceiling of the left atrium using the 5 Fr ablation cateter by delivering 20 W with a target temperature 60 °C under the guidance of the 3D electroanatomic mapping system. No tachycardia was induced after the ablation. The procedure and fluoroscopy time were 240 and 5 minute, respectively. No complication was observed. During follow-up of 6 years, she has no recurrence and ist on no antiarrhythmic treatment.

Conclusion: Although atrial flutter is seen very rarely in the children, it should be considered that the ceiling of the left atrium could be a substrate of atrial flutter. In adddition this case showed that radiofrequency ablation of left-sided atrial flutter could be safely performed in a small child.

Keywords: arrhythmia, children, atrial flutter, ablation, tachycardia

Figure In this picture it can be seen a counterclockweise macroreentry atrial tachycardia with cycle length 290 msn charactherized by negative flutter waves in II and aVF and the termination of the tachycardia by radiofrequency catheter ablation (top) as well as the picture of the ablation area by 3D-electroanatomic mapping system (bottom).

P-235

Machine learning-based arrhythmia detection tool for postoperative junctional ectopic tachycardia

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Background and Aim: Junctional ectopic tachycardiac (JET) is a prevalent life-threatening arrhythmia in children with congenital heart disease (CHD), with a marked resemblance to normal sinus rhythm (NSR) often leading to delay in diagnosis. There are no current bedside diagnostic algorithms to detect post-operative JET.

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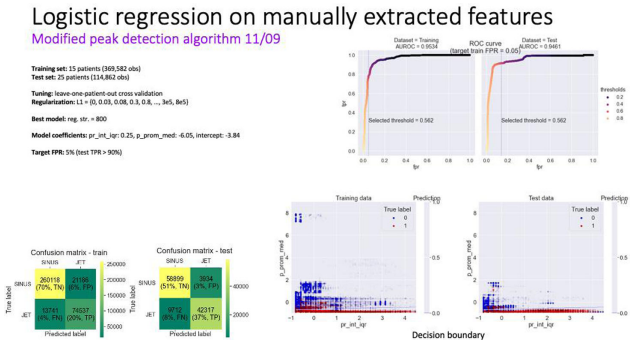
AIM: To develop a novel machine learning-based automated arrhythmia algorithm/tool to detect JET from real-time ECG signals.
Methods: A single-center retrospective cohort study of children with CHD was performed. Electrocardiographic (ECG) data produced by bedside monitors are captured automatically by the SickbayTM platform. Based on the detection of R and P wave peaks, 5 interpretable ECG features are calculated: P prominence median, PR interval interquartile range (IQR), P prominence divided by corresponding PR interval, P wave width, and P wave height. These features are used as input to a multivariate logistic regression classification model, and trained to distinguish JET from NSR. L1-regularization with tuned strength is applied to avoid overfitting.
Results: This study analyzed a total of 64.5 expert-labeled hours consisting of 509,833 cardiac cycles (R-R intervals), from 40 patients with CHD. Our classification model has an AUROC of 0.9449 for the detection of JET. A true positive rate of 90% can be achieved for a false positive rate of 5% on the test dataset.
Conclusion: This novel arrhythmia detection tool identifies JET, using two distinctive features of JET in ECG – the loss of a normal P wave and the PR relationship with high precision, which will allow for early detection and timely interventions for JET. Looking forward, error analysis performed on the best models strongly suggests that even higher performance is achievable upon improving the accuracy of PQRST detection algorithms with more interpretable physician-driven detection algorithms.

Keywords: Arrhythmia, machine learning

Logistic Regression Results



Confusion Matrix and AUROC curve differentiating results of LR-1

P-237

Reduced cardiorespiratory capacity in adolescents with ventricular arrhythmia

Radosław Pietrzak, Katarzyna Luczak Wozniak, Bożena Werner; Department of Pediatric Cardiology and General Pediatrics

Background and Aim: The prevalence of ventricular arrhythmia (VA) in young population is extremely common. If the arrhythmia burden is not high and heart morphology is normal it is considered as a benign phenomenon. On the other hand, VA in young individuals may cause dyssynchronous contractile activity and cardiac dysfunction. It is not known to what extent VA affects physical performance in those patients. The aim of the study was to assess to what degree VA affects physical performance in adolescents, using cardiopulmonary exercise testing (CPET)
Methods: The study group consisted of 33 asymptomatic children, aged 8-17 years with VA burden above 5% compared to 28 healthy controls matched according to sex and age. All patients underwent routine cardiological assessment including 24 hours ECG Holter monitoring and echocardiography and then CPET.
Results: The average number of ventricular extra beats in the Holter monitoring was 18584 (±15276) beats, arrhythmia burden was 18.8 % (±13.9%). Routine echocardiography showed there were no differences in the left ventricular size and function between the study and control group. During the CPET both, patients and controls achieved maximal effort. Patients in the study group showed statistically significantly lower ($p<0.05$) values of maximal oxygen consumption (VO2) when compared to the controls (33.9± ml/kg/min vs 38.5± ml/kg/min respectively). VO2 at anaerobic threshold (AT) was also statistically significantly lower ($p<0.05$) in the study group (17.1± ml/kg/min) comparing to this value in the controls (20.4± ml/kg/min). Although, oxygen pulse was lower in adolescents with VE (10.6 ml/beat) comparing to their healthy peers (11.0 ml/beat), this difference was not statistically significant. The correlations between the arrhythmia burden and such parameters of physical performance as maximal VO2 ($r = -0.1$) and VO2 at AT ($r = -0.1$) were week and not statistically significant ($p>0.05$). Throughout the exercise, adolescents in the study group with complex arrhythmia were not more affected by the low aerobic fitness than those with simple premature ventricular extra beats.
Conclusion: Adolescents with ventricular arrhythmia are affected by lower physical performance compared to their healthy peers. Physical fitness does not deteriorate with the increase of arrhythmia burden and complexity.

Keywords: ventricular arrhythmia, physical performance, adolescents

P-239

Temporary transvenous pacing in the sick preterm neonate

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Background and Aim: In infants with cardiac failure secondary to bradycardia, first line treatment includes medical management with isoprotenerol. In exceptional cases, patients may require a longer-term pacing strategy to assure higher heart rates that allow them to face transient higher metabolic demands without permanent commitment to a permanent pacing system. Potential options for pacemaker insertion in preterm neonates can be limited by patient size and co-morbidities.
Methods: We report two cases of temporary transvenous pacemaker insertion in preterm, low birth weight neonates with congenital complete heart block secondary to maternal autoimmune disease. Both patients had structurally normal hearts and required inotropic support for cardiac failure secondary to bradycardia.
Results: Patient 1:
Born in poor condition at 29 weeks gestation, birth weight of 1.7kg. On day one of life a temporary pacing wire (2 Fr Arrow quad) was inserted via the right femoral vein and attached to an external pacemaker generator (VVI 130ppm). Inotropic support successfully weaned. They underwent permanent epicardial single chamber pacemaker insertion (Microny K SR, VVI 120ppm) on day 4 of life. At the time of surgery it was noted that the temporary transvenous pacing wire had perforated the right ventricle wall without causing haemodynamic instability.
Patient 2:
Born at 29+4 weeks gestation, birth weight of 1.37kg. Transoesophageal pacing was attempted unsuccessfully.

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Isoproterenol resulted in a worsening lactic acidosis. A temporary transvenous pacing lead was inserted on day 8 of life via the right subclavian vein and attached to an external pacemaker generator (Medtronic Adapta, VVI 120 ppm). Complications included perforation of the right ventricle without haemodynamic compromise and pacing lead infection at 5 weeks of age. The infected pacing lead was successfully removed and they have remained haemodynamically stable without a pacemaker for over 18 months.
Conclusion: Temporary transvenous pacemaker lead insertion with an external pacing box allows to acutely stabilise sick preterm neonates with high performance and low risks. We recommend close monitoring of the system for adequacy of pacing parameters, signs of skin/ pacemaker lead infection as well as to exclude myocardial laceration and perforation.

Keywords: temporary pacemaker preterm neonate

P-240
Ablation of atrioventricular accessory pathways in pediatric patients: the single-centre experience

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Background and Aim: Accessory pathways are muscular bundles which connect atrium and ventricle and bypass the normal conduction system. They can cause Wolff-Parkinson-White syndrome and life-threatening arrhythmias. Ablation of the accessory pathways is the most effective therapy. Data on ablation and using 3D mapping systems (3D-MS) in children is limited. The aim of this study is to present our experience with ablation and 3D mapping systems.
Methods: The retrospective study enrolled 53 pediatric Wolff-Parkinson-White syndrome patients (32 boys, 21 girls; average age 13.2 ± 3.45 years, minimal age 3 years) with 61 intracardiac electrophysiologic studies: 59 radiofrequency ablations and 2 cryoablations of accessory pathways were performed.
Results: Ablations were performed for 61% right septal; 21% left lateral, 13% right lateral, 3.5% left septal accessory pathways. The average procedure time was 46.4 ± 17 minutes (from 20 to 105 minutes), average fluoroscopy time was 3.82 ± 3.75 minutes and the average number of energy ablation applications was 8.8 ± 8 (from 1 to 34). 3D-MS: EnSite NavX was used in 10 procedures, CARTO system in 1 procedure. The average procedure time with 3D-MS guiding was 43.2 ± 14.9 minutes, the average fluoroscopy time 1.1 ± 1 minutes and the average number of energy ablation applications was 6.1 ± 5. Fluoroscopy time using 3D-MS was statistically significantly shorter ($p = 0.005$). There was no significant difference in total procedure time and number of ablation applications in 3D-MS guiding ($p > 0.05$). Acute success of ablation rate was achieved in 77%. Total rate of complications was 1.6%.
Conclusion: Ablation of accessory pathways is a procedure with high success rates (77% after primary, 87% after repeated procedure) without major complications. 3D-MS is a useful tool to reduce fluoroscopy time.

Keywords: Atrioventricular accessory pathways, Children, Ablation, Electrophysiologic study, Mapping.

P-241
Jervell and lange-nielsen syndrome: a russian single centre study

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Background and Aim: Jervell and Lange-Nielsen syndrome (JLNS) is an autosomal recessive type of long QT syndrome characterized by congenital deafness and a high risk for sudden cardiac death at an early age. We aimed to analyze clinical and genetic characteristics from Jervell and Lange-Nielsen syndrome patients from our referral center for hereditary arrhythmic diseases.
Methods: Medical data from 12 Jervell and Lange-Nielsen syndrome patients were studied. The clinical information, 12-lead ECG, echocardiography, stress-test, 24-hour Holter recordings, blood tests, thyroid ultrasound and assessment of thyroid hormone levels were analyzed at least once per year during the follow-up period (mean 8.3 ± 5.1 7.5 years).
Results: The mean age was 3.0 ± 2 years old at the first visit, girls prevailed (67%). All patients are alive at the moment. The mean QTc was 578.9 ± 57.3 ms. Cardiac events had ten patients (83%), mean age at the first event was 1.9 ± 1.6 years. In 67% of children, syncope was initially regarded as epileptic. Eleven patients are genotyped by now, five of them have a homozygous variant in KCNQ gene (one has the whole exon deletion), three patients have compound heterozygous variants, and three patients are under the experimental confirmation of digenic origin of the disease. Beta-blockers were prescribed at mean age 3.0 ± 2.3 years (from 1 month to 7 years). In 8 patients, cardiac events recurred despite regular and adequate beta-blocker therapy. Cardioverter-defibrillator (ICD) was implanted for all of them. Patients with the first syncope under the age of 1.5 and delayed initiation of therapy (at the mean age 4.3 ± 1.7 years) had multiple syncope recurrences despite therapy. Non-cardiac symptoms include hypochromic anemia in 75% of cases. One patient developed juvenile absence epilepsy.
Conclusion: Early identification of patients and careful monitoring of risk factors allows to delay the age of manifestation of cardiac events and ICD implantation. ECG screening is crucial in solving the early diagnosis of the disease. If high risk of cardiac events persists in very young patients despite beta-blockers preferable next step would be a left cardiac sympathetic denervation as an alternative to ICD. Extracardiac pathology in patients with Jervell-Lange-Nielsen syndrome necessitates the interaction of doctors of different specialties when managing a patient.

Keywords: sudden cardiac death, Jervell and Lange Nielsen syndrome, children

P-242
A rare potentially life-threatening complication of subcutaneous implantable cardiac defibrillators in childhood: lead dislocation due to body growth

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Background and Aim: Subcutaneous implantable cardiac defibrillators (s-ICD) are increasingly implanted in selected pediatric patients, since short and long-term complications of transvenous ICDs associated with endovascular lead placement and later replacement can be avoided.
Methods: We report on a 14-year-old boy who received an s-ICD for secondary prevention after aborted sudden cardiac death. Diagnostic work-up (electrocardiogram, 24-hour Holter rhythm-monitoring, transthoracal echocardiography, exercise stress test, cardiac magnetic resonance imaging, coronary angiography, myocardial biopsy) did not reveal any abnormalities. Recently the results of the genetic testing revealed a mutation

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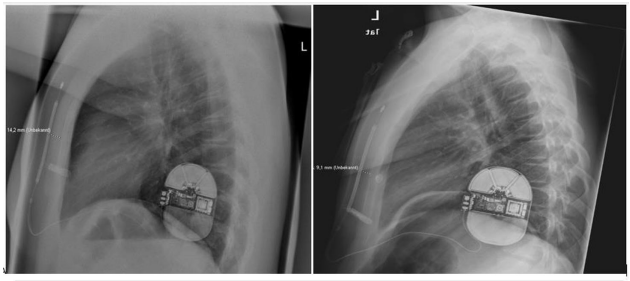
in the ryanodine type 2 receptor (RYR2) which is associated with catecholaminergic polymorphic ventricular tachycardia (CPVT). A sudden death of his mother at the age of 24 was reported which was not associated with exercise. S-ICD implantation in the adolescent was uneventful. Intraoperative defibrillation testing was successful with an impedance value of the subcutaneous lead of 91 Ohm. The patient was lost to follow-up despite several attempts to contact the patient's family. Three years after implantation, he presented in our outpatient clinic because of an episode of dizziness and loss of consciousness after a heated argument with another adolescent at school. S-ICD interrogation revealed an episode of ventricular fibrillation with five subsequent shock deliveries. Only the fifth shock was effective. The impedance of the subcutaneous lead had increased to 156 Ohm.
Results: The lateral x-ray of the thorax revealed a 5 mm growth of the subcutaneous fat tissue between sternum and shock coil compared to implantation. The lead of the s-ICD was surgically revised and defibrillation test was effective after revision.
Conclusion: Here we report for the first time that after s-ICD implantation body growth can cause ineffective shock delivery. We conclude that defibrillation testing should be performed yearly in children and adolescents with s-ICDs and this recommendation should be suggested by the manufacturer.

Keywords: subcutaneous implantable cardiac defibrillators, CPVT, ineffective shock delivery, childhood, body growth

pictures



Chest radiograph with lateral view highlighting the distance between sternum and shock coil with 9,1mm after the initial surgery (on the right) and 14,2mm after three years (on the left)

P-243

The therapeutic dance into the electrical storm of the TANGO2 disease: a case report

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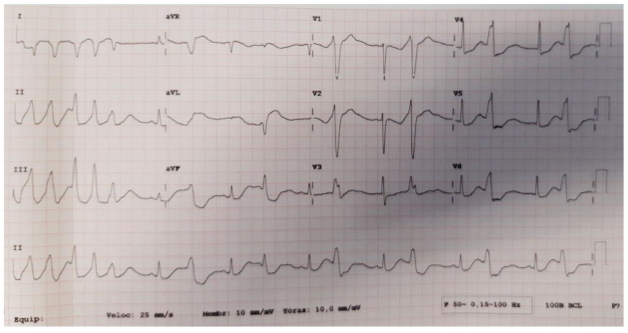
Background and Aim: TANGO2-related metabolic encephalopathy and arrhythmia is a rare, newly recognized and likely under-diagnosed condition. First described in 2016, the phenotype is characterized by developmental delay and recurrent metabolic crisis associated with rhabdomyolysis. During this episodes, patients may present QTc prolongation and ventricular arrhythmias.
Methods: We report a 13-year-old, with developmental delay who presented with severe rhabdomyolysis. The initial electrocardiogram (EKG) was normal.
Results: Due to worsening of rhabdomyolysis, QTc prolongation was identified (QTc 570ms) and oral β-blocker therapy started. A non-sustained ventricular tachycardia developed (fig.1), initially managed with magnesium bolus and impregnation dose of lidocaine, followed by perfusion. The echocardiogram did not identify structural or functional alterations. After a short period, frequent polymorphic ventricular extrasystoles (VPB) in short-long-short sequences developed. An arrhythmic storm of VPB induced torsade de Pointes (TdP) was triggered, leading to cardiac arrest. After resuscitation and due to the bradycardia dependent nature of the TdP, a temporary percutaneous pacing lead was placed and esmolol infusion started. Normal plasma levels of potassium, calcium and magnesium were maintained. Despite therapeutic optimization, new episodes of TdP developed, requiring defibrillation. The electrical instability ran in parallel with increasing severity of rhabdomyolysis and systolic ventricular function decline. Genetic testing identified a pathogenic variant in homozygosity in the TANGO2 gene. Stable sinus rhythm was finally achieved with optimization of the caloric intake, stable blood glucose levels and normal serum electrolytes. EKG showed normalization of the QTc interval.
Conclusion: The full TANGO2-related phenotype emerges over time and the prognosis is linked to the appearance of EKG abnormalities. QT interval prolongation can lead to life-threatening ventricular tachycardias.
The arrhythmia mechanism seems to be secondary to metabolite build up in cardiomyocytes, what can explain the cardiac phenotype during the crises witch subside after their resolution. In this patients, avoiding bradycardia is fundamental, since long QT related TdP seems to be triggered by bradycardia and short-long-short VPB.
During acute metabolic crises, the management of arrhythmias relies on metabolic control.

Keywords: Torsade de pointes, TANGO2 disease, TANGO2-related metabolic encephalopathy and arrhythmias



Recording of non-sustained polymorphic ventricular tachycardia and ventricular premature beats in bigeminy pattern.

P-244

Short-term electrocardiographic atrial remodelling after percutaneous atrial septal defect closure with the GCO device in a pediatric population

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Background and Aim: The GORE® CARDIOFORM septal occluder (GCO) is an atrial septal defect/patent foramen ovale

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(ASD/PFO) closure device with theoretical advantages over other commercialized devices thanks to its softness and anatomical compliance.
Our aim was to evaluate the short and medium-term electrocardiographic changes after percutaneous ASD closure with GCO in a pediatric population.
Methods: We enrolled 39 patients with isolated ASD submitted to trans-catheter closure with GCO from January 2020 to June 2021. EKG was performed before (T0), at 24 hours (T1) and 6 months (T2) after ASD transcatheter closure. P wave dispersion was calculated as the difference between maximum and minimum P-wave duration, PR interval as the interval between beginning of the P wave and beginning of the QRS complex and QT dispersion as the difference between maximun and minimun of QTc intervals. At 6-months from device implantation, the patients were submitted to ambulatory EKG Holter recording.
Results: Patients' age and BSA were 8.2 ± 4.2 years (IQR 4.2-8.3, median 7.0) and 1.0 ± 0.3 m2 (IQR 0.7-1.7, median 0.9), respectively. The strectched ASD diameter was 16.3 ± 4.5 mm (median 16), resulting in QP/QS of 1.7 ± 0.6 (median 1.5). At the baseline mean P wave dispersion was 40 ± 15 msec and decreased to 30 ± 13 msec ($p<0.002$) at 24h, without any further change at 6 months (30 ± 13 msec, $p<0.002$). PR conduction significantly improved at 24 h from device implantation (from 175.0 ± 20.8 to 144.0 ± 22.7 msec, $p = 0.018$) and did not significantly change at 6 months (164.0 ± 19.5 msec, p = NS). QTc dispersion decreased at 24 hours (31.7 ± .20.3, $p<0.02$) and at 6 months (28.0 ± 18.1, $p<0.002$) from device implantation. After device deployment, 2 pts (5%) developed transient, self-limited junctional rhythm and one of them needed a short course of anti-arrhythmic therapy for supra-ventricular tachycardia. No tachy/brady-arrhythmias were recorded at the 6-months follow-up EKG Holter monitoring.
Conclusion: Percutaneous ASD closure with the GCO device results in significant, sudden improvement of intra-atrial, atrio-ventricular and intraventricular electrical homogeneity. This benefit persists unaltered over a medium term follow-up. It might be due to a favourable volumetric remodelling that was not hindered by mechanical impact of the occluding prosthesis and could explain the low rate of arrhythmias found at the mid-term EKG evaluation.

Keywords: Atrial septal defect, electrocardiographic cardiac remodelling, cardiac arrhytmias

P-245
Tachycardia-induced cardiomyopathy in infants with multifocal atrial tachycardia

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Background and Aim: Multifocal atrial tachycardia (MAT) in most children has a mild course and a good prognosis, but sometimes can be the cause of tachycardia-induced cardiomyopathy (TIC).
PURPOSE: To analyze a clinical course of MAT complicated by TIC.
Methods: We observed 15 infants with MAT in 2011-2020. 7 (46.7%) patients were boys. The age of arrhythmia's primary registration was 2.04 ± 2.27 months (Me 1.5), 4 - in utero. Physical examination, laboratory monitoring, 12-lead ECG recording, 24-hour ECG monitoring, echocardiography at baseline and during follow-up were performed.
Results: Among 15 patients with persistent MAT 7 (46.7%) had a TIC. The average heart rate (HR) in group with TIC was 161.7 ± 26.9 bpm, the maximum – 262.9 ± 24.5 bpm; without TIC – 150.5 ± 19.6 bpm and 239.4 ± 18.5 bpm. LVEF was 37-55%, 3 patients had atrium dilatation (z-score>2,0) also. Nobody had LV dilatation (z-score -0.28 ± 1.19, Me -0.49). NTproBNP was elevated in 5 of them (370-13003 pg/ml). All patients with TIC received antiarrhythmic therapy (AAT): propranolol (n = 4) or amiodarone (n = 3; 1 – per os, 2 – intravenous) as a start, in the next stages also propafenone, digoxin and sotalol. Monotherapy was effective in 1 patient (propranolol). Combined AAT was used in 6 patients, including simultaneous use of 3 drugs (amiodarone +digoxin and propranolol or propafenone) in 2 of them.
As a result, sinus rhythm (n = 2), sinus rhythm with atrial ectopic activity (n = 2); MAT with HR control (n = 2) were observed. In one case daytime tachysystole persisted despite three-component AAT, but echocardiography parameters improved.
The follow-up period was 29.8 ± 8.98 months. Echocardiography was normalized during the one year. The arrhythmia disappeared in a period from 1 to 15 months (Me 5 months). The duration of AAT was from 4 to 26 months (Me 9.5 months). There were no therapy complications, lethal outcomes and recurrence of MAT after AAT withdrawal.
Conclusion: In our study TIC was observed in 46.7% infants with MAT manifested as a reduced LVEF and in 3 cases also atrial dilation. Combined AAT and HR control strategy can be justified to cope the TIC. It's necessary to continue further studies of molecular genetic predisposing factors of TIC.

Keywords: Multifocal atrial tachycardia, tachycardia-induced cardiomyopathy, infants, antiarrhythmic therapy

P-246
QTC intervals are not prolonged in former ELBW infants at pre-adolescent age

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Background and Aim: Survivors of preterm birth have an increased risk for cardiovascular mortality later in life. Whether preterm birth is associated with cardiac conduction or repolarization abnormalities in later life is still poorly explored, with conflicting data on QTc prolongation in former extreme low birth weight (ELBW, <1000 g) infants.
Methods: Twelve lead electrocardiograms (ECG) at rest, collected in the PREMATurity as predictor of children's Cardiovascular-

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renal Health (PREMATCH) study in former ELBW cases and term age-matched controls during pre-adolescence (8–14 years) were analyzed on corrected QT-time (QTc, Bazett) and QT-dispersion (QTd). ECG findings were compared between groups (Mann−Whitney), and associations with clinical and biochemical findings were explored (Spearman). In ELBW cases, associations between QTc and perinatal characteristics (at birth, neonatal stay) were explored (Mann−Whitney, Spearman).
Results: QTc and QTd were similar between 93 ELBW cases and 87 controls [409 (range 360–465) versus 409 (337–460); 40 (0–100) versus 39 (0–110)] ms (Table 1). Age, height, weight, or body mass index were not associated with the QTc-interval, while female sex (median difference 11.4 ms) and lower potassium ($r = -0.26$) were associated with longer QTc-interval. We could not observe any significant association between QTc-interval and perinatal characteristics.
Conclusion: There were no differences in QTc or QTd between ELBW and term controls in ECGs at rest in pre-adolescents. It is therefore unlikely that alterations in repolarization explain the observed increase in cardiovascular mortality. Also, our study in pre-adolescent former ELBW cases does not support a priori avoidance of QT prolonging medicines in this population. Future perspectives include the pooling of QT-data with other published but conflicting cohorts.

Keywords: QT, repolarization, ELBW, preterm birth

Table 1. Comparison of ELBW cases versus controls for clinical measurements, ECG-related measurement and biochemical measurements, data reported by median and range.

Characteristic	ELBW cases $n = 93$	Controls $n = 87$	*p* value
Clinical measurements			
Age, year	11 (9–14)	10.8 (9–14)	0.025
Height, cm	145 (124–168)	147 (129–181)	0.027
Weight, kg	35.5 (21.4–71.4)	37.7 (23.5–67.8)	0.043
BMI, kg/m2	16.5 (12.6–27.5)	17.2 (12.7–24.9)	0.053
Male/female	47/46	42/45	
Blood pressure, systolic, mmHg	114 (94–146)	107 (88–132)	<0.001
Blood pressure, diastolic, mmHg	69 (48–88)	65 (52–80)	<0.001
Medicine use, any	18/93	7/87	0.019
Medicine use, QTc prolongation	4/93	4/87	0.635
ECG-related measurements			
Heart rate, bpm	71 (47–104)	67 (48–111)	0.052
PR, ms	124 (94–188)	134 (94–196)	0.005
QT, ms	382 (320–438)	384 (320–442)	0.046
QTc (*Bazett*), ms	409 (360–465)	409 (337–460)	0.870
QTd, ms	40 (0–100)	39 (0–110)	0.673
QRS time, ms	82 (70–104)	84 (68–112)	0.015
QRS axis, °	78 (−24–104)	77 (28–117)	0.628
Biochemical measurements			
Sodium, mmol/L ($n = 127$)	141 (137–146)	140 (134–146)	0.185
Potassium, mmol/L ($n = 127$)	4.38 (3.72–5.92)	4.4 (3.87–5.36)	0.589
Calcium (total), mmol/L ($n = 127$)	2.52 (2.24–2.82)	2.46 (2.17–2.67)	0.001
Phosphate, mmol/L ($n = 127$)	1.52 (0.91–1.96)	1.48 (1.13–1.84)	0.550

ELBW extreme low birth weight, <1000 g, *BMI* body mass index, *ECG* electrocardiogram, *bpm* beats per minute.

P-247 / Moderated Poster
Epicardial device migration in children – a rare but important complication

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Background and Aim: Epicardial access is often used in the paediatric population for implanting cardiac devices. Permanent epicardial pacing has been in practice for many years but comes with potential risks and complications. Migration of the device generator box is a recognised complication but has only been described in isolated case reports in the literature. We aim to study the incidence and associated risk factors for this complication.
Methods: All paediatric patients implanted with epicardial devices at the Royal Brompton Hospital between 1992 and 2018 were retrospectively reviewed and included in this study. Patient characteristics recorded include pacing indication, age and weight at time of implantation, device type, surgical approach, original device position, any subsequent pacing intervention and any other abdominal surgery. The operation notes, pacing checks, chest X-rays and/or CT scans were reviewed with special attention to the device position. Migration of the device was confirmed if the device was visualised outside the area where it was originally implanted, either on imaging or during subsequent pacing intervention.
Results: 80 patients were included in our analysis with a mean follow up duration of 91 months (1-271mths). Device migration occurred in 7 patients (8.75%), with an incidence of 1.15 per 100 patient-years. The mean age and weight were lower in the patients who suffered from device migration [0.34 (7days-1year) vs 4.03 (1day – 18years) years - $p<0.01$] and [4.3 (2.1-8kg) vs 14 (2.1-80kg) kg; $p<0.01$ respectively]. Further abdominal surgery performed after device implantation was also more common in these patients (3/7(43%) vs 6/73(8%); $p = 0.02$). There were no statistically significant differences between the groups with regards to gender, pacing indication, surgical approach and original device position.
Conclusion: Our study suggests that young age and low weight at the time of implantation as well as abdominal surgery performed after epicardial device implantation, are important risk factors for device migration. In these patients, it is especially important that this complication is considered and monitored for during subsequent device follow up.

Keywords: epicardial pacemaker, pediatric, migration

P-248 / Moderated Poster
Clinical characteristics and mid-term follow-up findings in children with isolated complete heart block: a single-center ten-year experience

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Background and Aim: Isolated complete atrioventricular block (CAVB) is a rare disease often associated with maternal autoantibodies. It may occur with or without structural heart disease.

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Although some of the patients are diagnosed in the prenatal and neonatal periods, the diagnosis rate in childhood is quite high. In this study, we aim to present the data of our patients diagnosed with isolated CAVB.

Methods: In this study, we evaluated 108 patients diagnosed with isolated CAVB between 2011 and 2021. Demographic data of the patients, electrocardiography, echocardiography, 24-hour Holter monitoring data and follow-up and complications of the patients who underwent pacemaker implantation were evaluated retrospectively from hospital records.

Results: The mean age at diagnosis of the patients was 5.51 ± 5.05 years. At the time of diagnosis, 74.8% of the patients had no symptoms associated with AV block. The most common symptom was fatigue. 25% of the patients were diagnosed in the first month of life. Echocardiography were normal in 42 patients, left ventricle (LV) dilatation in 14 patients at baseline. Mitral valve regurgitation was mild in 48 patients, significant in one, and moderate in 2 patients. Pacemaker implantation was needed in 87 patients during follow-up. The most common pacemaker implantation indication was significant bradycardia. The mean battery life was 5.41 ± 2.65 years, Replacement-free period of 68 patients who underwent pace implantation and continued their follow-up was 4.18 ± 2.89 (0.1-10) years. The replacement-free period was 98.5% in the 1st year, 85% in the 5th year, 74.5% in the 7th year. Pacemaker-related complications developed in 8 patients during follow-up. LV dysfunction developed (dyssynchrony induced) in 3 patients at follow-up, and all were paced from the right ventricle anterior wall. All of these patients underwent cardiac resynchronization therapy (CRT) and LV dysfunction improved.

Conclusion: Isolated CAVB is a rare disease requiring careful clinical follow-up. Patients are often asymptomatic and the most common indication for pacemaker implantation is significant bradycardia. LV dysfunction is an important cause of morbidity, especially in patients with right ventricular anterior wall pacing. Physicians should be aware of LV dysfunction during follow-up. CRT should be considered as a treatment option for LV dysfunction.

Keywords: Isolated complete atrioventricular block, Pacemaker, Congenital complete atrioventricular block, Cardiac resynchronization therapy

P-249
Clinical characteristics, and course of sinus node dysfunction in children: A ten years single-center experience

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Background and Aim: To evaluate the clinical characteristics and outcomes of children diagnosed with sinus node dysfunction (SND).

Methods: This was a retrospective review of patients diagnosed with sinus node dysfunction in a tertiary paediatric cardiology center in Turkey from January 2011 to November 2021.

Results: In all, 51 patients (34/66.6% males) were included, with a mean age of 9.3 ± 6.4 years and a mean weight of 29.6 ± 19.2 kg.The age inappropriate bradycardia and pauses were the most common rhythm disturbance, and syncope and dizziness (n:23, 45%) were the most frequent initial symptoms.Forty of the 51 patients (78%) had structural heart disease, thirty two of them had congenital heart disease, most commonly AV septal defect (n:10), atrial septal defect (n:5) and transposition of great arteries (n:4). Seven of them had also left atrial isomerism.The remaining eleven patients were isolated. Two of our patients were siblings and had SCN5A mutation.Of the total patient population, 29 patients (57%) had previously undergone a cardiac operation and except one patient sinus node dysfunction developed after a surgical procedure. This patient had left atrial isomerism and SND was diagnosed during 24 hours holter monitorization before surgery. The most common surgical procedures were fontan operation in six, closure of atrial septal defect in five, correction of AV septal defect in four, Senning operation in three, and abnormal pulmonary venous connection anomaly repairment in two. Twenty nine (56%) patients (17 of them were after cardiac surgery) were underwent pacemaker implantation. The mean interval between pacemaker implantation and the previous operation was 5,1 years (range, 0-18.4 years). Five of them had pacemaker implantation within the first 30 days after surgery. All symptomatic patients noted resolution of symptoms after pacemaker implantation. During the mean follow-up time of 43 ± 33.7 months, no mortality was seen.

Conclusion: Although SND is rare in children, it has been diagnosed with increasing frequency with structural heart disease especially in patients who have undergone corrective cardiac surgery related with atrial tissue. Since SND can occur at any time postoperatively, these patients should be kept under constant control. If symptomatic sinus node dysfunction is confirmed, permanent pacing is an effective therapeutic modality.

Keywords: Sinus node dysfunction, children

Age at diagnosis (months) Mean (±SD)	113.7±78.9
Weight at diagnosis(kg) Mean (±SD)	29.6±19.2
Structural heart disease	**40 (78.4)**
✓ **Congenital heart disease***	
o *Atrioventricular septal defect (Complete or partial)*	10
o *Atrial septal defect*	5
o *Transposition of great arteries*	4
o *Partial/total anomalous pulmonary venous connection*	3
o *Others*	11
o *Left atrial isomerism*	7
✓ **Cardiomyopathy**	
o Left ventricular noncompaction cardiomyopathy	3
o Hypertrophic cardiomyopathy	2
o Restrictive cardiomyopathy	1
✓ **Myocarditis**	1
Additional arrythmia substrates **	**21(41.1)**
✓ *Supraventricular tachycardia*	
o Atrial fibrilation	4
o Intraatrial reentran tachycardia	4
o Short RP SVT	4
o Focal atrial tachycardia	3
o Multi focal atrial tachycardia	1
✓ Ventricular *arrythmias*	
o Ventricular fibrilation	
o Idioventricular rhythm	1
✓ Others	1
o Complete AV block	2
o Long QT	1
Pacemaker implantation	**29 (56.8)**
✓ *Postoperative*	17
✓ *Isolated*	12
Pacemaker indication	
✓ *Symptoms correlate with* severe bradycardia or pauses	23
✓ Significant bradycardia that does not improve in the early postoperative period	5
✓ *Preoperative symptomatic SND (in left atrial isomerism petient)*	1

Values mean±SD or n (%)

*: Some patients had multipl or complex congenital cardiac disease| **: Some patients had more than one *arrythmia* substrat.

Figure 1. Clinical characteristics of patients

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P-250

Catheter ablation in patients with congenital heart disease using three-dimensional electroanatomic mapping system with limited fluoroscopy

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Background and Aim: The purpose of this study was to evaluate arrhythmia substrates, outcomes, and complications of catheter ablation using a three-dimensional (3D) electroanatomical mapping system in children with congenital heart disease (CHD).

Methods: The medical records and procedural data of patients with CHD who underwent invasive electrophysiological study (EPS) and catheter ablation between November 2013 and 2021 were evaluated. Clinical characteristics, echocardiographic examination, ablation procedures, outcomes and follow-up were obtained from hospital records using the Filemaker database. EPS were performed using the EnSite™ system (St Jude/Abbott Medical, St Paul, MN, USA) with limited fluoroscopy.

Results: 132 patients (73 males, 55.3%) with CHD and treated with catheter ablation were evaluated retrospectively. The mean age was 14.5 ± 7.8 years (range, 2-43 years) and the mean weight was 47.3 ± 20.4kg (range, 11-98 kg). The most common diagnoses were Ebstein's anomaly (n:32), atrial septal defect (n:25), ventricular septal defekt (n:16), tetralogy of fallot (n:21), single ventricle (n:7) and transposition of great arteries (n:5). 72 (54.5 %) patients had undergone prior corrective or palliative cardiac surgery. Arrhythmia mechanisms included Wolff-Parkinson-White syndrome (n:41; 28%), intra atrial reentran tachycardia (IART, n:30; 20.5%), atrioventricular nodal reentry tachycardia (AVNRT, n:29; 19.8%), focal atrial tachycardia (FAT, n:14; 9.5%), PVC (n:13; 8.9%), AVRT due to concealed accessory pathway (n:11; 7.5%), ventricular tachycardia (n:2; 1.3%) and others (n:6; 4.1%). Of the 132 procedures, 45 (34%) were radiofrequency ablations, 33 (25%) were irrigated radiofrequency ablations and 26 (%19,6) were cryoablation. Both non irrigated and irrigated radiofrequency ablation were used in 21(15.9%) and radiofrequency ablation and cryoablation were used in eight (6%) procedures. The mean procedure time was 178.4 ± 70.6 minutes.Fluoroscopy was used in 78 (%59) patients. The mean floroscopy time was 5.2 ± 7.8 minutes. Procedural success was complete in 92.4% (n = 122) and partial in 5.3% (n = 7) of patients, whereas ablation failed in 7% (n = 2.3). During a mean follow-up period of 38.1 ± 24.9 months 12 patients had recurrence (%9.8). Except for two, the procedure was successful with the second ablation performed in the others. No major complications were observed,except for the patient who underwent balloon angioplasty after narrowing of the Cx during the procedure.

Conclusion: Despite the complex anatomy,age,and limited vascular intervention possibilities in patients with CHD,transcatheter ablation treatment with 3-D electroanatomical mapping and limited fluoroscopy seems to be a safe and effective option.

Keywords: pediatric, congenital heart disease, transcatheter ablation, limited flouroscopy, 3-D electroanatomic mapping system

Male sex	73 (55.3)
Age,year	14.5±7.8 (2-43)
Weight, kg	47.3±20.4 (11-98)
Weight <20 kg	11 (8.3)
Congenital heart disease	
Ebstein's anomaly	32 (24.2)
Atrial septal defect	25 (18.9)
Tetralogy of fallot	21 (15.9)
Ventricular septal defekt	16 (12.1)
Single ventricle	7 (5.3)
Transposition of the great arteries	5 (3.8)
Congenitally corrected transposition of the great arteries	4 (3)
Atrioventricular septal defect	3 (2.2)
Partial anomalous pulmonary venous connection	2 (1.5)
Others	17 (12.8)
Procedural time, min	178.4±70.6
Fluoroscopy time, min	5.2 ± 7.8
Arrhythmia mechanism(s)*	
Wolff- Parkinson-White syndrome	41(28)
Intra atrial reentran tachycardia	30 (20.5)
Atrioventricular nodal reentry tachycardia	29 (19.8)
Focal atrial tachycardia	14 (9.5)
Ventricular ectopy	13 (8.9)
Concealed accessory pathway	11 (7.5)
Sustained ventricular tachycardia	2 (1.3)
Mahaim fibers	2 (1.3)
Junctional ectopic tachycardia	2 (1.3)
Multifocal atrial tachycardia	1 (0.7)
Atrial fibrillation	1 (0.7)
Ablation energy source	
Nonirrigated-tip radiofrequency	45 (34)
Irrigated-tip radiofrequency	33 (25)
Cryoablation	26 (19.6)
Cryoablation&Nonirrigated-tip radiofrequency	8 (6)
Nonirrigated& Irrigated-tip radiofrequency	21 (15.9)
Acute Success	122 (92.4)
Procedural complications	1 (0.7)
Follow up time,months	38.1 ± 24.9
Recurrence	12 (9.8)

Values are presented as n (%), mean±SD and range(min-max)

*: Some patients had more than one substrate

Figure 1. Clinical and Procedural Characteristics (N=132)

P-251

Utility and outcomes of ajmaline provocation test for brugada syndrome in pediatric patients; single center 8 year experience

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Background and Aim: Brugada syndrome (BrS), an inherited arrhythmogenic disease which can cause syncope or sudden cardiac death (SCD), is characterized by ST-segment elevation in right precordial leads. Sodium channel blocking drugs (Ajmaline, Flecainide) are used to reveal ECG findings in unidentified forms. In this study, we aimed to share our clinical experience of pediatric patients who were suspected of BrS and performed Ajmaline provocation test (APT).

Methods: The data of 78 patients who underwent APT between 2013-2021 were analyzed retrospectively. In the test protocol, Ajmaline was given to each patient as a six to ten-minute infusion

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(1mg/kg). 12-channel ECG recordings were taken at the beginning of the test and at two-minute intervals. During this whole period, the test was considered positive in case of the appearance of type-1 Brugada sign, especially on lead V1 and V2.
Results: 51/78(65%) patients included in the study were male. The mean age at admission was 13.02 ± 4.34(0.98-21.27) years and the mean body weight was 47.06 ± 19.48(9-95)kg. On admission 15 patients(19.2%) had syncope, 15(19.2%) had palpitations and 15(19.2%) had a history of cardiac arrest. 20(25.6%) patients had a family history of sudden death. 12 lead basic ECG, suspected Brugada sign was observed in 23 patients, borderline long QT interval in 3 patients, and right bundle branch block in 2 patients. Echocardiographically revealed normal cardiac findings except asymmetric septal hypertrophy in one patient and mitral valve prolapse in another. APT was positive in 13/78 (16.6%) patients (Table-1) and there were no arrhythmias or adverse events during testing. In 7 (53.8%) of the 13 patients with a positive test result, ICD was implanted (syncope (n = 6) and cardiac arrest (n = 1)). During to mean 5 years follow-up period, there was no major arrhythmic event in these 13 patients.
Conclusion: APT test can be performed safely and effectively in suspected BrS, aborted cardiac arrest, and a family history of sudden death. In positive cases; medical treatment ± ICD decision should be made by evaluating together with other clinical findings and risk factors.

Keywords: Brugada syndrome, pediatric, Ajmaline, provocation test,

Table-1: Characteristics of the patients with a positive Ajmaline provocation test result

	Sex, Age	symptom	Family history of SCD or BrS	Therapy
Patient 1	m,12.2 year	Syncope	+	ICD
Patient 2	m, 16.3 year	Cardiac Arrest	-	ICD
Patient 3	f, 14.1 year	Palpitation, Syncope	+	ICD
Patient 4	f, 20.6 year	(asymptomatic)	+	-
Patient 5	m, 15.9 year	(asymptomatic)	-	-
Patient 6	m, 15.2 year	Syncope	-	ICD
Patient 7	m, 21.2 year	Syncope	+	ICD
Patient 8	f, 13.9 year	(asymptomatic)	+	-
Patient 9	m, 16.6 year	(asymptomatic)	-	-
Patient 10	m, 10.1 year	Syncope	-	-
Patient 11	f. 5.2 year	Palpitations	+	-
Patient 12	m, 9.4 year	Cardiac Arrest	-	ICD
Patient 13	f, 15.6 year	Syncope	+	ICD

BrS: Brugada Syndrome, f: female, m: male, SCD: sudden cardiac death

P-252
Clinical characteristics and management of aborted sudden cardiac arrest in children: a 10-year single cardiac center experience

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Background and Aim: Sudden cardiac arrest (SCA) in children is very rare, it has various causes. In this study, we provided an overview of SCA survivors and evaluated investigations for etiology with management in our center.
Methods: We identified 59 pediatric patients with aborted SCA (aSCA) only documented by the healthcare professionals (emergency physician, pediatrician, and pediatric cardiologist) between March 2010 and December 2021. Clinical history, non-invasive and invasive diagnostic tests and therapies were noted from FileMaker® Database.
Results: The mean ± standard deviation age of patients was 9,6 ± 5.5 years (2 month-21 years) and 55.9% were male. Echocardiography and other imaging tests revealed structural heart disease (group 1) in 18 patients (30.5%). 37 patients (62.8%) had electrical heart disease (group 2) and no cause was found in the remaining four patients (Table 1). Hypertrophic cardiomyopathy was the most common cause (4/18, 22%) in the first group. Long QT syndrome (LQTS) was the most common cause (12/17, 32.4%) in the second group. 46 patients (78%) had SCA as the first manifestation of heart disease. While SCA occurred with exertion in 14 patients (23.7%), SCA developed in 38 patients (64.7%) at rest. According to the information obtained from the data, the most common documented cardiac arrest rhythm was ventricular fibrillation in 15 patients (25.4%). The median intensive care unit follow-up period of survivors after sudden cardiac arrest was 7 days (2-46 days). Neurological sequelae remained in 10 (16.9%) patients after aSCD. 15 patients (25.4%) underwent electrophysiological study and/or ablation. ICD implantation was performed in 44 (74.6%) patients (29 transvenous, 14 epicardial, 1 cardiac resynchronization therapy-defibrillator (CRT-D)). ICD was not implanted in 15 patients. Of these, 7 patients did not require ICD due to definitive treatment (successful ablation therapy, surgical coronary reimplantation, pacemaker implantation, sympathectomy). ICD could not be inserted because 4 patients died. 4 patients and their families refused ICD. 9 patients (15.3%) died during the follow-up.
Conclusion: The etiology of aSCA is very variable and present at any age. Detailed non-invasive and invasive investigations are crucial in revealing the diagnosis. Appropriate treatment and secondary preventive ICD implantation are life-saving route for selected SCA survivors.

Keywords: aborted sudden cardiac death, children, cardiomyopathy, long QT, implantable cardioverter-defibrillator

Baseline characteristics of the patients	(n=59)
Median age(months)	9,6 (2months- 21 years)
Male gender	33 (55.9%)
Weight (kg)	39.47±20.36
First manifestation	46 (78%)
Sudden cardiac arrest	10 (16,9%)
Syncope	2 (3.4%)
Palpitation	1 (1.7%)
Exercise intolerance	
Family history	12 (20.4 %)
Sudden death	9 (15.3%)
Arrhythmia	7 (11.8%)
Pacemaker/ICD	2 (3.4%)
Syncope	1 (1.7%)
Cardiomyopathy	28 (47.4%)
No history	
ECG finding at the time of the event	15 (25,4%)
VF	10 (16,9%)
VT	4 (6.8%)
Torsades de pointes	2 (3.4%)
Atrial fibrillation+ VF	1 (1.7%)
Asystole	27 (45.8%)
Unkown	
Sekondary prevention	29 (49,2%)
Transvenous ICD	14 (23.7%)
Epicardial ICD	1 (1.7%)
CRT-D	

Table 1. Baseline characteristics of the patients

Sudden Cardiac Arrest Etiology	n	%
Group 1. Structural causes	18	30.6
Cardiomyopathy	11	18.7
• Hypertrophic Cardiomyopathy	4	6.8
• Dilatative Cardiomyopathy	3	5.1
• Restrictive Cardiomyopathy	1	1.7
• Arrhytmogenic right ventricular dysplasia/cardiomyopathy (ARVD/C)	2	3.4
• Left ventricular non compaction	1	1.7
Opere Congenital Heart Disease	6	10.2
• Opere TOF	2	3.4
• Opere Aort Coarctation, mitral valve repair	1	1.7
• Opere VSD	2	3.4
• Opere VSD, Bentall procedure	1	1.7
Anomalous left coronary artery from pulmonary artery (ALCAPA)	1	1.7
Group 2. Non-structural (Electrical) causes	37	62.8
LQTS	12	20.4
CPVT	10	16.9
Idiopathic Ventricular Fibrillation	4	6.8
Ventricular Tachicardia	3	5.1
Wolf-Parkinson-White Syndrome	3	5.1
Brugada Syndrome	2	3.4
Early repolarization	2	3.4
Atrioventricular Block	1	1.7
Unexplained causes	4	6.8
Total	59	100

Table 2. Sudden Cardiac Arrest Etiology

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P-253
Electroanatomic mapping guided cryoablation of anteroseptal-midseptal manifest accessory pathways in children: single center 11-years experience

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Background and Aim: Catheter ablation of anteroseptal and midseptal manifest accessory pathways (AP's) is challenging due to proximity to HIS region, causing atrioventricular block as complication, and high recurrence due to inadequate lesion delivery. The aim of this study is to present electrophysiologic characteristics and catheter ablation results of these AP's in children at our center.
Methods: We reviewed retrospectively the electrophysiological study and ablation data for anteroseptal-midseptal manifest AP's in our clinic between 2010-2021. All ablation procedurs were performed via cryoablation (8mm and 6mm-tip Freezor MAX, Medtronic Inc., Minneapolis, MN, USA) and with near-zero floroscopy approach using 3-D mapping system (EnSite™, St. Jude Medical Inc., St. Paul, MN, USA).
Results: A total of 104 procedures in 100 patients were included (63 anteroseptal, 37 midseptal). 56 (56%) were male. Mean age was 12,04 ± 4, 10 years (6 months-21years) and mean body weight 44.96 ± 17.88kg (8-88 kg). 20 (20%) patients were asymptomatic and 18(18%) had intermittant AP. The AP was found having risk for sudden cardiac death in 16 (16%) cases and supraventricular tachycardia was induced in 60 patients (60%). Acute success rate was 95/100 (95%). 6mm-tip cryocatheter was used in 59 cases (59%) and 8mm-tip in 41 cases (41%). In one patient the midseptal AP was ablated successfully on left atrial side, requiring transseptal puncture. In 2 patients the noncoconary aortic cusp was also mapped, requiring floroscopy use. No floroscopy was used in the rest (n = 97, 97%). Mean cryoablation lesion number was 6.35 ± 1.77 and mean cryooablation lesion duration was 1902.60 ± 622.34 seconds. Mean procedural duration was 160.06 ± 45.92 minutes (95-330). During the procedures, AV block occured during cryolesions in 15 patients (1. degree in eleven, 2. degree in two and 3.degree in one patient) and right bundle branch block (RBBB) in 7 patients (incomplete RBBB in six and complete RBBB in one patient). One 1. degree AV block and one incomplete RBBB persisted, while the other blocks were temporary, recovering during the procedure. Mean follow-up time was 61.75 ± 39.24 month (2-136). In 11 patients the AP was recurred (recurrance rate: 11%), and 4 of them were ablated successfully later. The cumulative procedure success rate was 99/104 (95.2%).
Conclusion: Cryoablation is an effective and safe method for the treatment of anteroseptal and midseptal AP's, although recurrance rates seem to be a little high.

Keywords: anteroseptal, midseptal, manifest accessory pathway, pediatric, cryoablation

Table-1: Electrophysiologic characteristics of the AP's

AP characteristic	Value/Number	Notes
Location; Anteroseptal/midseptal (n=)	63/37	In one patient left anteroseptal AP detected
APERP, mean±SD [range] (ms) (n=93)	312.04±52.39 [200-360]	In 7 patients APERP couldn't be evaluated due to incessant SVTs, or intermittant AP.
SPERRI during Afib, mean±SD [range] (ms) (n=93)	303.23±49.80 ms (220-408)	in 7 patients Afib could'nt be inducted at all.
Response to IV Adenosine*** (n=82)	Positive in 7 patients Negative in 73 patients Partial response in 2 patients	
AP risk evaluation	No risk for SCD in 84 patients Risk for SCD in 16 patients	
SVT TCL mean±SD [range] (ms)	300.03±50.76 [225-450]	
Intermittant AP (n=)	16	All of them with 'no risk for SCD'

Afib; Atrial fibrillation, AP: Accessory pathway, APERP: Accessory pathway effective refractory period, IV: intarnenous, ms; milliseconds, SCD: Sudden cardiac death, SD: standard deviation, SPERRI; Shortest preexcited R-R interval, SVT: Supraventricular tachycardia, TCL: Tachycardia cycle length

P-254 / Moderated Poster
Clinical course and electrophysiological characteristics of permanent junctional reciprocating tachycardia in children: ten years experience

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Background and Aim: In this study, we aimed to evaluate the clinical features, electrophysiological study, and ablation results of permanent junctional reciprocating tachycardia (PJRT) in children.
Methods: Twenty nine pediatric patients followed up with the diagnosis of PJRT in two pediatric electrophysiology centers between 2011-2021 were included. The basic demographic characteristics of the patients, electrocardiographic and echocardiographic findings were obtained retrospectively from the file records. The medical treatment responses of the patients during the follow-up, the electrophysiological study, and the ablation data of the patients who underwent electrophysiological study were evaluated.
Results: The mean age at diagnosis of the patients was 3.13 ± 4.43(0-18) years and the mean weight was 18.22 ± 19.68kg (3.8-94). 62.1% of the patients were girls. At the time of admission, 11 patients (37.9%) had tachycardia-induced cardiomyopathy (TIC) and their mean ejection fraction was 34 ± 14%. In 15 (51.7%) patients, tachycardia was incessant and all patients except one received medical treatment before ablation. 22 patients were using multidrug therapy (2 or more antiarrhythmics). The most commonly used agents were amiodarone, beta-blockers, class 1 c drugs, and their combinations. A total of 26 ablation procedures were required in 22 patients. The most common indications for ablation were TIC and multidrug-resistant tachycardia. Accessory pathway localization was right posteroseptal in 18 (81.81%) patients, the intracoronary sinus-middle cardiac vein in 2 patients, midseptal in one patient, and posterior-posteroseptal oblique pathway in one patient. The acute procedure success rate was 100% (22/22). The recurrence rate was 18% (4/22), and three of them underwent successful ablation again. The overall success rate was %95.4 (21/22). There were no complications in any of the patients. Cumulative success was 21/22 (95.4%). The mean follow-up period was 4.39 ± 3.05 years. The spontaneous resolution was observed in only 1 of the 7 patients who were followed up clinically with antiarrhythmics, and 6 of the patients are still under follow-up on medical therapy.
Conclusion: Although PJRT is rare, often incessant, resistant to medical treatment, and has a high risk of developing TIC. In such patients, catheter ablation can be performed at any age with low complication and high success rates. It is important to follow the patients in terms of recurrence.

Keywords: Permanent junctional reciprocating tachycardia, Catheter ablation, Supraventricular tachycardia, Tachycardia induced cardiomyopathy

P-255
Comparison of transseptal puncture and pfo route for left-sided accessory pathway ablation in pediatric patients

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Background and Aim: Antegrade or retrograde approaches can be used for left sided accessory pathway (AP) ablation. Because of

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the important arterial complications related to puncture in retrograde approach, some experienced centers prefer to access the left atrium via patent foramen ovale (PFO) or transseptal puncture (TSP). In this study we share our experience with TSP and to compare it with PFO.

Methods: We compare single center data of 322 pediatric patients with left-sided accessory pathway (AP), between 2013 and 2021. Antegrade approach (TSP or PFO) was preferred in all patients. Type of AP, duration of procedure, fluoroscopy time, success, recurrence and complications were compared. Fluoroscopy was used in addition to 3D mapping system in all cases.

Results: The ablation procedures were performed via TSP in 233 (72.3%) cases and PFO in 89 (27.7%) cases. In both groups, 196 (60.9%) of the patients were male. Median age and weight in TSP group were 12.55 ± 3.73 years and 49.36 ± 19.14 kilograms. Median age and weight in PFO group were 11.26 ± 4.79 years and 45.07 ± 21.42 kilograms. In total, 205 (64%) of the patients had manifest, 117 (36%) of them had concealed AP. All TSP procedures were performed successfully, and no major complications were observed, except for one self-limiting pericardial effusion. The most common AP locations were lateral in 111 (%34.4) cases and posterolateral in 80 (24.8%) cases. The mean procedural duration was 141.2 ± 51.69 minutes in TSP cases, and 138.1 ± 52.18 minutes in PFO cases. Mean fluoroscopy duration was 3.99 ± 3.91 minutes in TSP cases and 1.54 ± 3.27 minutes in PFO cases. Number of procedures that considered unsuccessful is 4 in each group (success rate was 98% in TSP and % 95.5 in PFO groups). In follow-ups, recurrence was observed in 7 (3%) TSP and 7 (7.8%) PFO cases. When both groups were compared, no statistically significant difference was found between the success ($p = 0.228$), complication ($p = 0.328$) and recurrence rates ($p = 0.073$) and procedural time ($p = 0.463$); but the fluoroscopy time was found shorter in PFO group ($p<0.0001$) as expected.

Conclusion: Left-sided AP ablation in children can be performed safely via TSP or through PFO in experienced centers. The presence of PFO had no effect on procedure time, success and recurrence rates; but the fluoroscopy time predicted to be slightly shorter.

Keywords: transseptal puncture, patent foramen ovale, manifest accessory pathway, concealed accessory pathway, pediatric ablation

P-256

Low-voltage bridge strategy after first sucessfull cryolesion during tachycardia in children with typical avnrt; comperative study

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Background and Aim: The aim of this comperative study was to evaluate the mid-term efficacy of 8 mm tip cryoablation with / without low voltage mapping of Koch's triangle after first sucessfull cryolesion during typical atrioventricular nodal reentrant tachycardia (tAVNRT) in children.

Methods: From 2013 to December 2021, 204 pediatric patients who underwent cryoablation with 8 mm tip cryocatheter for tAVNRT in our center were included. EnSite™ (St. Jude Medical Inc., St. Paul, MN, USA) was used in all patients. Standard electroanatomical approach without voltage mapping was performed prior to October 2020 (Control group). In the voltage mapping group, Ensite system was used to develop a "bridge" of lower voltage gradients (0.2-0.8 mV) of the inferoposteroseptal right atrium to guide cryoablation. Patients who had congenital heart disease, other arrhythmia substrates and prior ablation attempts with either RF or cryoablation were excluded. First cryoablation lesion was applied during tachycardia in all patients.

Results: 204 patients were included (58 low voltage, 146 control). There was no difference between groups with regard to age (13.86 ± 2.78 vs 13.6 ± 2.77; p: 0.559), gender (38 / 65.5% vs 79 / 54.1%; p: 0.159), or catheter-tip size (8 mm). Fluoroscopy was not used in any procedure. Total procedural time was similar in voltage and control groups (135.8 ± 30.2 min vs 138.68 ± 33.61 min; $p = 0.581$). Acute success was similar in both groups (100% Voltage vs 99% Control; P = 0. 0.991) The overall recurrence rate were similar (1, [1.7%] and 2, [1.4%]; p:0.380) Follow-up time was shorter in the Voltage group (5.98 ± 3.66 months vs 42.3 ± 18.01; p:0.001) There was no major or minor complications were reported during or after the procedures in either group.

Conclusion: Use of a slow pathway voltage map in Koch's triangle with 8 mm tip criyo ablation catheter is a safe and effective method in children with tAVNRT. Although there is no significant difference between both groups in acute success rate, procedure duration, cryolesion number, and recurrence rate, a longer follow-up period is needed to evaluate the long-term results.

Keywords: Low voltage bridge mapping, cryoablation, AVNRT, children

	Control(n:146)	Low voltage(n:58)	P value
Mean age (years)	13.6±2.77	13.86±2.78	0.559
Mean weight(kg)	51.5±12.17	53.5 ± 11.86	0.270
Gender (n= females, %)	79 / 54.1%	38 / 65.5%	0.159
Total number of lesions	7.8±2.02	7.5±1.78	0.380
Procedure time (min)	138.68±33.61	135.8 ±30.2	0.581
Fluoroscopy time (min)	0	0	
Acute Success (n, %)	145/146 (99%)	%100	0.991
Complications	0	0	
Recurrences (n, %)	2(1.4%)	1(1.7%)	0.380
Follow-UpTime (months)	42.3 ±18.01	5.98 ± 3.66	0.001

Figure 1 Patient Characteristics, Procedural and Long-Term Outcomes

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P-257 / Moderated Poster

Cardioneuroablation in a pediatric patient with type IIB cardioinhibitory vasovagal syncope and functional advanced AV block

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Background and Aim: In severe vasovagal syncopy (VVS) with long sinüs pauses or AV blocks, back-up cardiac pacemaker therapy should be considered. Nowadays transcatheter cardioneuroablation (CNA) is comming into prominance, especially in young patients. We present a pediatric case successully treated with transcateheter CNA.
Methods: CASE: A seventeen years old girl admitted to our clinic with frequent VVS attacks. 12-lead ECG was normal. On 24-hour ambulatory-ECG frequent 2.degree mobitz-type-1 and 2:1 AV blocks,and also advanced 2.degree AV blocks, up to 8,9 seconds of ventricular pause were detected. Tread-mill exercise test and echocardiography revealed no pathology. During tilt testing the patient had an type IIb cardioinhibitory type VVS with 8 second sinuse pause. Atropin challenge testing revealed >30% increase in heart rate (from 75 bpm to 140 bpm). We decided to perform CNA.
The procedure was performed under general anesthesia.EnSite Precision 3-D mapping system (EnSite Precision,Abbott, Chicago, Illinois) and limited floroscopy was used during procedure. After transseptal puncture high density multipolar mapping catheter (Advisor HD Grid, Abbott, Chicago, Illinois) through a 8,5f steerable long sheath (Agilis, Abbott, Chicago, Illinois) was used to mapp left atrium, left- and right upper pulmonary vein (LUPV-RUPV) orifices, right atrium around superior vena cava(SVC) for high amplitude fractional electrogram (HAFE) and low amplitude fractional electrogram(LAFE) signals from epicardial parasympathic ganglions. Using an 8f contact-force irrigated radiofrequency catheter (TactiCath, Abbott, Chicago, Illinois) lesions with 20-30 watt 34-36°C 107-110ohm and 8-10gram contact force were given to marked points, by following a 15% decrease in impedance and until the HAFE and LAFE signals disappear, showing effectiveness of the lesions. While a slow junctional rhythm developed during lesion delivery on LUPV, sinus tachycardia was seen on RUPV, carina and SVC-right arial junction. After 24 lesions, each for 30 seconds, control atropin challenge test was negative and the procedure was ended with success and without complication.
Results: Two weeks later, control Tilt testing and 24-hour ambulatory-ECG revealed normal results, and the patient is still asymptomatic under follow-up after 3 months of ablation.
Conclusion: CNA is now an alternative treatment to cardiac pacemaker therapy in severe type IIb cardioinhibitory type VVS, especially in selected young patients.

Keywords: Vasovagal syncope, cardioneuroablation, parasympathic ganglion, pediatric ablation

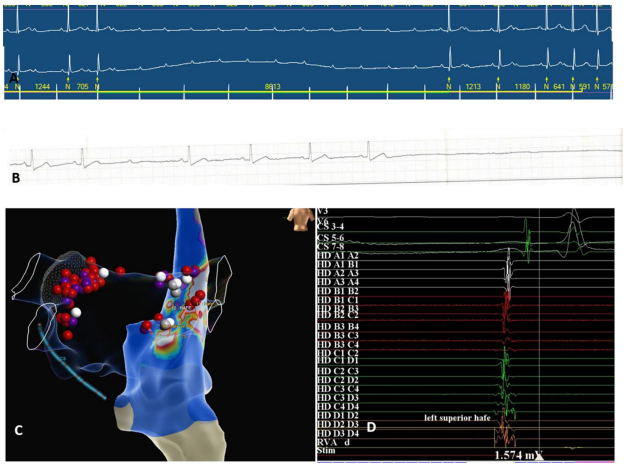

Figure 1. A) Preprocedural 24-hour ambulatory-ECG revealing advanced AV block with a ventricular pause of 8,9 seconds. B) Preprocedural tilt testing revealing 8 seconds sinus pause with type IIb cardioinhibitory type VVS. C) Marked points on HAFE and LAFE lesions detected on left atrium, left and right upper pulmonary vein orifices, right atrium and septal side of superior vena caval orifice seen on 3-D electroanatomic mapping (EnSite) D) HAFE signal on left upper pulmonary vein orifice, targeted for irrigated radiofrequency ablation.

P-258

Arrhythmia / Electrophysiology

Life-threatening rhythm and conduction disorders by a child with MIS-C

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Background and Aim: Cardiac involvement is seen in the majority of cases with multisystem inflammatory syndrome in children (MIS-C). Various rhythm and conduction disturbances, as well as repolarization abnormalities, have been described by more than 50% of the patients, while there are few cases with complete heart block or with asystole.
Methods: Case report
Results: 8-year old girl presented with a 5-day history of fever, cough, headache, and abdominal pain. Because of the critical condition, with respiratory insufficiency and heart failure symptoms, the child was intubated and started on inotropic support. ECG showed complete AV-block with a ventricular rate of 75/min

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and with ST-T changes; echocardiography revealed dilated left ventricle with reduced contractility, CT-scan of the lungs showed bilateral pneumonia, the inflammatory markers were elevated, in combination with high troponin levels, and positive SARS-CoV2-IgG antibodies. The diagnosis MIS-C was made and treatment with immunoglobulins, antibiotics, corticosteroids, and anticoagulants was initiated.
During the next 2 days, the cardiac function deteriorated further, and while still on mechanical ventilation and inotropic support, extreme bradycardia with a ventricular rate of 35/min was registered, and the patient was indicated for temporary emergency pacing. Upon induction of anesthesia, the child became asystolic, requiring extensive resuscitation. After circulation recovery, the ECG showed nodal tachycardia with a heart rate of 140-170/min. A temporary transvenous pacemaker (PM) was inserted, and the patient was started on intravenous amiodarone which resulted in a slower ventricular rate of 70/min. 3 days later sinus rhythm was restored, with first-degree AV-block, which allowed removal of the PM 5 days after its insertion. Left ventricular dimensions were normalized and contractility remained low-normal (EF 56%). During the 6-month follow-up, the ECG and the Holter-monitoring showed sinus rhythm with first-degree AV-block. Magnetic resonance imaging (MRI) on day 15 of the hospital stay demonstrated scattered areas of myocarditis and ischemia predominantly in the left ventricle, as well as thickening of the basal septum. Six months later the MRI changes were reduced but still persistent.
Conclusion: MIS-C can present with serious and life-threatening rhythm and conduction disturbances in children; this is why extensive cardiac monitoring is obligatory by all patients.

Keywords: COVID-19, MIS-C, complete AV-block

Heart failure

P-259
Establishing a cardiomyopathy registry in an african country:preliminary results

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Background and Aim: Data about pediatric CMP in Africa are limited. This study aims to describe clinical and echocardiographic (echo) patterns and short-term outcomes of pediatric CMP from a hospital registry.
Methods: A CMP registry was initiated at 2 pediatric cardiology referral centers in Khartoum in 2018 including all children 0-18 years with an echo diagnosis of CMP. A retrospective study was carried from Jan 2013 to January 2019 and a prospective part was carried from January 2019 to December 2021. Clinical and echo data were collected on presentation and on at least one follow-up. Genetic studies were conducted in selected families.
Results: : A total of 206 patients were diagnosed with CMP during the study period. Most patients (48%) presented at 1-6years of age, neonatal CMP was present in 4%. Familial incidence was detected in 11% of cases. Heart failure was the most common mode of presentation in 96% of patients; 65% needed hospitalization; a third of them needed intensive care unit admission. The most common type of CMP was the dilated type (67%), others include noncompaction CMP (19%), restrictive CMP and hypertrophic CMP (both of them in 6%) of patients. Hospital mortality was 20% for the whole cohort and 100% for neonates. Other complications occurred in 21% of patients including arrhythmias, intracardiac thrombi (Figure) and cerebrovascular accidents. Clinical and echo follow for a mean period of 6 months showed that most patients (52%) remained the same, 26% worsened and 21% improved. Genetic studies were conducted in 3 families and revealed Pompe's disease, an autosomal dominant dilated cardiomyopathy type 1EE and autosomal recessive familial hypertrophic CMP type 8.
Conclusion: Pediatric CMP imposes a high burden on hospital resources and yet has a high mortality. Genetic and metabolic studies that could help to improve management and outcomes are needed.

Keywords: Cardiomyopathy, Africa

P-260
Discriminative ability of irisin and apelin in prediction of heart failure phenotypes in type 2 diabetes mellitus patients

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Background and Aim: Irisin and apelin are autocrine regulators of cardiac and vascular reparation. The aim of the study was to investigate whether serum levels of both irisin and apelin predict HF with preserved ejection fraction (HFpEF) in patients with type 2 diabetes mellitus (T2DM)
Methods: One hundred and eight HF patients with T2DM having HFpEF (n = 58), HF with mildly reduced ejection fraction (HFmrEF; n = 22), HF with reduced ejection fraction (HFrEF, n = 28) aged from 41 to 62 years and 20 non-HF patients with T2DM. Measurement of irisin, apelin, N-terminal pro-brain natriuretic peptide (NT-proBNP) by ELISA was.performed at baseline.
Results: The levels of irisin were significantly higher in HFpEFto HFrEF, whereas healthy volunteers and T2DM non-HF patients demonstrated lower concentrations of these peptides. Apelin levels were significantly increased in HF patients mainly with HFrEF. There were not significant differences between the levels of these biomarkers in HFrEF and HFmrEF (P = 0.42 for all cases). Using ROC curve we revealed that cut-off points for irisin and apelin that distinguished HFpEF from HFrEF/HFmrEF were (6.50 ng/mL; AUC = 0.78; 95% confidence interval [CI] = 6.85 - 10.66 ng/mL and 4.12 ng/mL, AUC = 0.72; 95% CI = 3.90-5.75 ng/mL, respectively). Then we divided all patients with HF having elevation of NT-proBNP > 750 pmol/mL into three subgroups depending on the biomarkers' levels. Patients from subgroup A had both irisin and apelin levels higher cut-off points, individuals from group B had higher concentration of one of two biomarkers, and patients from subgroup C demonstrated levels of both peptides lower cut-off points. Multivariate logistic regression analysis revealed that discriminative value of irisin and apelin to predict HFpEF in subgroup B (Odds Ratio [OR] = 2.18; 95% CI = 1.26-3.14; P = 0.001) were substantially higher compared with subgroups A and C (OR = 1.03; 95% CI = 1.00-1.05; P = 0.64 and HR = 0.92; 95% CI = 0.89-1.01; P = 0.62, respectively). Adding irisin and apelin to NT-proBNP as independent variables to the predictive model sufficiently improved discriminative ability of whole model for HFpEF.
Conclusion: We found that multidirectional changes in the levels of irisin and apelin in T2DM patients had better predictive value for HFpEF that simultaneous increase and decrease in the circulating levels of these peptides.

Keywords: Heart failure; type 2 diabetes mellitus, irisin, apelin, biomarkers.

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P-261

Sacubitril/valsartan treatment for pediatric heart failure

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Background and Aim: Sacubitril/valsartan is the first agent approved in a new class of drugs called angiotensin receptor neprilysin inhibitor. The medication is FDA-approved to treat patients with chronic heart failure with reduced ejection fraction with New York Heart Association (NYHA) class II, III, or IV. Recently, ACEI, ARB, or ARNI are now recommended in patients with chronic symptomatic heart failure with reduced ejection fraction (HFrEF) to reduce morbidity and mortality (class I recommendation). However, there are insufficient data on the use of sacubitril/valsartan in the childhood age group. This study aims to share our experience on sacubitril/valsartan treatment in ARVD patients.
Methods: ARVD patients who have initiated sacubitril/ valsartan treatment from January 2020 to December 2021 were included in our study. ARVD patients with a systemic RV ejection fraction of ≤35% and symptomatic despite treatment with β-blocker and ACE-inhibitor were started on sacubitril/valsartan. Renal function, NT-pro-BNP and serum potassium levels were monitored at the initiation of therapy. A 6-minute walking test was performed for all patients. If the medication was well tolerated, the dose of sacubitril/valsartan was increased stepwise until the highest tolerated dose was reached. During the study period, echocardiography, a 6-minute walking test, laboratory investigation including hemoglobin levels, kidney function, electrolytes and NT-pro-BNP were repeated in every outpatient clinic visit.
Results: A total of 6 patients with ARVD who were initiated sacubitril/valsartan and followed for six months were enrolled in the study. Four of them were boys, 2 were girls, and the mean age was 8.4 years. Four of the patients were NYHA class II, and 2 were NYHA class III. The mean RV ejection fraction was 21%, and LV ejection fraction was 35%. N-terminal pro-B-type natriuretic peptide decreased significantly from an average of 11370 to 4830. The average 6-minute walking distance improved from 240 to 376 meters. Sacubitril/valsartan significantly improved the NYHA class. Adverse effects were not detected during the study period.
Conclusion: is study suggests that sacubitril/valsartan may be an effective and safe strategy to improve the clinical symptoms and reduce HFrEF hospitalization in patients with ARVD.

Keywords: heart failure, sacubitril, valsartan, congenital heart disease, pediatric

P-262

Safety and tolerability of SGLT-2 inhibitor in pediatric refractory heart failure listed for transplant- a case report

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Background and Aim: Paediatric heart failure (HF) is not an uncommon entity. It is characterised by ventricular dysfunction, and either volume or pressure overload, alone or in combination. The two most common pathophysiological entities resulting in end stage HF in children are cardiomyopathy and congenital heart disease (CHD). The aim of this case report is to show the effect of sodium-glucose co-transporter inhibitors(SGLT-2i) in addition to standard HF therapy including angiotensin converting- enzyme (ACE) inhibitors or Angiotensin Receptor-Neprilysin Inhibitor (ARNi), mineralocorticoid receptor antagonists (MRA) and a β-adrenergic receptor antagonist (β-blocker).
Methods: We present a case of a 11 year old boy with refractory HF for four years who was referred for cardiac transplantation. He presented with symptoms of Class IV dyspnoea with severe Bi-ventricular dysfunction and left ventricular Ejection fraction(EF) of 8-10%. Right ventricular(RV) Tricuspid annular plane systolic excursion (TAPSE) was 10mm and N-terminal-pro hormone BNP (NT-proBNP) was 16,000 pg/ml. He had associated mild Budd-chiari syndrome with good collateralisation and normal liver function. He was stabilised with intravenous milrinone and diuretics. Sacubitril-Valsartan and beta-blockers were also added eventually. EF remained at 20%. Patient continued to be symptomatic with class III dyspnea inspite of high dose diuretics and maximum tolerated doses of ARNI, MRA and β-blockers. As a last resort before listing for transplant, he was started on SGLT-2i dapagliflozin 5mg as an off-label indication which was later increased to 10mg. Precautions to prevent hypoglycaemia and genital infections were taken.
Results: The child improved symptomatically from class IV to class II dyspnea. His LVEF improved to 30%, RV TAPSE to 20mm and his NT-proBNP reduced from 16,000 to 8,000 pg/ml. The diuretic dosage was also significantly reduced. At three months his effort tolerance and walking distance improved and he is even able to go shopping with his parents. The patient tolerated dapagliflozin well with no tolerability and safety issues.
Conclusion: The SGLT-2i dapagliflozin can be considered in children who are on maximum tolerated dose of standard guideline directed medication as an off-label indication with adequate precautions. To the best of our knowledge this is the first case of SGLT-2i in refractory pediatric HF.

Keywords: Sodium-glucose co-transporter inhibitors, Heart failure, Paediatric population.

P-263 / Moderated Poster

Systemic small vessel disease following pediatric heart transplantation

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Background and Aim: Cardiac allograft vasculopathy (CAV) is an important predicrtor of outcome in patients following pediatric heart transplantation (pHTX). It is standard of care at most pediatric heart transplant centers to perform regular coronary angiograms to detect early coronary artery changes and adjust medical management accordingly. It was the aim of this study to characterize systemic arterial characteristics in patients following pHTX compared to healthy controls.
Methods: Patients following pHTX (n = 39) and controls (n = 31) were recruited prospectively. Common carotid intima media thickness (cIMT) was analyzed with ultrasound. Central blood pressure, central augmentation index corrected to a heart rate of 75/minute (cAIx75) and carotid-femoral pulse wave velocity (PWV) were measured (SpygmoCor XCEL). Reactive hyperemia index (RHI) as a measure of endothelial function and peripheral

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AIx75 (pAIx75) were determined (EndoPAT). Retinal vessels were assessed quantitatively using the arteriolar-to-venular ratio (AVR; iMedos). Data are expressed as mean +/- standard deviation and number (percent) as appropriate. Group analyses were performed using the student t-test and Fisher exact test, respectively.
Results: Mean age overall was 19+/-7 years. PHTX patients were evaluated 12+/-7 years post transplant. The groups were of similar age ($p = 0.523$) and sex (58% female, $p = 0.808$). Brachial blood pressure as well as central pressure were significantly higher in pHTX patients (central mean pressure 97+/-11 vs 88+/-10 mmHg, $p<0.001$). However, there was no significant difference in cAIx75 ($p = 0.291$) or PWV ($p = 0.440$). PAIx75, though, which evaluates arterial wave reflection peripherally, was significantly increased in pHTX patients (-3+/-11 vs -14+/-10, $p<0.001$). Likewise, there was evidence of microvascular impairment based on a significantly decreased retinal AVR (0.82+/-0.06 vs 0.88+/-0.05, $p<0.001$). Interestingly, AVR had a moderate correlation with pAIx75 ($r = -0.375$, $p = 0.009$) but not with blood pressure parameters (all $p>0.05$). Focusing on endothelial characteristics, neither cIMT nor RHI were significantly different between the groups.
Conclusion: Patients following pHTX appear to have systemic small vessel disease, but there is no evidence of large arterial stiffening, abnormalities in cIMT or endothelial dysfunction. Further analyses will determine whether AVR and pAIx75 may serve as non-invasive predictors of CAV.

Keywords: Cardiac allograft vasculopathy, pediatric heart transplantation, endothelial function, arterial stiffness, microvasculature

P-264

The opportunities and difficulties of the bridge to transplantation in pediatric cardiac transplantation

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Background and Aim: The mechanical support of circulation was implemented parallel with the onset of the paediatric cardiac transplantation program in Hungary in the year 2007. The first implantation of a long-term assist device (BerlinHeart) took place in 2008. We present our clinical experience in line with the data published in the literature over the corresponding time interval.
Methods: Over the considered period (2007 to 2021) 25 of the total of 57 paediatric patients were supported mechanically prior to the transplantation. Age at initiation of the device implantation was 9ys2mo, with a mean weight of 36 kg. The median duration of pre-Tx mechanical support was 194 days, their median post Tx follow-up being 45 months, summing up a total of 1179 patient-months. The first period of the program was characterized by frequent use of biventricular assist devices, as the patients were considered for this therapy in very advanced stage of their underlying cardiac disease (INTERMACS 1-2) In the most recent period the implantations were effectuated more and more in a planned, elective manner (INTERMACS 2-3), and as a consequence LVADs were resorted to almost exclusively. Pulsatile devices in patients over 20kg weight were progressively replaced by continuous flow devices, which also led to a significant decrease of the overall complication rate.
Results: At a low overall mortality rate (8 %), despite strict adherence to anticoagulation and antiseptic protocols, the cumulative haemorrhagic, thrombo-embolic and infection related complication rates (42/4/19%) were not trivial, but the numbers were in line with data of the PEDIMACS registry.
Conclusion: VAD became integral part of transplantation programs worldwide. By applying it in 43% of our patients listed for HTx, the reduction of the waiting-list mortality was impressive. Our results must be viewed through the optics of reduced number of patients and limited follow-up, a shortcoming which also characterizes the majority of the data from the literature.

Keywords: transplantation, heart failure, mechanical circulatory support, VAD

P-266

Mortality and heart disease in children at a reference center

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Background and Aim: Death from congenital heart disease(CHD) represents 6.4% of global infant mortality, but causes of death in children with cardiac disease remain scarcely reported.
To describe distribution of causes of death in children with heart disease in a single institution, and to estimate evolution of mortality rates over 11 years(2010-2020).
Methods: Data on circumstances of death were retrieved from medical file. All patients with heart disease and aged 0 to 18 years at death were included. Mortality rates are presented by type of heart disease and by immediate cause of death. Evolution of mortality rates per underlying cause over the study period was analyzed.
Results: 929 patients died over the study period: 704 (76%) CHD, 147 (16%) cardiomyopathies, 40 (4%) pulmonary arterial hypertensions (PAH), 20 (2%) isolated arrhythmias or conduction disorders, 3 cardiac tumors, 3 post-traumatic heart contusions, 5 systemic hypertensions, 6 sudden unexplained deaths and 1 infectious endocarditis. 186 (20%) neonates who received primary compassionate care were excluded from subsequent analyses. For the remaining 743 patients, age at death was younger than one year for 479 (64%) patients, and mortality related to CHD remained significantly higher until the age of 12 years. Approximately 50% of the cardiac mortality occurred within 30 postoperative days for CHD with 58% of children having complex CHDs. Causes of death were heart failure (39%), PHA crisis (12%), sepsis (11%), sudden death (9%), respiratory failure (8%), neurological impairment (6%), arrhythmia or conduction disorder (4%), hemorrhage (3%) and thrombotic event (3%). Heart failure was also the leading cause of death in four selected CHD: transposition of the great arteries, tetralogy of Fallot, functionally univentricular heart, and aortic coarctation (32%, 34%, 39% and 46%, respectively). There was no significant evolution of global or selected mortality rates over the study period except for mortality of hypoplastic left heart syndrome that decreased significantly (40% in 2010 vs 20% in 2020).
Conclusion: Postoperative early mortality in neonates and infants with complex CHDs represents the major cause of cardiac death in this tertiary center cohort. A tenth of deceased patients experienced sudden cardiac death. Prevention of such event and efforts on management of acute and chronic heart failure and pulmonary hypertensions are crucial to reduce cardiac mortality in children.

Keywords: CHD mortality, heart failure, sudden death,

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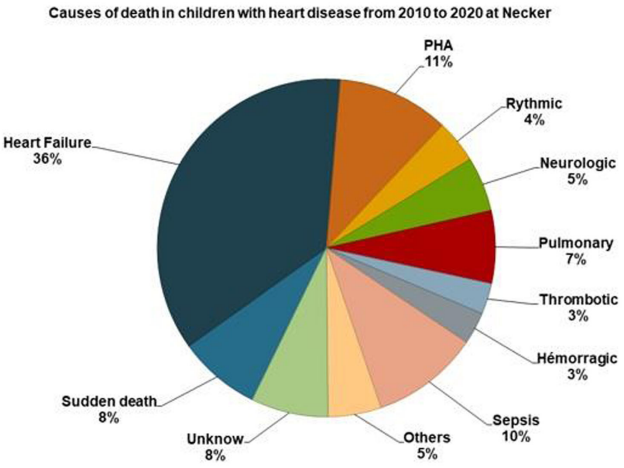
Cause of death



P-270

Right ventricular wall tension in the assessment of pediatric pulmonary arterial hypertension

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Background and Aim: Echocardiographic determination of the right ventricular wall-tension (RVWT) is a novel parameter for the assessment of pulmonary hypertension (PH) in adults. We aimed to investigate RVWT in pediatric PH patients, and to correlate RVWT data with invasively obtained hemodynamics, echocardiographic variables, NT-proBNP, and NYHA functional class (FC).
Methods: Prospective echocardiographic study in two independent cohorts of children with PH and age-and gender-matched control subjects (Pediatric PH patients: n = 49; matched controls: n = 49).
Results: RVWT was increased in children with PH as compared to healthy controls ($p<0.01$). Higher RVWT values were associated with less favorable invasive hemodynamics, i.e. increased mean pulmonary artery pressure (mPAP) and pulmonary to systemic vascular resistance ratio (PVR/SVR). RVWT values also increased with worsening NYHA-FC and increasing plasma NT-proBNP levels. In addition, RVWT values correlated with other echocardiographic variables, such as pulmonary artery acceleration time (PAAT), tricuspid annular peak systolic excursion (TAPSE), RV/LV dimension ratio, and left ventricular eccentricity index (LVEI).
Conclusion: RVWT is a novel and easily obtained bedside indicator of RV remodeling and dilatation. Higher RVWT values were associated with lower FC, and worse hemodynamics, demonstrating that this index provides additional information on right ventricular performance in the setting of increased RV pressure load in pediatric PH.

Keywords: Echocardiography, Pulmonary Hypertension

P-271

The level of superoxide dismutase and catalase in acyanotic congenital heart disease in children with heart failure

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Background and Aim: The most common complication in acyanotic congenital heart disease (CHD) is heart failure which definitive diagnosis and therapy remain unsatisfactory. Heart failure's progression is often associated with oxidative stress process. Superoxide dismutase (SOD) is the first line antioxidant of defense against superoxide anion. While Catalase (CAT) breaks down hydrogen peroxide into water and oxygen molecules which complements previous detoxification carried out by SOD.
OBJECTIVE: This study is aimed to compare the differences of SOD and CAT levels in acyanotic CHD patients between those with and without heart failure.
Methods: A case-control study was conducted on three to ten years old children with a left-to-right shunt acyanotic CHD with and without heart failure in the Pediatric Cardiology outpatient clinic, ward, and emergency room of Dr. Soetomo Hospital Surabaya from April-July 2020. Echocardiography examination was used to establish the diagnosis of CHD, while Ross' Modified Pediatric Heart Failure Score (PHFS) criteria was used to indicate heart failure. T-test was undertaken for analyzing the difference between both groups.
Results: The total samples were 41 children, consisted of 29 subjects in the case group (CHD with heart failure) and 12 subjects in the control group (without heart failure). The level of SOD in CHD with heart failure was lower (74.670 U/ml +- 15.705) than those without it (109.163 U/ml +- 3.111) ($p<0.05$). In contrast, level of CAT in CHD with heart failure was higher (25.895 μM) than those without it (13.976 μM) ($p<0.05$).
Conclusion: There was a significant difference of SOD and CAT levels in acyanotic CHD between those with and without heart failure.

Keywords: SOD, CAT, acyanotic CHD, heart failure, oxidative stress

P-272

Use of bromocriptine in two adolescent cases with peripartum cardiomyopathy

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Background and Aim: Peripartum cardiomyopathy (PPCMP) is a rare cause of heart failure characterized by left ventricular (LV) systolic dysfunction that occurs in the last month of pregnancy or up to the fifth month postpartum.
Methods: We present the use of bromocriptine in the treatment of postpartum cardiomyopathy in two adolescent cases in this report.
Results: Case 1: A 15-year-old girl presented with complaints of dispnea and swelling in her legs on the second postpartum day.

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Her physical examination revealed tachypnea, tachycardia, rales in the bilateral lung bases, 2/6 pansystolic murmurs at the apex and pretibial edema. Diffuse LV hypokinesia (Ejection fraction (EF): 30%, shortening fraction (SF): 15%) and dilatation (Diastolic inner diameter: 54 mm, Z score: +2.7), increased wall thickness, mitral regurgitation, pericardial effusion (PE) in echocardiography. On cardiac MRI, marked hypokinesia and fibrosis in inferolateral wall and apex were detected. LV dilatation and function improved significantly (Diastolic inner diameter: 45 mm, Z score: -0.12, EF: 60%, SF: 31%) with the use of bromocriptine (1x2.5 mg) in addition to the standard heart failure treatment for two weeks. In the third month of the follow-up, she did not have any complaints and LV systolic function persisted to be normal.

Case 2: A 17-year-old girl, who had complaints of dispnea, fatigue at the sixth hour of postpartum and was referred to us for PE detected in the thorax CT scan, had signs of heart failure in the physical examination. Echocardiography revealed LV systolic dysfunction (EF: 43%, SF: 22%) and mild PE. In addition to these findings, mild thickening of the interventricular septum (Septum: 12.5 mm, Z score +1.64) was observed on cardiac MRI. Her heart returned to normal by the first month of follow-up with bromocriptine treatment and LV systolic function was normal at the sixth month.

Conclusion: It was aimed to draw attention to the effect of bromocriptine on heart failure findings and improvement of left ventricular systolic disfunction in adolescent patients with PPCMP. Early diagnosis and use of bromocriptine in the treatment of PPCM eliminates the need for mechanical circulatory support and/or heart transplantation, and reduces morbidity and mortality.

Keywords: adolescent, bromocriptine, cardiomyopathy, peripartum

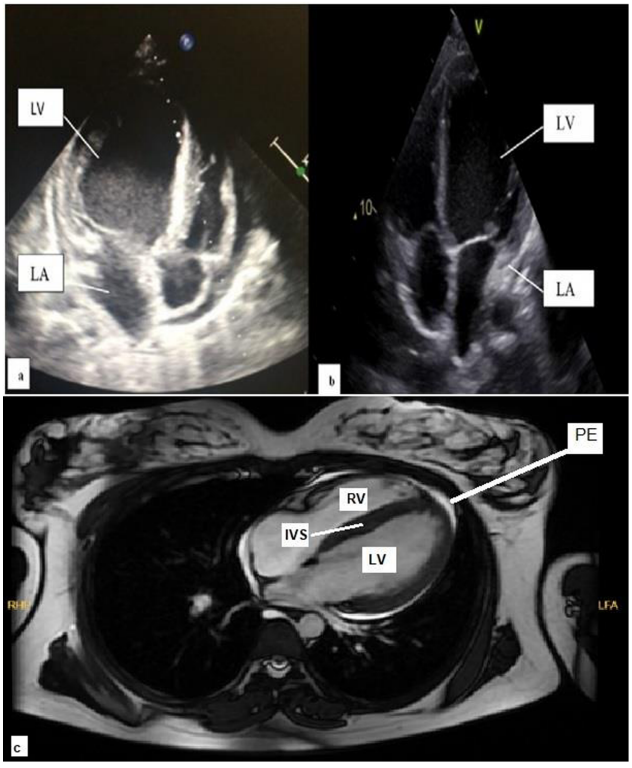

Figure 1. Echocardiographic images of the case 1 before treatment with bromocriptine (a) and two weeks after bromocriptine treatment (b), cardiac MRI image of the case 2 (c). (LA: left atrium, LV: left ventricle, RV: right ventricle, IVS: interventricul

P-273

Clinical features and treatment of mixed cardiogenic and distributive shock in patients with multisystem inflammatory syndrome in children (MIS-C)

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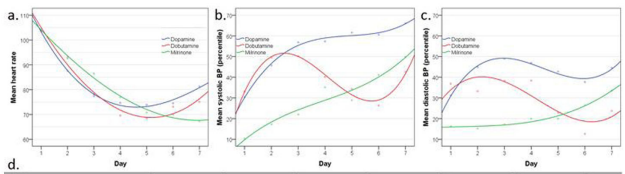
Background and Aim: Mixed shock in multisystem inflammatory syndrome in children (MIS-C) associated with COVID-19 is consequence of acute heart failure, inflammation-induced vasodilation and potential volume loss.

Methods: Retrospective analysis included 25 patients (7 girls) with MIS-C-related combined shock, treated in period from April 2020 to December 2021.

Results: Mean age of patients was 12.6 ± 4.0 years. Admission was 6.1 ± 1.6 days after symptoms onset. Systemic inflammatory response was manifested with neutrophilia (10.7 ± 4.2 x109/), lymphopenia (1.1 ± 0.7 x109/L), elevated CRP (220.9 ± 86.1 mg/L), ferritin (684.5 ± 549.5 μg/L) and D-dimer (1528 ± 1254 ng/mL). One third of patients had acute kidney injury with glomerular filtration rate of 64 ± 22 mL/min/1.73 m2 and urea level of 16.0 ± 8.4 mmol/L. All patients had acute heart failure with ejection fraction 47.2% ± 7.7% and fractional shortening 23.6% ± 4.9%, 92% of patients had NTproBNP >1500 pg/mL and 58% had elevated troponin I (1.34 ± 1.47 ng/mL). Z-scores for end-diastolic left ventricle, interventricular septum and posterior wall diameters were 0.7 ± 1.1, 1.7 ± 1.3 and 0.6 ± 0.7 respectively. All patients had mild/moderate mitral regurgitation, and 60% had mild pericardial effusion. Inotropes, administered during first 3.7 ± 1.6 days, were divided in three groups: 1) dopamine (n = 14), 2) dobutamine + dopamine (n = 5), 3) milrinone ± dopamine (n = 6). Additional treatment included diuretics and captopril. Total fluid balance (including insensible loss of 300 mL/m2/day) through days 1-7 was +860 mL/m2, +128 mL/m2, -108 mL/m2, -36 mL/m2, -306 mL/m2, -335 ml/m2, -298 ml/m2 (total -95 ml/m2). Methylprednisolone/intravenous immunoglobulin and low-molecular-weight heparin/acetylsalicylic acid were administered and fever persisted 1.2 days averagely. Oxygen supplementation was needed in 71% of patients. Transitory bradycardia was noticed and there was no difference in heart rate between treatment groups. Profound hypotension was revealed on admission and correction differed regarding treatment ($p < 0.05$) (Figure 1). All patient survived with clinical improvement (one had mechanical ventilation, and one had stroke).

Conclusion: Mixed shock is the most severe manifestation of MIS-C, and treatment of heart failure should be combined with cautious fluid resuscitation.

Keywords: mixed shock, MIS-C, inotropes, fluid balance



Day	1	2	3	4	5	6	7
Heart rate	104 ± 13	90 ± 15	80 ± 15	74 ± 13	72 ± 13	73 ± 8	77 ± 11
SBP (mmHg)	99 ± 11	105 ± 10	109 ± 9	110 ± 10	110 ± 10	110 ± 10	113 ± 10
DBP (mmHg)	54 ± 10	58 ± 11	60 ± 9	59 ± 9	58 ± 7	56 ± 8	59 ± 8

Figure 1. Differences in mean heart rate (a), systolic (b) and diastolic blood pressure (c) among three treatment groups. Daily average heart rate, systolic and diastolic blood pressure in all patients (d).

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P-274
Catheter-based treatment of pulmonary embolism after pneumectomy in a child with univentricular physiology

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Background and Aim: Pulmonary artery stump thrombosis is a well-known complication after pneumectomy, as well as thromboembolic events in patients with superior cavopulmonary connection (SCPC). Management protocols are lacking, especially for antithrombotic therapy in children.
Methods: A 7-year-old boy with right isomerism syndrome with complete non-balanced AVSD was referred for evaluation before total cavopulmonary connection (TCPC). At the age of 15 months, he underwent surgical correction of a partial abnormal pulmonary return of both right pulmonary veins, as well as a SCPC. Cardiac catheterization showed correct drainage of the superior caval vein to the right pulmonary artery (RPA). Left pulmonary artery (LPA) had retrograde perfusion from multiple aorto-pulmonary collaterals. Left pulmonary veins were totally occluded. In the meantime, several collaterals were embolized in preparation surgical left pneumectomy. After left pneumectomy, he was treated with Sildenafil and low molecular weight heparin LMWH (Nadroparine). LMWH was switched to Aspirin 10 days later. On post-operative day 12, he developed acute respiratory distress with severe hypoxia (SpO2 40%). Angiography showed multiple thrombi in the right superior and inferior lobe and an important thrombus in the left stump of the ligatured LPA. LPA stump was totally occluded proximally by a 16mm vascular plug, to exclude the thrombus and the thrombi on the right side were aspirated. At the end of the procedure, pulmonary pressure dropped from 20 to 13mmHg. LMWH was relayed by Xarelto 2 weeks after percutaneous thrombectomy. 4 weeks later, pulmonary pressure remained low and angiography showed homogenous perfusion and drainage of the right lung, permitting TCPC.
Results: Antithrombotic therapy after pneumonectomy in patients with CHD is not standardized. Pulmonary embolisms (PE), typically originating from thrombi in the contralateral stump, are life-threatening in this population. The risk of thromboembolisms is even higher in univentricular physiology where decreased flow and absence of pulsatility characterize the pulmonary vasculature. Percutaneous treatment of massive PE by endovascular embolectomy is feasible. Moreover, we showed that occluding the contralateral pulmonary artery stump could avoid recurrence.
Conclusion: Patients after SCPC and pneumectomy are at high risk of PE. Adequate anticoagulation therapy is mandatory. We showed that endovascular embolectomy can be life-saving.

Keywords: pulmonary embolism, pneumectomy, superior cavopulmonary connection, percutaneous thrombectomy

Image A

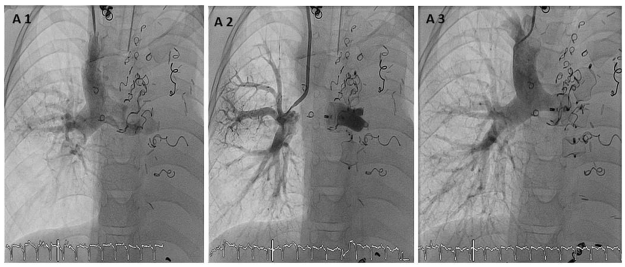

Figure 1 A. Angiography after contrast injection in SCV after SCPC. A-1 Multiple thrombi in the right superior and inferior lobe and a important thrombus in the left stump of the ligatured LPA. A-2 Result after aspiration of thrombi on the right side and occlusion of LPA stump by a 16mm vascular plug, to exclude the giant thrombus. A-3 Four weeks later, control angiography showing homogenous perfusion with no recurrence of long embolism.

P-275
High incidence of primary cardiomyopathy features in children and adolescents with marfan syndrome

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Background and Aim: Marfan syndrome (MFS) is an inherited connective tissue disorder that causes serious cardiovascular complications. In addition to secondary cardiac manifestations, such as valvulopathies, a possible primary cardiomyopathy is still under debate. In a previous retrospective analysis we found signs of cardiomyopathy in about 50% of pediatric MFS patients in our clinic. This is a first interim analysis of our prospective validation study looking at characteristics that could be associated with an increased risk of cardiomyopathy.
Methods: In a single center, prospective, observational cohort study we are currently enrolling children and adolescents with secured MFS, and collecting prospective data from their electronic medical records, physical exam, echocardiography, and genetic evaluation. The patients in this group were defined to have a cardiomyopathy if they had any one of the following: a cardiac index (CI) below the age-adjusted lower limit of normal (LLNaa), an ejection fraction (EF) below 50%, diastolic dysfunction (abnormal E/E' for age), or clinical symptoms of CHF (NYHA > 1).
Results: In so far 18 recruited patients with secured MFS (age 8.1 ± 3.2 years), 66.7% exhibited cardiomyopathic features. Only 16.7% (n = 3) showed symptoms of congestive heart failure (NYHA>1). A reduced CI was found in 22.2% (n = 4), an EF<50% in 11.1%

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(n = 2), and 27.8% (N = 5) had signs of diastolic dysfunction. All tested positive for an FBN1 mutation, with 38.9% (n = 7) carrying a dominant-negative variant without cysteine involvement, 44.4% (n = 8) a dominant-negative variant involving cysteine, and 11.1% (n = 2) a haploinsufficient variant. In these three categories we observed an incidence of cardiomyopathic features of 57.1%, 62.5% and 100% respectively.
Conclusion: This first interim analysis of our prospective cohort study confirms our earlier findings of cardiomyopathic features in children and adolescents with MFS. The class of underlying FBN1 mutation might adversely contribute to higher severity of primary cardiac disease burden in the same way that genotype-phenotype correlation is described in the overall disease. Understanding these correlations better will help providers focus resources on preventing the deterioration of cardiac function in children and adolescents with MFS.

Keywords: Marfan Syndrome, Marfan Cardiomyopathy, FBN1 Mutations, Genotype-Phenotype Correlation

P-276
Pulmonary artery banding as adjunct therapy for ventricular recovery after alcapa-repair: a case report

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Background and Aim: Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital coronary abnormality commonly associated with severe but reversible left ventricular dysfunction, even after restoring coronary perfusion.
Methods: We present a case of persisting left ventricular failure after successful corrective coronary reimplantation, with inability to wean off ventilator and inotropes. Considering the preserved right ventricular function, pulmonary artery banding was forwarded as valid option to aid weaning. Almost four weeks after corrective surgery, banding of the pulmonary artery was performed to achieve mild shift of the interventricular septum without compromising circulation.
Results: Pulmonary artery banding provided successful bridging to myocardial recovery. Early mechanical improvement allowed weaning after a few days, after which gradual myocardial recovery resulted in further resolution of clinical symptoms after several months. Partial debanding was performed percutaneously around one year after primary surgery.
Conclusion: Pulmonary artery banding can be considered as a bridge-to-recovery in selected cases of severe left ventricular dysfunction after ALCAPA-repair.

Keywords: ALCAPA, left ventricular failure, bridge-to-recovery, pulmonary artery banding

P-277 / Moderated Poster
Differences between qrs-t angle in children with idiopathic dilated cardiomyopathy - a pilot study

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Background and Aim: Idiopathic dilated cardiomyopathy (IDCM) is a rare but potentially fatal disease in children, as heart failure often progresses and malignant arrhythmia appears. In adult patients with non-ischemic dilated cardiomyopathy greater QRS-T angle derived from electrocardiogram (ECG) has been associated with: increased mortality, risk of ventricular arrhythmia, sudden cardiac death (SCD) as well as rehospitalization due to heart failure. The aim of this pilot study was to assess electrocardiographic and echocardiographic factors associated with unfavorable outcomes of pediatric IDCM such as ventricular tachycardia (VT) or listing for heart transplant (HTx). The differences between the spatial QRS-T angle in children with and without the end-point of the study were assessed. To our knowledge no such study has been previously conducted in children.
Methods: Patients under the age of 18 with IDCM were included into the study. ECG analysis was performed by two independent observers; discrepancies were resolved by the third reviewer. Echocardiographic parameters (left ventricular ejection fraction (LVEF), left ventricular end-diastolic dimension (LVEDD)) and a cardiac biomarker (NT-proBNP) were also analyzed.
Results: There were 17 children with IDCM included into the study; the mean age was 8,9 years (IQR 3-14 years), 8 females. Four children with IDCM had a VT; these patients had significantly greater QRS-T angle (143° vs 44° p = 0,006), higher NT-proBNP value (2232 pg/ml vs 69 pg/ml; p-value 0,006) as well as significantly lower LVEF (23,5% vs 46%; p = 0,008) and greater LVEDD z-score (7,35 vs 2,43; p = 0,002) when compared to patients without malignant arrhythmia. Children qualified for HTx (5/17) also had a significantly greater QRS-T angle (128° vs 41°; p <0,001), as well as higher NT-proBNP (2448pg/ml vs 63,5pg/ml; p <0,001) and lower LVEF (24% vs 46,5%; p = 0,002) and higher LVEDD z-score (7,21 vs 2,38; p = 0,001). There was a strong correlation between QRS-T angle and LVEDD (rs 0,76), a strong inverse correlation between QRS-T angle and LVEF (rs -0,66) and a moderate correlation between QRS-T and NT-proBNP (rs 0,53).
Conclusion: QRS-T angle appears to be a new promising parameter associated with both ventricular arrhythmia and end-stage heart failure in pediatric IDCM.

Keywords: idiopathic dilated cardiomyopathy, pediatric cardiomyopathy, ventricular arrhythmia, QRS-T angle

P-278
Defined structural features potentially favor infective endocarditis in bovine jugular vein valved conduits

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Background and Aim: Late onset endocarditis (IE) is enhanced in pulmonary valved conduits made from bovine jugular veins (BJV), as compared to valved conduits from other sources. The mechanism of IE onset remains unclear and less is known about in vivo tissue integration of implanted conduits. The aim of this study was to compare cellular infiltration and extracellular matrix remodeling within cryopreserved homografts (CH) and BJV valves in a sheep model.

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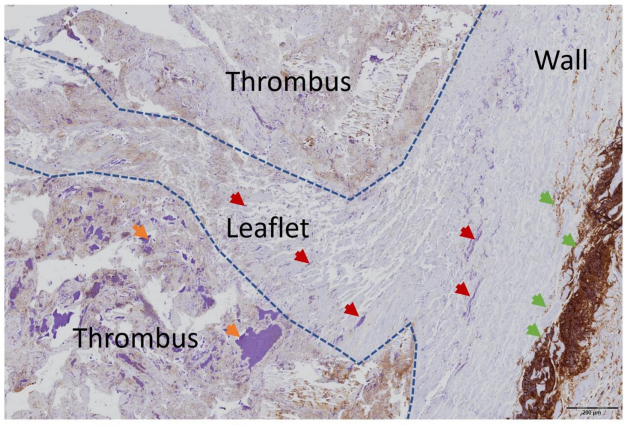
Methods: Four ovine CHs and 11 BJVs were implanted in pulmonary position in sheep. Hemodynamic function was confirmed by MRI. After 4 month, valves were explanted, fixed in formalin and prepared for movat verhoeff staining to differentiate extracellular components and for immunohistology to detect lymphocytes, macrophages, granulocytes, contracting cells and fibroblasts.
Results: In homografts, as compared to native pulmonary valves, a general reduction of donor cells was observed. With relatively low cellular infiltrations, the graft-wall-diameter was lowered by 20%, while the diameter of the CH-leaflets was raised by 60%, due to enhanced infiltration of immune cells. In BJVs, a high number of macrophages, that partially presented as foreign body giant cells, was detected between displaced and fragmented graft-collagen fibers, leading to a thickening of the wall-diameter by 57% as compared to an unimplanted controls. In contrast, no infiltrating cells were detected within BJV-leaflets. Bacterial infection occurred as unexpected complication in 1 CH and 3 BJVs. This enabled the detection of the locations of bacterial proliferation and cellular response. In the homograft, bacteria were seen within a thrombus-like formation in close proximity to neutrophils. However, the tightly packed collagen fibers of BJV leaflets rarely showed any immune cell infiltrations, while bacteria were regularly seen here.
Conclusion: In homograft tissue, donor cells are depleted rapidly, while recipient cells can infiltrate the entire graft to properly react to bacterial infection. In contrast, the tightly packed collagen fibers of BJV leaflets seem to prevent the infiltration of immue cells, while bacteria can enter the tissue and proliferate, not being offended by the cellular immune system.

Keywords: Homograft, BJV, heart valve, Infective Endocarditis

Figure Abstract II



Tissue section of infected BJV, explanted from sheep after 4 month of implatation. Staining: CD45 (immune cells appear in brown, green arrows). Red arrows point to bacterial colonies within graft tissue. Organe arrows: bacteria in thrombus. Dashed line borders BJV explant from surrounding thrombus.

P-279

Acute heart failure in children: it is always myocarditis?

Laura Castro González[1], Cristina Navarro Sánchez[1], Pedro Agudo Montore[2], María Rollán Martínez Herrera[1], María Carmen Gallego Muñoz[1], María Ángeles Murillo Pozo[1]
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Background and Aim: Heart failure (HF) is the inability of the ventricle(s) to fill with and/or eject blood. HF is estimated to affect 12,000 to 35,000 children under 19 years in the US annually. It results from structural or functional cardiac disorders. The presentation of HF in paediatrics is diverse because of the numerous underlying cardiac aetiologies and varying clinical settings.
Methods: One clinical case description and literature review.
Results: A 2-year-old patient is brought to the Paediatric Intensive Care Unit from the Emergency Room with haemodynamic instability and obnubilation. She had previously 24 hours of fever, no other symptoms. On X-Ray Chest a "bat wings" pattern and an enlarged heart silhouette were seen; the echocardiogram showed a decreased heart function: left ventricle ejection fraction (LVEF) of 23%. BNP and troponins were also elevated on the laboratory tests. Acute HF due to infectious myocarditis was suspected and treated: inotropic and vasopressor agents, diuretics, invasive mechanical ventilation, intravenous (iv) immunoglobulins, iv steroids, and iv antibiotics. During the next hours, the instability and fever persisted, and body temperature control device was started, permitting a decrease in the inotropic support. The following 48h, the patient maintained low LVEF, and levosimendan (calcium sensitiser) was initiated, without improvement. The patient had been studied in the Rheumatic Unit for recurrent fever without a diagnosis, adding then Anakinra (antileukine-1 inhibitor), suspecting underlying autoimmunity. Complementary tests showed no acute infection. Autoantibodies (anti-Ro and ANAs) were positive. Cardiac MRI showed LVEF of 33%, oedema (inflammation), and fibrosis (old ischemic lesions) of the LV. Cardiac catheterization showed small aneurysms of the right and circumflex coronary artery (beaded aspect), finding compatible with autoimmune coronaropathy. The patient is now awaiting a heart transplant.
Conclusion: HF diagnosis is based on clinical features, cardiac imaging, and laboratory findings. It is mandatory to have a correct approach to the underlying cause. An intensive approach of the cardiogenic shock and an incessant search of the aetiology, even performing early invasive techniques like cardiac catheterization, is essential to establish the correct treatment and improve the prognosis of these patients. Reports of individual cases may help with those patients management.

Keywords: Paediatric Cardiac Pathology, Heart Failure, Rheumatic Disease.

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Acute Heart Failure in Children Image

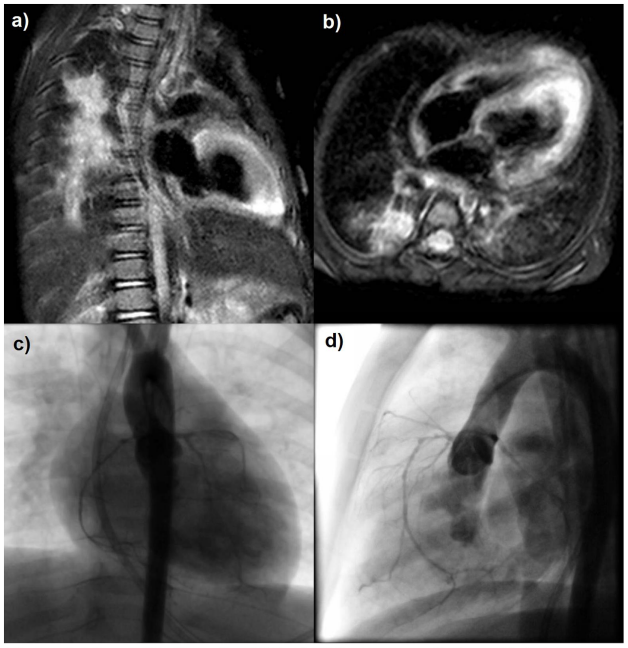

Figure 1. T2-weighted cardiac MRI: a) Oblique coronal plane; b) Axial plane. Coronarography: c) Posteroanterior projection; d) Lateral projection

Cardiac MRI showing decreased LVEF, oedema (inflammation), and fibrosis (old ischemic lesions) of the LV. Cardiac catheterization showing small aneurysms of the right and circumflex coronary artery (beaded aspect).

P-281
Vaccination against pneumococcal infections in children with congenital heart defects

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Background and Aim: Study the vaccination status of CHD children, the tolerance of pneumococcal vaccination and its effectiveness
Methods: 50 parents of children with CHD were surveyed. The post-vaccination period was followed up with 30 unvaccinated or partially vaccinated children with a 13-valent pneumococcal conjugate vaccine. In a catamnesis a year later the incidence of respiratory infections in these children was assessed.
Results: According to the results of the survey, 11 (22 per cent) children with UPU were vaccinated against pneumococcus. The remaining 39 children (78 per cent) did not receive vaccinations for the following reasons: lack of information - 44.5 per cent, medical excuse - 42.8 per cent - from the attending physician: Refusal by parents - 12.7 per cent. In the Vaccination Unit, 30 children received one or two doses of conjugate pneumococcal vaccine. All patients were vaccinated remotely after the surgery. Only 2 (6.6 per cent) patients received the first dose of vaccine before the age of 1. Most of the patients - 17 children (56.6%) were vaccinated in the second year of life, 11 children (36.8%) of the patients were vaccinated after 2 years of age. There were no post-vaccination complications. Twelve (40 per cent) of the children showed a rise in body temperature to sub-febril numbers lasting up to two days, and 18 (60 per cent) had a slight increase in hyperemia and compaction at the injection site. Before vaccination, all children were in the high-prevalence group of children, 14 children (46.6 per cent) were suffering from acute pneumonia, 2 children were suffering from acute otitis and 1 child was suffering from meningitis. In the first year after vaccination, the incidence of acute respiratory infections declined more than twofold, with two children suffering from pulmonary hypertension suffering from acute pneumonia. No child has been diagnosed with acute middle otitis or meningitis, and no aggravation or aggravation of the main disease has been detected.
Conclusion: Pneumococcal vaccination is safe and effective in children with congenital heart disease. Consideration of recommendations for vaccination of this category of patients at an earlier date, including before prompt treatment, remains relevant.

Keywords: Vaccination against pneumococcal infection, congenital heart defects

P-282
Predictive wall stress as a prognostic marker for left ventricular function 1–2 years after surgery in congenital mitral regurgitation

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Background and Aim: Predictive wall stress (pWS), calculated from preoperative diastolic dimensions, diastolic wall thickness, and aortic diastolic blood pressure, helps predict hemodynamics just after surgery for mitral regurgitation (MR). As pWS can predict postoperative left ventricular (LV) afterload and afterload is major determinant of postoperative LV pump function, it is useful for determining the timing of the operation in children and adolescents with congenital MR (Murakami T et al. Pediatr Cardiol 1999). Because persistent increases in wall stress after surgery can cause left ventricular injury, we investigated the correlation between pWS and postoperative LV function, including 1–2 years after surgery.
Methods: This study included 11 pediatric and adolescent patients who underwent mitral valve repair at Hokkaido University Hospital in the past 20 years. The patients' echocardiographic recordings were retrospectively analyzed, and the relationship between pWS and postoperative LV function was investigated.
Results: The mean age of the patients at surgery was 10.2 ± 7.0 years, the preoperative fractional shortening (FS) was 0.36 ± 0.04, and the pWS was 167 ± 24.1 kdyn/cm^2. Although the FS before surgery did not correlate with the FS immediately after surgery (0.31 ± 0.06) ($rs = 0.511$, $p = 0.108$), the pWS before surgery was significantly correlated with the FS immediately after surgery ($rs = -0.62$, $p = 0.04$). Additionally, we investigated the correlation between pWS and LV function in 1–2 years after surgery. In seven cases, a significant correlation was found between preoperative pWS (165 ± 25.4 kdyn/cm^2) and FS 1–2 years after surgery (0.38 ± 0.05) ($rs = -0.83$, $p = 0.02$). Moreover, preoperative pWS correlated strongly with postoperative FS recovery (i.e., postoperative FS at 1–2 years after operation minus postoperative FS immediately after surgery) (0.06 ± 0. 03) ($rs = -0.95$, $p < 0.01$; Figure).
Conclusion: We suggest that the pWS of congenital MR not only helps predict hemodynamics immediately after surgery, but may also predict the prognosis of LV function, including 1–2 years after surgery. Our findings could be applied to preoperative and postoperative management of congenital MR patients.

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Keywords: Mitral regurgitation, predictive wall stress, left ventricular wall stress, left ventricular afterload, mitral valve repair

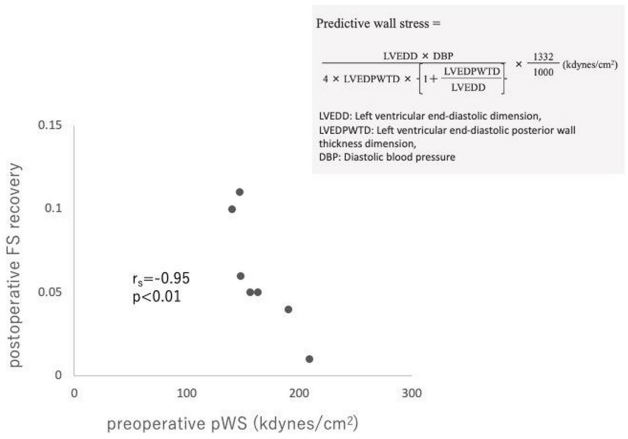

Figure 1 Relationship between preoperative pWS and postoperative left ventricular FS recovery. Correlation was analyzed using Spearman's test (n = 7). FS: Fractional shortening, pWS: Predictive wall stress, postoperative FS recovery: Postoperative FS at 1–2 years after operation minus postoperative FS immediately after surgery

P-283

Premature closure of the ductus arteriosus – prenatal right heart failure and postnatal ductal aneurysm requiring surgery

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Background and Aim: In utero constriction of the arterial duct is a rare condition and mostly associated with prostaglandin-inhibiting drugs such as Indomethacin. However, spontaneous closure has been reported to occur as well, although the incidence remains unknown. Ductal aneurysms with thrombus formations on the other hand are significantly more common and are even debated as a physiological form of postnatal closure. Both conditions are potentially life-threatening, especially in case of premature closure. We report a case of a term infant after routine fetal echocardiography revealed right heart failure and pseudoatresia of the pulmonary valve.
Methods: After birth the infant showed severe respiratory distress, respiratory acidosis, massive hypertrophy and dysfunction of the right ventricle as well as elevated pressure in the pulmonary circuit. Echocardiography failed to show blood flow through the ductus arteriosus, so premature ductus constriction was suspected. Despite opening of the pulmonary valve could be demonstrated postnatally, inotropes as well as endotracheal intubation were deemed necessary. The following echo controls showed significant improvement of heart function which first led to an extubation and cessation of medical therapy. However, reintubation was required on the same day due to the presence of marked inspiratory stridor. Subsequent diagnostics revealed paresis of left laryngeal nerve secondary to ductal aneurysm (DA) compression and additionally obstruction of the left pulmonary artery (LPA). Consequently, it was considered necessary to excise the thrombus within the arterial duct and enlarge the LPA.
Results: Excision of the ductus revealed a 5x6mm thrombus formation within the DA. The LPA insertion site of the arterial duct was augmented with autologous pericardium. Postoperatively, echo controls showed an unobstructed laminar blood flow in both main branches of the pulmonary artery. However, inspiratory stridor was ameliorated but still present. It must be assumed that the DA was causative for the paresis, due to the proximity of the laryngeal nerve to the arterial duct. Right ventricular function resolved slowly. A complete restoration of cardiac function can be expected.
Conclusion: Premature closure of the ductus arteriosus can lead to impaired RV-function before birth, and postnatal sequelae might need surgical intervention.

Keywords: Premature Ductus closure, Right heart failure

P-284

Increased aortic stiffness in fontan patients: impact on hemodynamics and clinical course

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Background and Aim: Arterial pulse wave velocity (PWV) is a surrogate marker for elasticity and rigidity of the arterial vessels and increases with arterial stiffness, which in turn affects the cardiac function. It is not known whether aortic stiffness is altered in univentricular Fontan circulation. We therefore investigated invasively measured PWV in "Fontan-patients" in comparison to patients with "normal heart" physiology. Furthermore, the impact of PWV on other Fontan hemodynamic parameters with possible clinical significance was studied.
Methods: We investigated 19 Fontan patients (mean age 18,5 ± 9,3 years; 9 men) with an underlying condition of: hypoplastic left heart syndrome (n = 12), tricuspid atresia (n = 3), double inlet left ventricle (n = 2), unbalanced AVSD (n = 1) and double outlet left ventricle with uncommitted VSD (n = 1). The control group of 28 individuals was age-, sex- and BMI-matched with a normal heart physiology (heart-transplant patients, 17,5 ± 5,2 years, 14 men). The PWV was assessed in the ascending aorta. Pressure pulse waves were simultaneously documented from the catheter in the aortic position and femoral artery sheath. Travelled distance and time delay between both pulse waves was measured. Subsequently PWV was calculated Furthermore central arterial blood pressure (cBP), central venous pressure (CVP) or mean pulmonary artery pressure (mPAP) in Fontan patients, systemic enddiastolic pressure (EDP), and Cardiac index (CI) were assessed.
Results: Fontan patients showed a significantly higher PWV in comparison to the control group (5,6 ± 1,3 vs. 4,9 ± 0,6 m/s; $p<0,05$; see Figure 1). The other arterial and left ventricular hemodynamic parameters were not significantly different between both groups. PWV correlated significantly to cBP (Pearson coefficient ≥ 0,4; $p<0,05$) in the controls but not in the Fontan-patients. However, we identified a moderate but not significant positive correlation of PWV with mPAP within the Fontan-cohort (r = 0,3).

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Conclusion: PWV is a surrogate parameter for arterial stiffness and is increased in Fontan patients. The reason for this is unclear, however surgical arch reconstruction, the use of patch material and subsequent scarring may be causative. As there was a moderate correlation with mPAP monitoring of the arterial stiffness using PWV measurement could be an additional tool to early predict failing Fontan physiology. Further research is however needed to verify our results.

Keywords: PWV, Pulse-wave-velocity, central blood pressure, pediatrics, Fontan, univentricular

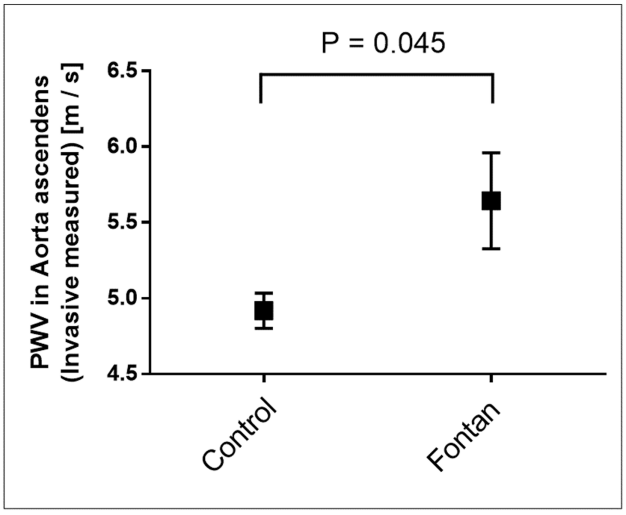

Figure 1 Box plot to illustrate the difference between invasively-measured PWV [m/s] in Fontan and Control-group

P-285

Role of circulating cardiac autoantibodies in pediatric non-compaction cardiomyopathy

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Background and Aim: Circulating cardiac autoantibodies was previously described in adults with myocarditis and dilated cardiomyopathy and it correlates to a lower rate of cardiac recovery. Anti–muscarinic receptor antibodies have also been detected in adult patients with arrhythmias in other autoimmune diseases. Autoimmune activation was also described in children with myocarditis and it correlates with non-recovery.
However, the presence of cardiac autoantibodies in dilated cardiomyopathy and their relationship to clinical outcomes have not been evaluated in children. We aim to describe the presence of cardiac autoantibodies and their correlation with outcome in a subset of pediatric patients with non-compaction dilated cardiomyopathy.
Methods: From January to December 2019, all consecutive patients with non-compaction cardiomyopathy were enrolled. Blood samples were collected during clinical evaluation and specific anti-cardiac autoantibodies were evaluated: Anti-Heart Autoantibodies (AHA), Anti-Intercalated Disk Autoantibodies (AIDA) and Anti Endothelial Cell Antibodies (AECA). Data analysed were: family history of cardiomyopathy or autoimmune disease, admission for heart failure, arrhythmias, fibrosis evaluated at cardiac magnetic resonance (CMR) and outcomes (VAD, heart transplant or death). Patients with negative autoantibodies were considered as a control group.
Results: 11 consecutive children were enrolled (mean age 11 ± 6; male 63%). Three of these patients showed elevated level of cardiac antibodies: patient 1 with AHA; patient 2 with AIDA; patient 3 with both AHA e AIDA; no patients showed elevated levels of AECA. Comparing to control group admission for heart failure, adverse outcome and presence of fibrosis were present in patients with elevated circulating antibodies. No differences were notice in arrhythmias or presence of family history of autoimmunity or non-compaction disease.
Conclusion: Although the study was limited by small sample size, we can speculate that the presence of circulating cardiac autoantibodies in children with non-compaction cardiomyopathy may help to identify patients at risk of adverse outcomes.

Keywords: cardiac autoantibodies, non-compaction cardiomyopathy, pediatric

P-287

The impact of a novel mobile driving unit on quality of life in children treated with a paracorporeal vad: interim results from the e-motion study

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Background and Aim: The use of Ventricular Assist Devices (VAD) in pediatric patients has significantly grown over the last years. Although VAD therapy shows good outcomes, lack of mobility remains a major concern in this young cohort. To improve mobility, a mobile driving unit (MDU) was recently developed for the only approved paracorporeal VAD-system for children of all sizes and ages.
This observational clinical study evaluates the safety and performance of MDU as well as its impact on patient mobility and Quality-of-Life (QoL).
Methods: Twelve patients (5 males, 7 females) from 5 international sites were included in this interim analysis. Patients were eligible for study participation on paracorporeal VAD support connected to a stationary driving unit (SDU). Enrolled patients remained on the SDU for 7 ±5 days and were then switched to MDU with a follow-up of further 45 ±7 days. To evaluate QoL and psychological condition, patients or their caregivers completed the Pediatric-Quality-of-Life-Inventory (PedsQL), Depression-Anxiety-Stress-Scale (DASS-21) and a customized questionnaire to assess family satisfaction. In addition, the patients/caregivers kept a diary to record daily activities.
Results: The median age was 3.4 years (range 0.1–17.4 years), median Body-Surface-Area was 0.55 m2. The diagnoses were cardiomyopathy (9 patients, 75.0%), myocarditis (2 patients,

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16.7%) and one patient had a congenital heart defect (8.3%). Eight patients (66.7%) received a leftventricular VAD the others were treated with biventricular VAD. At the end of the observation period, 10 patients (83.3%) were still on-system and 2 (16.7%) were transplanted.
The study found significantly increased patient activity ($p = 0.020$), mobility ($p = 0.008$), and a significant higher independency from the healthcare-professional ($p = 0.027$) for MDU in comparison to SDU. In addition, an increase in the QoL was observed by all questionnaires while on MDU support.
The safety profile showed a comparable adverse event rate between both units (per 10 patient days, SDU: 0.294 vs MDU: 0.244). None of the adverse events was classified as serious.
Conclusion: The preliminary results suggest that the novel mobile driving unit is safe and effective for paracorporeal VAD support in pediatric patients. The increase in patient mobility indicates an elevated QoL in this cohort.

Keywords: ventricular assist device; driving unit; qualitiy of life

P-288
Acute cardiac compromission in patients with MIS-C

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Background and Aim: Multisystem inflammatory syndrome in children (MIS-C) is a late manifestation of SARS-CoV-2 infection. Cardiac involvement is common and presents as ventricular dysfunction, shock, and coronary anomalies. The aim of the study is evaluate the influence of cardiac disfunction on clinical presentations and outcomes in a single center.
Methods: A retrospective study on patients diagnosed with MIS-C and referred to Buzzi Children's Hospital in Milan from November 2020 to February 2021. Patients were treated with intravenous immunoglobulins, corticosteroids and anti-thrombotic prophylaxis, in respect to our approved multidisciplinary protocol. According to the admission cardiac left ventricular ejection fraction (LVEF), the patients were divided into group A (LVEF <45%) and group B (LVEF ≥45%).
Results: We collected 32 consecutive patients. Group A included 10 patients (9M/1F, aged 13 years [IQR 5–15]), and group B included 22 patients (15M/7M, aged 9 years [IQR 7–13]). At the presentation, significant differences were observed among shock (group A 6/10 vs group B 2/22, $p<0.01$), gastrointestinal involvement (9/10 vs 11/22, $p = 0.04$) and duration of fever (5.3 vs 6.9 days, $p = 0.02$). All patients in group A required intensive care hospitalization (10/10 vs 12/22, $p = 0.01$). Interestingly, despite good cardiac function, two patients in group B presented with shock, probably due to vasoplegic/distributive cardiocirculatory impairment secondary to the inflammatory state.
Among biochemistry parameters, leukocytes, neutrophils, and CRP were significantly worse in group A ($p = 0.001$, $p = 0.001$ and $p = 0.008$, respectively). Pathological level of troponin T and NTproBNP were detected in all patients in group A and also in 33% and 77% of group B; with statistically significant higher median values in group A (Troponin T 72 [40-243] ng/L vs 22 [8-49] ng/L, $p = 0.01$; NTproBNP 14825 [11340-17810] ng/L vs 5921 [1114-11243] ng/L, $p = 0.01$).
In group A, mitral regurgitation was more frequent ($p<0.01$) and one patient had transient left main coronary dilation (Boston z-score +2.39). At the discharge, cardiac function normalized in all patients. Total length of hospital stay and cardiac recovery time were not statistically different between groups.
Conclusion: If correctly diagnosed and early treated, all the MIS-C patients completely recovered, regardless of the initial cardiac involvement.

Keywords: Multisystem inflammatory syndrome, children, heart, cardiac, COVID-19

P-289
Experience with safety and efficacy of levosimendan in pediatric patients

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Background and Aim: The objective of this study was to describe the tolerability and results of treatment of critically ill children with levosimendan infusion (LEVO).
Methods: The study is a retrospective single-center analysis of all patients aged 0 to 18 years, who received one or more levosimendan infusions, for failure to wen off inotrope and/or ECMO support. Demographic data and clinical, biological, and echocardiographic parameters (LVSF, subaortic VTI and TAPSE) were serially collected at onset of LEVO (TO) and at 2 days (T2), 10 days (T10), 30 days (30) and
Results: 71 patients (42 males, mean age 3.2y) received 112 levosimendan infusions (LEVO): 34 (48%) had myocardial dysfunction including 5 acute myocarditis (CMP group), and 37 (52%) in the early postoperative course after surgery for congenital heart disease (CHD). Mean time from surgery to LEVO was 8.3days (median 3), from hospitalisation to LEVO was 30.5days (median 15); 90% of cases were dependent of at least one inotrope support (77% milrinone). Mean dose of LEVO was 0.2microgrammes per kg per mn. Inotrope support was successfully discontinued in 34% of 32 LEVO (60% maintained off support) and 65.6% of 32 LEVO on ECMO support were weaned off assistance. Mean heart rate decreased from 137 to 131 and 116 bpm at T0, T2 a,d T30), mean blood pressure increased (64 to 67 and 79mmHg at T0, T2 and T30), and mean lactates decreased from 2.17 to 1.9 to 1.5.
Mean LVSF increased from 18.3% to 21.2%, 22.9%, 25% and 31.9%, at respectively TO, T2, T10, T30 and M6. VTI increase was +1.7cm, +3.1cm and +6cm, at T2, T10, T30 and TAPSE increase was +2.2mm and +2.8mm at T10 ans T30. LEVO was uncomplicated in 109 among 112 infusions (97.3%), 3 experienced adverse events (tachycardia and hypotension). Mean CICU time was 66 days (median 46) and hospitalisation time was 76days (median 65). Fourteen patients underwent heart transplantation and 30 died, i.e. failure occurred in 42 cases (59.2%). Survival rates was 50% at 6th month after termination of LEVO.
Conclusion: Levosimendan is safe and can allow weaning from inotrope and/or ECMO support in hemodynamically compromised pediatric patients with CMP and/or CHD.

Keywords: levosimendan, cardiomyopathy, congenital heart disease, efficacy, tolerability

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P-290

Acute myocarditis in children: features and outcomes of COVID-19 and NON-COVID patients

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Background and Aim: The objective of the study was to assess the features of acute myocarditis and compare Covid19 and non-Covid19 cases.
Methods: Patients <18y with acute myocarditis (proved by virology and/or MRI and/ or complete recovery of myocardial function) were included. Clinical data, echocardiographic parameters and outcomes were collected. Cases were divided in groups I (non-Covid), II (Covid).
Results: From 1983 to 2021, 139 patients were included: 76 patients in group I and 63 in group II, 67males (31 in group I = 40% vs 36 in II = 57%). Mean age at diagnosis was 6.8 years: 4.2 years in group I vs 9.9 years in II. Heart failure (HF) was present at onset in 78% of cases in group I and 50% in group II: severe HF was more frequent in group I, chest pain was more frequent in II. Mean left ventricular shortening fraction (LVSF) at diagnosis was 23.8%: 18.4% in groups I vs 31.6% in II ($p<0.05$). Mitral regurgitation was present in 63.8% of cases = 76.5% vs 43.8% respectively in groups I and II, pericarditis in 16.4% (no difference between groups), thromboembolic events occurred in 7% and arrhythmias in 10% ((all in group I). Virus was positive in 37.5% in group I and SARS-Cov2 positive in all of group II. Inotrope support was needed in 47%, mechanical circulatory support in 8% in group I only. Eleven patients died in group I, no death occurred in group II. One was transplanted(3rdmonth) and 19 have sequellae in group I. Complete recovery occurred in 74% of all cases: 40 of group I (58%) and all of group II (100%): time to recovery was longer in group I (2 years) than in group I (2 weeks). Mean LVSF improved from 18.4% at onset, to 24.6% at 1st month, 26.5% at 3rd month, 30.7% at 6th month and 38% at last FU in group I, while mean LVSF normalized within 2 weeks after onset in group II.
Conclusion: Myocardial dysfunction and heart failure were less frequent, and complete recovery occurred promptly in COVID cases, while myocardial improvement progressed slowly within first 6months and beyond in half of non-COVID cases.

Keywords: acute myocarditis, children, SARS-Cov2, outcomes

P-291

Hidden in plain sight: a case of arrhythmia and sudden cardiac failure masked by COVID-19 positivity

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Background and Aim: Whilst most commonly recognised as a respiratory pathogen, COVID-19 can also result in a variety of extrapulmonary manifestations including myocardial dysfunction and arrhythmia. We report a case of a 15 year old girl with repaired atrioventricular septal defect, presenting with arrhythmia and sudden severe cardiac failure masked by COVID-19 positivity.
Methods: A 15 year old girl with repaired atrioventricular septal defect and tetralogy of Fallot, under regular follow up with asymptomatic moderate to severe atrioventricular valve dysfunction, presented with one month's history of progressive breathlessness to her local hospital. Onset of illness coincided with typical COVID-19 symptoms; her family, attributing her deterioration to this, delayed seeking medical help. She was rapidly referred to our unit.
The heart failure severity, which included diffuse dependent oedema, large pleural effusion and severe biventricular dysfunction, could not be attributed to major valve function change. Moreover, deterioration had occurred rapidly without apparent ongoing cause, after extensive diagnostics. Tachyarrhythmia was suspected; initial adenosine challenge via peripheral vein was inconclusive. Diuretics, inotrope and empirical administration of amiodarone provided limited response; the patient was therefore ventilated, enabling further adenosine challenge via central line, which revealed atrial flutter. Shortly after conversion to sinus rhythm and pleural drainage, her biventricular function improved to near normal.
Results: Redo surgery was undertaken. Firstly, this included mechanical valve replacement of left and right atrioventricular valves and ablation of the isthmus. Secondly, a permanent pacemaker was inserted a few days later. The patient made a quick and uneventful recovery and was discharged on day 10 with good biventricular function on minimal medical therapy and no symptoms.
Conclusion: Our case highlights delayed presentation as a hidden effect of the COVID-19 pandemic, and that sudden deterioration in stable children with repaired congenital heart disease should prompt the clinician to exclude all reversible causes of de-stabilisation, and in particular to maintain high suspicion of arrhythmia.

Keywords: COVID-19; atrial flutter; atrioventricular septal defect; delayed presentation

Interventional Cardiology

P-292 / Moderated Poster

Institutional experience with the occlutech© atrial flow regulator (AFR) in children with congenital heart disease

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Background and Aim: Children with congenital heart disease (CHD) and heart failure (HF) with reduced (HFrEF) and with preserved ejection fraction (HFpEF) have limited treatment options to improve outcomes/quality of life. Long-term interatrial communication can be therapeutic option for children as it is for adults.
Methods: Atrial Flow Regulator device (AFR, Occlutech) was implanted in 9 patients aged 21-85 months (median 30 months) of body weight 9-20 kg (median 10 kg). Left ventricular diastolic disfunction related to left heart disease was the indication in 8 patients (aortic stenosis - 5, aortic stenosis/mitral valve disease - 2, hypoplastic left heart complex - 1) and dilated cardiomyopathy with severe left ventricular disfunction in 1 case. Prior to implantation of the AFR the patients underwent fetal aortic valvuloplasty (6), Ross-Konno procedure (2), (bilateral pulmonary artery banding + ductal stenting (2). All of the left heart disease patients had repeated balloon atrial septostomies. In the dilated cardiomyopathy patient implantation of the AFR was followed by pulmonary artery banding. The AFRs were implanted under general anaesthesia from right femoral vein access (7 patients) or transhepatic

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access (2 patients). Transseptal puncture with Brockenbrough needle was performed in 6 patients, existing interatrial communication was used in 2 patients. Landing zone was prepared using 12 mm balloon. The AFR fenestration diameter was 4 mm in 6 patients, 6 mm in 3 patients. Implantation of AFR was approved as compassionate use by the local Ethical Committee and Occlutech.
Results: Immediate results after AFR: mean gradient reduction from 10-25, med.13mmHg to 8-16, med.10mmHg. All patients were on dual antiplatelet therapy.
During follow-up 7-18, med.11mths in 8pts AFR flow was preserved with mean gradient 5-14, med.7,5mmHg. Serious restriction of flow (neointimal hyperplasia?) through the AFR occured in one pt 14mths after implantation, treated successfully with balloon dilation (gradient reduction from 16 to 8mmHg. NYHA class improved in 8pts, was unchanged in 1pt.
Conclusion: Compassionate use of the AFR device in children with HFpEF and HFrEF is technically feasible and produces beneficial short/midterm hemodynamic and clinical improvement. Late follow-up and further work is needed to evaluate its role for this diverse group.

Keywords: atrial flow regulator, AFR device, HFpEF treatment

P-293
Special vascular intervention in newborn with scimitar syndrome

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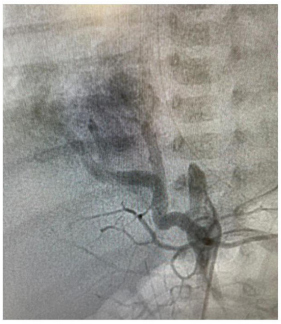
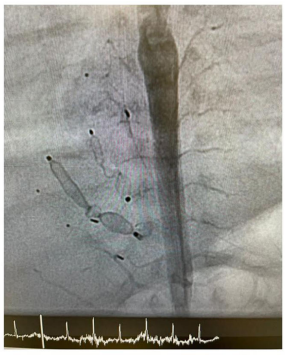
Background and Aim: Scimitar syndrome is a rare and complex association of congenital cardiopulmonary abnormalities with an estimated prevalence of 1 to 3 per 100,000 births. Literature demonstrated that patients presenting in infancy are more likely to have severe heart failure, Aorto-pulmonary Collaterals, and a poorer prognosis than patients presenting at a later age
Methods: Case History:
We are presenting a 4 weeks old neonate with weight of 2.7 kg, diagnosed as a non-obstructive scimitar syndrome (right lower pulmonary vein draining directly to IVC) and multiple massive aorto-pulmonary collaterals to the right lung and non-restrictive ASD secundum, hypoplasia of RPA (right pulmonary artery) and right lung. Diagnosis was confirmed by transthoracic Echocardiography, CT scan of abdomen and thorax with contrast and cardiac MRI. Patient was not diagnosed antenatally. Presented after birth with severe pulmonary hypertension requiring high setting of ventilation and high serum lactate level. Patient developed systemic circulation steel with bowel ischemia and infection (necrotizing enterocolitis). After failure of aggressive supportive medical treatment, patient was taken to the catheterization lab for diagnostic and interventional catheter.
Results: Therapeutic Interventions:
Angiographies showed multiple massive collaterals from abdominal aorta to the right lung with pulmonary venous return of the lower right lobe of the lung to the IVC and significant cardiomegaly.
We used multiple microvascular plugs, PDA occlusion devices, Piccolo device through 3 Fr then 4 Fr sheaths and catheter through right femoral artery to occlude the massive collaterals. Post- procedure patient was extubated in the NICU and heart failure symptoms disappeared.
Long term follow up showed grumbling lactate despite all screening tests (metabolic, genetic and gastrointestinal) were normal reflecting chronic ischemic changes of embolized lung tissue. lactate normalized after 6 months.
Follow up cardiac catheterization showed growing RPA.
Conclusion: Massive aortopulmonary collaterals transcatheter arterial embolization can be done safely in the catheterization lab in neonates with small body weight and represent a minimal invasive option compared to surgical option. Embolization of anomalous systemic arteries can dramatically improve symptoms of heart failure and pulmonary hypertension in neonates, and potentially allow growth of RPA. Lactate can remain high for long time following aortopulmonary embolization.

Keywords: Scimitar syndrome, Aortopulmonary collaterals, high lactate, Neonatal cardiac interventions

Aortopulmonary collaterals pre and post occlusion



Descending Aorta Angiography demonstrating multiple Major Aorto-Pulmonary Collaterals arising from Abdominal Aorta to the right lung.



An angiograph showing multiple occluders deployed in the AP collaterals

Imaging / Functional assessment

P-294 / Moderated Poster
Does pulmonary artery stenting in single ventricle patients affect lung growth and function?

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Background and Aim: Stenting of the left pulmonary artery (LPA) is often required during staged Fontan palliation in single ventricle (SV) patients, due to LPA compression from the dilated neo-aortic root. LPA stent could potentially compress the left main bronchus (LMB). We assessed the impact of LPA stenting on bronchial size, pulmonary function, and pulmonary volumes.
Methods: Forty-nine SV patients (61% males) underwent prospectively cardiovascular magnetic resonance (CMR) 11 (8-15) years after Fontan. Area and shape of the main bronchus, lung volume and surface were calculated on CMR images for each lung. These measures were transformed and expressed in right/left ratio to overcome the influence of age and body size. Body plethysmography and spirometry were performed concomitantly in 36 patients. LPA stents were present in 17 (35%) patients. CMR

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measures and lung function parameters were compared between patients with and without LPA stent. The effect of time of stent insertion, being early at stage II of palliation, was analyzed.
Results: Patients with LPA stent had a larger right/left ratio for main bronchial area (4.35 (2.82-6.56) vs. 2.02 (1.45-2.91); p<0.001) and a larger right/left ratio for lung volumes. However, the difference in lung volumes was only significant in patients with early LPA stenting (1.35 (1.32-1.44) vs. 1.26 (1.10-1.42); p = 0.04). No difference was observed for pulmonary surface and LMB compression. TLC and VC, obtained by body-plethysmography, were reduced in 44% and 50% of all patients and FEV1 and FVC, obtained by spirometry, were reduced in 33% and 36%. There were no differences in lung function between patients with and without LPA stent.
Conclusion: Early insertion of a LPA stent seems to have a negative effect on the left-sided bronchial and pulmonary growth in SV patients. Up to 50% of SV patients show an abnormal lung function; presence of a LPA stent did not have a negative influence on lung function, so that other causes need to be investigated. In LPA stented patients, the right lung may be able to compensate for the small left lung volume.

Keywords: Single ventricle, Fontan, lung function, lung growth, pulmonary artery stenting

Interventional Cardiology

P-296
Kill two birds with one stone; aorticoventricular fistula closure during percutaneous pulmonary valve implantation

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Background and Aim: Iatrogenic aorta to right ventricular fistula is an extremely rare complication following total correction of tetralogy of Fallot (TOF). The management of hemodynamically insignificant aorta-right ventricle fistula in an asymptomatic individual is controversial. Treatment options range from medical observation to transcatheter embolization or surgical ligation to complex repairs.
Methods: The patient diagnosed with TOF underwent surgical repair with a transannular patch at 1.5 years old. A progressively increasing pulmonary regurgitation and stenosis were observed on regular echocardiographic examinations. At 14 years old, aorta to right ventricle tunnel was detected on echocardiography before the percutaneous pulmonary valve implantation (PPVI). We confirmed the diagnosis by computerized tomography. Based on the past medical history and previous imaging, we assumed that the fistula is an iatrogenic one. It was not detected until the PPVI procedure as he already had right ventricular dilatation secondary to the free pulmonary regurgitation. We performed simultaneous prestenting and valve implantation with Meril's Myval 23 mm valve. Aortic root angiography before the PPVI demonstrated a fistula between the right coronary cusp to the right ventricle infundibulum. Amplatzer Vascular Plug II 6mm device was implanted by retrograde approach without forming a loop. Control angiogram after the device deployment revealed no residual leak and aortic regurgitation
Results: Although surgery is the treatment of choice, transcatheter device embolization might be an emerging alternative for aorticoventricular fistula occlusion.
Conclusion: To the best of our knowledge, this is the first successful transcatheter closure of an aorticoventricular fistula at the same session with the PPVI.

Keywords: percutaneous pulmonary valve implantation, aorticoventricular fistula, operated Fallot tetralogy, device embolization, Amplatzer vascular plug II

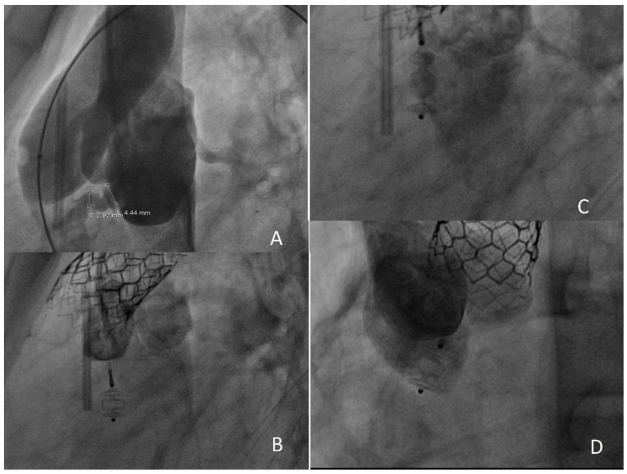

Figure (A) Aortic root angiogram shows aorticoventricular fistula (B) Amplatzer vascular plug II passing through aorticoventricular fistula (C) Amplatzer vascular plug II occluding aorticoventricular fistula (D) Angiogram after device release.

P-297 / Moderated Poster
Implantation of a new transcatheter heart valve for pulmonary position: meril's myval™

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Background and Aim: Transcatheter Pulmonary Valve Replacement (tPVR) into right ventricular outflow tract (conduit or native) is being increasingly performed in many centers. In this report we aimed to review the outcomes for the Meril's Myval™ (Navigator, Meril Life Sciences Pvt Ltd, India) for pulmonary position in patients with both native right ventricular outflow tracts (RVOTs) and conduits.
Methods: Between January 2020 and November 2021, 27 tPVR procedures with Meril's Myval™ were performed in two different centers. Among them 20 (74%) had native RVOT and 7 (26%) had conduits or valve. For native RVOT patients Andra XXL or Andratec XXL stents with Z-Med-II balloons preferred for prestenting. For counduits covered CP stent or Begraft™ stents were used. If RVOT is very large we prefer to wait 6-8 weeks after prestenting procedure before tPVR procedure.
Results: All the patients underwent RVOT stenting before tPVR. Procedure was unsuccessful in 1 of them. Median age and weight of the remaining patients were 18 (6.5-50 years) and 56 (23-85) kg, respectively. Primary underlying diagnosis was tetralogy of Fallot (n = 22/26). Stenting was performed simultaneously with tPVR in 22 (85%) cases whereas pre-stenting was performed 4 to 5 weeks

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earlier in 2 patients (2 patients had stents for a long time and referred to our hospital lately). Valve sizes were 23mm (n = 7), 26mm (n = 3), 29mm (n = 10), 30.5 (n = 1) and 32 (n = 5).One patient's valve was migrated to RV and couldn't be reposed to main pulmonary artery. One patient had renal artery bleeding and underwent left upper lobe renal arter embolization. Three patients had minimal paravalvular leakage. There were no deaths during a median follow-up of 7 (1-12) months.
Conclusion: The Meril's Myval™ may be an alternative for patients in patients with RVOT dysfuction both native and conduits. One of the major advantages over other tPVR is larger (30.5 mm and 32 mm) valve sizes which can be used effectively and successfully in patients with large RVOTs. The advantages of Myval™ can be stated that all sizes work from a 14F sheath, the valve size spectrum is very wide and it can be manipulated more easily compared to its equivalents.

Keywords: percutaneous pulmonary valve implantation, Meril's Myval, Tetralogy of Fallot

P-298
Percutaneous interventions in fontan circulation: a single center experience

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Background and Aim: Several percutaneous interventional procedures are needed to maintain adequate anatomical and physiological conditions for the Fontan circulation. We purpose to describe the experience gained at a single center hospital and to determine the type and incidence of hemodynamic abnormalities requiring catheter interventions.
Methods: Retrospective study of all patients with Fontan circulation completed between 2014 and 2021. Patients were included if they received any form of total cavo-pulmonary connection, including the lateral tunnel and extracardiac Fontan operations, as well as hepatic vein inclusion conduits in patients with a prior Kawashima operation. We analyzed the clinical characteristics and the different types of percutaneous interventions performed, considering the post-Fontan period of time.
Results: Between January 2014 to December 2021, 30 patients underwent 50 transcatheter interventions. All patients had undergone cardiac catheterization either because of hemodynamic instability (Low cardiac output syndrome: LCOS) or desaturation. Median age at catheter palliation procedure was 9.4 years (range 3-24 years) and the weight was 31.5 (13-104 kg). The procedures were performed after Fontan, or Kawashima surgery in 21 (70%), 9 (30%) patients respectively. More than one intervention was applied to 11 of the patients. Transcatheter procedures were performed in different sessions in 16% (n:5) of the patients. Among these patients the most performed intervention is the closure of veno-venous fistula 11/50 (%22). Transcatheter Fontan fenestration interventions were performed 17/50 (%34); fenestration closure in 9 and Fontan fenestration dilation/creation in 8 patients. Among the patients that branch pulmonary artery stenosis was determined, stent implantation was performed in 7 (%14) patients. Residual antegrade pulmonary flow (APF) was occluded in 4 (%8) patients with various devices. In 7 patients (%14) with aortopulmonary collaterals, coil occlusion was performed in order to reduce future systemic ventricular volume load. There were no procedural mortality and only temporary AV block was observed in only one patient, and no complications developed in the others.
Conclusion: Interventional catheterization procedures are often necessary to maintain the delicate Fontan circulation. These interventions can help many clinical problems and may avoid some operations. Interventions prolong life expectancy in failing Fontan and they play a critical role.

Keywords: Percutaneous intervention, Fontan, fenestration

P-299
Stenting of left main coronary artery stenosis in an infant after anomalous left coronary artery from the pulmonary artery repair

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Background and Aim: Anomalous left coronary artery from the pulmonary artery (ALCAPA) is a congenital coronary artery anomaly comprising 0.5% of all congenital heart diseases. The treatment of choice in patients with ALCAPA is establishing a dual coronary artery system with either surgical reimplantation of the left coronary artery into the left coronary sinus or creation of an intrapulmonary artery. This newly created coronary artery system is prone to stenosis or total occlusion in the postoperative period. Transcatheter intervention options are now being considered due to the high risk of reoperation.
Methods: Herein, we present the procedural technique in a two-month-old female patient with ALCAPA who had coronary stent implantation to relieve post-surgical LMCA obstruction.
Results: A two-month-old female patient diagnosed with anomalous left coronary artery from the pulmonary artery and severe congestive heart failure was referred to our center for surgery. Echocardiography revealed dilation of the left ventricle, reduced left ventricular ejection fraction (38%), a moderate degree of mitral regurgitation and endocardial fibroelastosis (grade 2). The electrocardiography showed pathological Q waves in the lateral leads. Cardiac catheterization demonstrated the anomalous origin of the left coronary artery. The patient underwent left main coronary artery reimplantation in the ascending aorta above the sinotubular junction. In the post-operative period, whenever the patient was tried to be weaned from the mechanical ventilator, her hemodynamics deteriorated. At that time, echocardiography showed that the left main coronary artery was filled with the retrograde flow. Also ST-T changes were observed in the electrocardiography. On the postoperative 23rd day, diagnostic cardiac catheterization was performed. Aortic root angiogram demonstrated almost total occlusion of the left main coronary artery ostium and retrograde filling from the right coronary artery. A 2.5x9 mm bare-metal stent was deployed in the left main coronary artery proximal to its bifurcation. Post-intervention angiography showed reestablished flow to the circumflex and the left anterior descending artery arteries.
Conclusion: Percutaneous coronary intervention is a feasible option in infants who develop coronary artery obstruction after surgical reimplantation for anomalous left coronary artery from the pulmonary artery if the risk of reoperation was believed to be unacceptably high.

Keywords: Left coronary artery, stent implantation, postoperative, anomalous left coronary artery from pulmonary artery

P-300

Bridge to surgery with pulmonary valve perforation and right ventricular outflow tract stenting in an infant with PA-VSD

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Background and Aim: In pulmonary atresia (PA)/ventricular septal defect (VSD), a rare congenital heart disease, blood flow to pulmonary arteries is provided by patent ductus arteriosus or major aorto-pulmonary collateral arteries (MAPCAs).Thus, in these cases palliative interventions are required in order to maintain the blood supply to the pulmonary arteries before the complete correction operation. Here, we report our case who underwent right ventricular outflow tract (RVOT) stenting and pulmonary valve perforation intervention as an alternative to systemic - pulmonary artery shunt (SPS), which is frequently used.
Methods: A 2.5-month-old cyanotic infant weighing 2500 g, followed up with the diagnosis of PA/VSD, was admitted to our institution. In the echocardiographic and cardiac CT examinations of the case, minimal antegrade flow was observed in the main pulmonary artery. In addition, well-developed branches of the pulmonary artery filled through the patent ductus arteriosus were observed. RVOT stenting and pulmonary valve perforation were applied to the case by cardiac catheterization. Moreover, it was sent to complete correction operation without any problems.
Results: When the patient was ten months old, catheterization was performed and she was referred to surgery for a complete correction operation and the surgery was done uneventfully.
Conclusion: Pulmonary valve perforation and RVOT stenting may be a safe and effective alternative in infants with PA/VSD.

Keywords: pulmonary valve perforation, pulmonary atresia, RVOT stenting, Tetralogy of Fallot

P-301

Transcatheter recanalization of the right pulmonary artery after surgical correction

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Background and Aim: An anomalous origin of the right pulmonary artery (RPA) from the ascending aorta (AORPA) is a rare cardiac malformation. Partial or complete occlusion of the RPA might be seen following re-implantation of the RPA on the main pulmonary artery (MPA). An occluded RPA can be recanalized either surgically or by transcatheter methods.
Methods: A female infant, who underwent surgery for AORPA during the newborn period, was admitted to our hospital at five months old. She had a history of RPA re-implantation on the MPA at first. Then embolectomy was performed for proximal RPA thrombosis one month after the operation. Transthoracic echocardiography revealed a disconnected RPA that was confirmed with computed tomography angiography (CTA).
Results: On catheter angiography, contrast injection through the right ventricular outflow tract and MPA showed RPA stump without flow to the distal branches. Then, hand injection of contrast material to the stump showed a tiny passage to the RPA. It was decided to perform revascularization with catheter intervention. A 0.014-inch extrasupport coronary guidewire was passed through the obstructed segment. 2Fr microcatheter passed over the guidewire and localization was confirmed by hand injection. Predilation of the target lesions was performed with 2 x 15 mm, 4 x 15 mm angioplasty balloons inflated up to 16 atm pressure for 30 seconds and 6 x 20 mm Numed TyShak II® balloon, inflated up to 2.5 atm. Following predilation, proximal RPA stenosis, measuring 2.7 mm, still persisted. A 5F guiding catheter was advanced over the extrasupport coronary guidewire up to the distal RPA. A 4.5 x

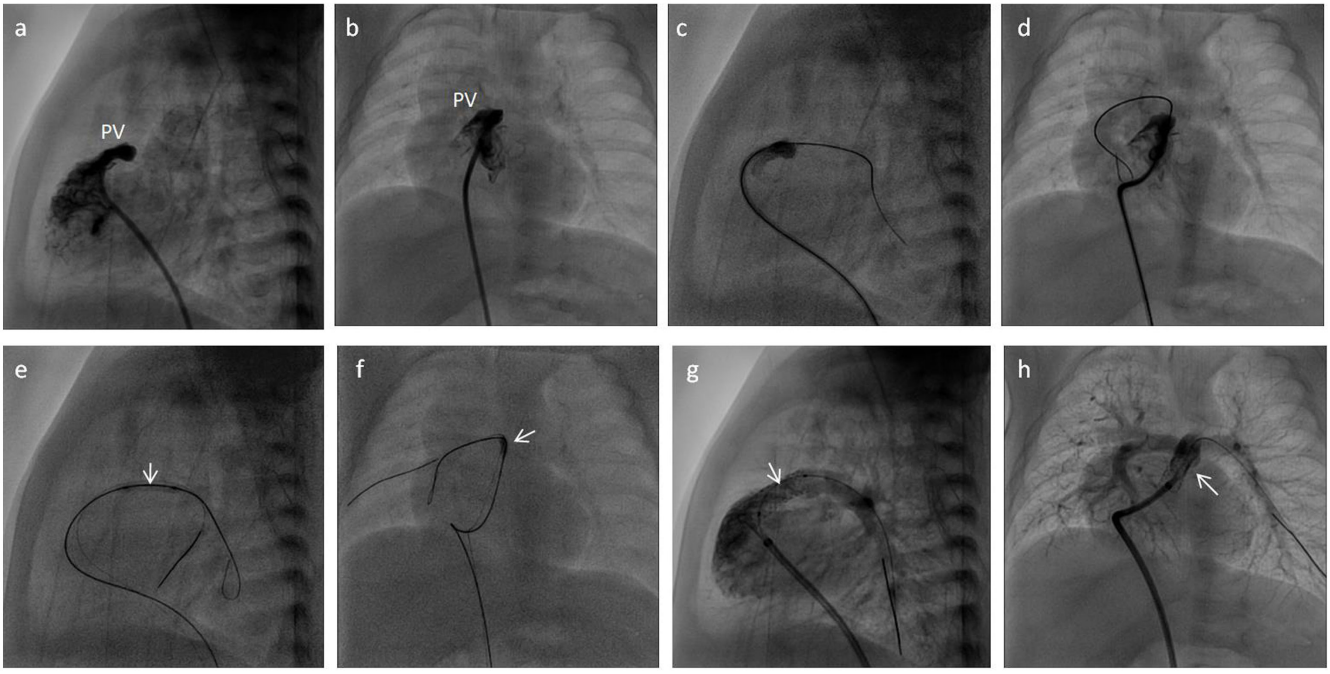

Figure 1. Catheterization images. Image of the atretic pulmonary valve (PV), lateral (a) and anteroposterior (b) view. Perforation of the pulmonary valve using the Conquest Pro CTO guidewire, lateral (c) and anteroposterior (d) view. Predilatation with coronary balloon (white arrow), lateral (e) and anteroposterior (f) view. Image of the adequate flow in the branch pulmonary arteries after the RVOT stent (white arrow), lateral (g) and anteroposterior (h) view.

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12 mm bare metal coronary stent was successfully deployed into the proximal RPA. The post-deployment angiogram showed optimal stent implantation with revascularization of RPA. The procedure was completed without any complications. CTA and transthoracic echocardiography three months after catheter intervention showed that RPA was patent and there was no stent obstruction. (Figure)

Conclusion: Transcatheter recanalization might be possible, especially in postoperative pulmonary artery obstructions.

Keywords: Transcatheter recanalization, right pulmonary artery from the ascending aorta, pediatric

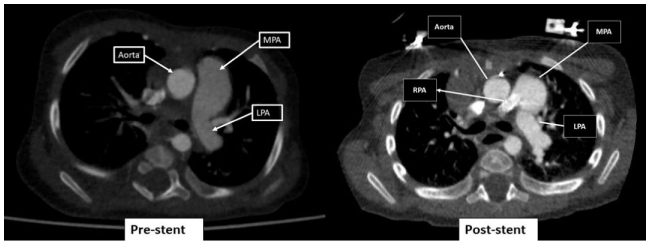

Figure 1 Pre-stent:Computed tomography angiogram demonstrating right pulmonary artery disconnection Post-stent: Computed tomography angiogram demonstrating right pulmonary artery connection and patent stent lumen.

P-302

Percutaneous pulmonary valve implantation in pediatric patients: a single center experience

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Background and Aim: Percutaneous pulmonary valve implantation (PPVI) is a feasible and low risk alternative to surgery in previously operated patients. Evolving technology allowed the development of various prostheses and expanded PPVI indications from an initial treatment exclusively of stenotic conduit to wide native right ventricular outflow tract (RVOT). Melody transcatheter pulmonary valve (TPV) and Edwards Sapien valve are the most commonly implanted prostheses. Indications for PPVI are the same as for surgical interventions but limited with the maximum diameter of the available percutaneous prosthesis. Therefore, an accurate preoperative evaluation is of paramount importance to select patients who could benefit from this procedure. This study aims to evaluate the PPVI experience in our center.

Methods: Patients who underwent PPVI between January 2015 and March 2020 were included in the study. The clinical and demographic characteristics of the patients, procedure details, complications and follow-up data were recorded retrospectively using the hospital database.

Results: Seventy-two of the patients were boys, and 55 were girls. The mean age was 18,5 years (5,8-63 years), and the median weight was 55,2 kg (17–100 kg). The primary underlying diagnosis was tetralogy of Fallot (n = 100/127). The most common symptom was fatigue. Stenting was performed simultaneously with valve implantation in 71/127 (56%) cases-six of which were hybrid procedures. Prestenting was performed 3 to 160 weeks earlier in 57/127 cases. Prosthetic valves were implanted to native RVOT or conduit in 85 and 42 patients, respectively. During the angiography, the stents' median right anterior oblique and lateral diameters were measured 24,5 mm (14–34,5 mm) and 23 mm (12,7–33 mm), respectively. Implanted valve sizes were 20-30 mm (n = 115) for Edwards Sapien XT and S3, 18-24 mm (n = 12) for Melody. There were no deaths during 86 months follow-up (median 43,5 months). Prosthetic valves were functional in all patients.

Conclusion: PPVI can be a valid and safe alternative to surgical interventions. A careful anatomical and hemodynamic assessment are mandatory to select ideal candidates for this procedure. Moreover, new valves and developing technology will also extend indications to patients with very large RVOT.

Keywords: Percutaneous pulmonary valve implantation, Pediatric, Melody transcatheter pulmonary valve, Edwards Sapien valve, tetralogy of Fallot

P-303

Trans-axillary artery approach for ductal stenting in neonates and infants; experience of two cardiac centers

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Background and Aim: Ductal stenting in arterial ducts with unusual anatomies and origins represents a challenge for pediatric cardiac intervention. The axillary artery route offers a safe and effective option for access to these ducts.

Methods: Between November 2018-June 2021, trans-axillary artery access was performed for ductal stenting on a total of 30 patients in two cardiac centers. The clinical and demographic characteristics of the patients, procedure details, complications, and follow-up data were recorded retrospectively using the hospital databases.

Results: A total of 30 patients, 12 female (%40) and 18 male (%60), were included in this study. The mean age was 24,8 days (3-131 days), the mean body weight was 3235 gr(2100-5000gr), the mean procedure time was 66,6 minutes (22-137 minutes). Eight had aortic outlet right ventricle with pulmonary atresia (PA)(%26), six had tricuspid atresia with PA (%20) six had PA with ventricular septal defect (%20). Two patients had aortic coarctation, and one had ductal dependent systemic blood flow due to aortic valvar obstruction. The others had other complex congenital heart defects with PA or near atresia.

The axillary arterial route was the first choice in 14 patients; for the others, after the femoral artery and/or venous routes were unsuccessful, the axillary arterial approach was preferred.

For five of the patients, ultrasound guidance was performed. Bare metal coronary stents were deployed successfully in 28 of 30. In one of them, the ductal spasm was developed while positioning the wire, and the other was taken to the operation emergently due to stent migration.

There were no procedural mortalities, but after one week following the procedure, one of the patients was died because of pulmonary hemorrhage. The long-term vascular sequelae of axillary artery catheterization were negligible, and there were no vascular pathologies. Only one had brachial plexus paralysis, and now well after physiotherapy. The mean follow-up time was 8,2 months.

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Conclusion: Alternative arterial access other than the femoral artery route is an option for neonates and infants with high surgical risk.

Keywords: trans-axillary artery access, ductal stent implantation, pediatric cardiac catheterization

P-304
Interventional cardiology in paediatric heart transplantation

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Background and Aim: Paediatric heart transplant is associated with several cardiac catheterizations. Analysis of the interventional procedures can offer a better understanding and management of these complex patients.
Methods: All cardiac catheterizations performed in heart transplanted children in a tertiary centre from 2002 to 2021 were included. All medical history, previous catheterisms with their complications and results were retrospectively reviewed.
Results: Sixty-four patients (31 males) were obtained. Dilated cardiomyopathy was the basal disease in 54.84% of the subjects. 55.55% of the patients had undergone a previous cardiac catheterization. It consisted on 92 procedures, a mean of 1.44 ± 2.00 standard deviation (SD) catheterization per person. 69.56% were diagnostic procedures, of which 11.11% endomyocardial biopsies. Regarding therapeutic catheterizations, the most prevalent techniques were major aorto-pulmonary collateral arteries closure (13.76%) followed by pulmonary arteries angioplasty with or without stenting (11.84%).
Transplantation occurred at a mean age of 96.24 months (± 89.47 SD). 19.00% of patients needed extracorporeal life support. Mean intensive care unit stay length was 33.65 (± 49.77 SD) days.
After a mean follow up of 6.55 years, 389 cardiac catheterization were performed, including 608 procedures (see figure 1). It resulted in 6.71 ± 4.13 catheterizations per person on average (SD). The most performed technique was endomyocardial biopsy (62.99%), obtaining a 0-1 grade cellular rejection in 351/383, grade 2 in 27/383 and 1 patient showed a grade 3 cellular rejection. 2 cases suffered a 2 grade class humoral rejection. The second most prevalent procedure was coronariography (18.25%) followed by coronary intravascular ultrasound (11.50%). The latter obtained a 31.42% of positive results, the most affected arteries were the anterior descending coronary (17/22 of cases) and left common coronary ostium (15/22), being both territories affected in 10 studies.
Angioplasties, were the fourth most performed technique, more frequently directed to cava veins, where stent implantation was considered necessary in 23.08% of cases.
90.00% of patients were alive at follow up.
Conclusion: This study, accordingly to previous bibliography, shows that endomyocardial biopsy is the most frequent procedure in children with heart transplantation. Added to that, these patients may need interventions on the surgical anastomosis, being cava veins the mainly affected territories.

Keywords: Heart transplant, Interventional Cardiology, enodmyocardial biopsy.

Interventional Cardiology after paediatric heart transplant

PROCEDURES	NUMBER
Endomyocardial biopsy	383
Coronariography	111
Coronary intravascular ultrasound	70
Other diagnostic procedures	7
Superior cava vein angioplasty	10
Superior cava vein stenting	2
Inferior cava vein angioplasty	3
Inferior cava vein stenting	1
Right ventricle-pulmonary artery tube angioplasty	1
Pulmonary artery angioplasty	9
Pulmonary arteries stenting	3
Angioplasty coronary	1
Coronary stenting	1
Marginal coronary artery thrombolysis	1
Cava vein thrombi related procedures	3
Pericardiocentesis	1
TOTAL	607

P-305
"Kissing Stents": An alternative interventional treatment for severe bilateral pulmonary branch stenosis following congenital heart disease surgery

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Background and Aim: Bilateral branch pulmonary stenosis is a common lesion in complex congenital heart diseases (CHD), e.g. Tetralogy of Fallot, or it can be a frequent residual lesion following CHD surgery, e.g. d-TGA s/p Arterial Switch Operation (ASO). Frequently, interventional procedures are used alternatively to open heart surgery. "Kissing Stents" have been introduced in the early 1990's as an alternative interventional treatment of patients suffering from aortoiliac occlusive disease. In our knowledge the use of simultaneous bilateral "Kissing Stents" for treatment of bilateral branch pulmonary stenosis is rare and only a few cases have been reported.
AIM: Present 2 cases of postoperative severe bilateral pulmonary stenosis treated successfully and followed-up for a year, using simultaneous bilateral "Kissing Stents" technique
Methods: Case1: male 9.5 years old, suffering from d-TGA, s/p ASO (02/2012). Residual neo-pulmonary valve stenosis, mPA, LPA stenosis, i/p balloon valvoplasty and LPA dilatation (11/2014). Restenosis evolved over time in both mPA, LPA, s/p (9 &10/2019) "Y" shape pulmonary homograph repair. Two years later sever stenosis in RPA (diameter 2.0mm) and LPA (diameter 5mm), by c-MRI.
Balloon valvoplasty and simultaneous placement of "Kissing Stents" 10/2mm to RPA 8/3mm to LPA percutaneous by (R) &(L) femoral's and (R) internal jugular veins.

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Case2: male 10months old, suffering from ToF s/p total repair 11/2020.Seven months later presented with severe bilateral stenosis of (R) &(L) BPAs at their branching area with RPA (diameter 2.0 mm) and LPA (diameter 3.0mm) by c-MRI. Balloon valvoplasty and simultaneous placement of "Kissing Stents" 7/2mm to RPA 8/2mm to LPA percutaneous by (R)&(L) femoral's and (R) internal jugular veins.
Results: Case1: Systolic pressures drooped i/p from 40 to 13 mmHg in RPA and 45 to 21 mmHg in LPA. No complications were noticed. Case2: Systolic pressures drooped i/p from 38 to 6mmHg in RPA and 40 to 6 mmHg in LPA. Transient second-degree A-V block was noticed for a few hours.
Conclusion: Simultaneous placement of "kissing Stents" following balloon valvoplasty can be an safe and excellent, minimal invasive alternative in treating bilateral branching PA stenosis following surgery for complex CHD.

Keywords: Simultaneous "Kissing Stents" placement, bilateral branching PA stenosis

P-306
Percutaneous closure of a giant coronary fistula

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Background and Aim:. The management of the cornary artery fistulas is controversial, especially in asymptomatic patients. Treatment options are conservative, surgery and trancathter closure.
Transcatheter occlusion of CCF is now considered the treatment of choice for most fistulae.
Methods: We present the case of a 3 year old girl with a giant coronary cameral fistula who was admitted for possible interventional catheterisation. She was diagnosed echocardiographically with a giant coronary cameral fistula from the left coronary artery to the right atrium at the neonatal age. She had been admitted to the neonatal unit of a different hospital with tachypnea, tachycardia and poor weight gain. A continuous precordial systolic high frequency murmur was audible. Due to her small weight, she was managed conservatively and was followed up periodically every three to six months.
Subsequently, at 3 years of age with her weighting 18 kgs, interventional closure of the defect was attempted. She was well grown, appropriate for her age and height, without any signs of heart failure. s1+s2 were normal, with an ejection systolic murmur at the lower left sternal edge. The resting electrocardiogram was normal, with no evidence of strain or ischaemia.
Results: Under general anesthesia, the right femoral vein was percutaneously cannulated with a 5 French short (pediatric) sheath and right femoral artery with a 4 French sheath. Heparin 50 Units/Kgr was administered after vascular access was obtained. The fistula was accessed from the femoral artery, using a 4F Judkins Left catheter and a 0.014" straight, 320 cm long guidewire. The wire was stabilized in right atrium and an arteriovenous loop was formed.The venous branch of the loop an Amplatzer Duct Occluder II 4/4 was advanced, introduced and deployed in the fistula. Total fluoroscopy time was 7 minutes There were no complications during the procedure.
Conclusion: The majority of coronary cameral fistulas are from the RCA, LCA, or both coronary arteries and usually drain into the RV, Pulmonary artery, RA and LV.Transcatheter occlusion of CCF is now considered the treatment of choice for most fistulae.

Keywords: coronary fistula

P-309
First serie of transcatheter correction of sinus venosus atrial septal defect using the new covered optimus XXL 99mm stent

Clement Batteux[1], Vlad Ciobotaru[2], Nicolas Combes[3], William Arditi[4], Clement Karsenty[5], Jerome Petit[1], Gregoire Albenque[1], Emre Belli[1], Philippe Brenot[1], Alain Fraisse[6], Sebastien Hascoet[1]
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Background and Aim: Transcatheter correction using customized covered, Cheatham-Platinum stents has emerged as an alternative to open-heart surgery for sinus venosus atrial septal defects. The 50 and 60 mm-long stents subsequently received CE marking and FDA approval. The longer sizes are nowadays unavailable despite that the anatomical configuration of the defect often requires a stent longer than 70 mm. Using the newly developed, covered, 99 mm-long, Optimus XXL stent, we report the first serie of 3 successful transcatheter correction of sinus venosus atrial septal defects.
Methods: We first assessed the mechanical properties of the uncovered and covered Optimus XXL 99mm stents on 3D-printed models of the 3 patients. (Figure, 2)
For the 3 patients (a 43-year-old man, a 25 year-old woman and a 55 year-old woman), we assessed procedure feasibility by following a step-by-step simulation program that included 3D CT scan modeling, virtual simulation, 3D printing, and hands-on simulation training. (Figure, 1, 2, 3)
Results: Procedures were performed with compassional agreement of the Agence Nationale Pour la Santé et le Médicament, the french authority for medical device regulations.
Case 1 and 2 (Figure, 1 and 2): Covered Optimus XXL 99mm stents were mounted on a customized balloon-in-balloon (BIB, NuMED, Hopkington, New York) 22 mm in diameter and 80 mm in length. The stent was stable and well positioned. Post-dilations of the two ends of the stent was needed to fully open the stent. The lower end was also flared using a large Coda Balloon. A tiny residual shunt was deemed acceptable. The patient experienced no adverse events and was discharged on day 2
Case 3 (Figure, 3): A covered Optimus XXL 99mm stent was mounted on a BIB balloon 24 mm in diameter and 80 mm in length and inflated with the same technic of case 1 and 2. The upper end was anchored by an additional uncovered Optimus 37mm stent flared toward the innominate vein. The lower end was also flared, using a large Coda Balloon. The patient experienced no adverse events and was discharged on day 2.
Conclusion: The new Covered Optimus XXL 99mm showed a satisfying and expected mechanic profile for SVASD percutaneous correction. Its specific lenghts allowed good stability during inflation and its conformation seemed to be accurated for this new

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procedure. Biggest study is needed to settle this new device for a day-a-day use.

Keywords: Sinus venosus atrial septal defect – new device – covered stent - 3D modeling – Percutaneous correction

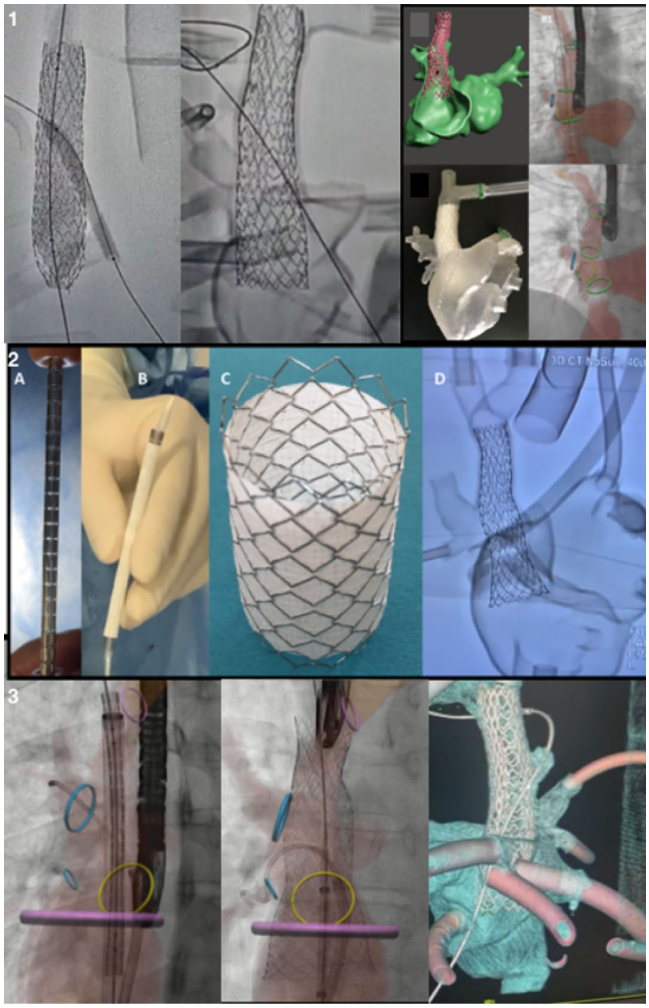

Figure 1 virtual simulation, bench-testing and in vivo implantations of the covored Optimus XXL 99 mm.

P-310

The effects of pulmonary branches stenting on the pulmonary arteries hemodynamic and growth kinetic

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Background and Aim: Pulmonary arteries (PA) stenting in children is controversal because of the need for reintervention and the impact on PA growth. The aim of this work is to study the PA growth after pulmonary arteries stenting (PAS).

Methods: A retrospective monocentric study including all patients who had PAS between 1992 to 2017. Hemodynamic evaluation was done during catheterization. PA growth was assessed by CT angioscan with the measurement of CT derived Nakata Index (CDNI) and the PA surface on multiplaner reconstruction (PAS-MR).

Results: 47 patients were included. Median age was 12 years [range 0.2 – 46years], median weight was 36.1kg [range 5.1 – 104kg]. Outlet malformations were the predominant pathologies (70.2%). Genetic disorders were presents in 7 cases (14.9%). Reintervention was necessary in 31 patients (65.9%) and in all patients < 2 years. Median invasive gradient across the stenosis decreased from 34.0 ± 21.4 mmHg to 18.6 ± 15.0mmHg ($p<0.001$) for the left stented PA and from 39.0 ± 23.9mmHg to 18.2 ± 13.6mmHg ($p<0.001$) for the right stented PA. Mean right ventricle to aortic systolic pressure ratio dropped from 68.4% ± 25.9% to 50.9% ± 17.4% ($p<0.001$). At follow up, 30 patients had CT control after PAS with median interval of 2.18 years. Median CDNI increased from 71.0mm/m² [IQ: 50.7 – 107.7mm²/m²] to 95.0mm²/m² [IQ: 69.5 – 141.0mm²/m²] ($p = 0.003$), and median PAS-MR increased from 69.5mm²/m² [IQ: 46.5 – 100.0mm²/m²] to 96.0mm²/m² [IQ: 67.5 – 124.0mm²/m²] ($p = 0.013$) on the stented PA. The rate of growth of the stented PA was similar to the controlateral normal PA and the expected PA growth in general population (figure 1). CDNI was correlated to PAS-MR for the left PA (93,0mm²/m² [54.5 – 150.0mm²/m²] vs 95,0mm²/m² [56.7 – 159.0mm²/m²] respectively ($p = 0.78$). Contrarily, CDNI overestimated the surface of the right PA comparing to PAS-MR (167.0mm²/m² [76.5 – 322.0mm²/m²] vs 214.0mm²/m² [78.5 – 363.0mm²/m²] respectively ($p<0.001$).

Conclusion: PAS is efficient to improve hemodynamic conditions. Distal PA growth remains normal comparing to the controlateral branch and the general population. PAS-MR is more accurate than CT derived Nakata index to measure the PA real surface, mainly for the right PA.

Keywords: Pulmonary arteries; Stent; CT derived Nakata index; Multiplanar reconstruction; Growth kinetic; Predictive model

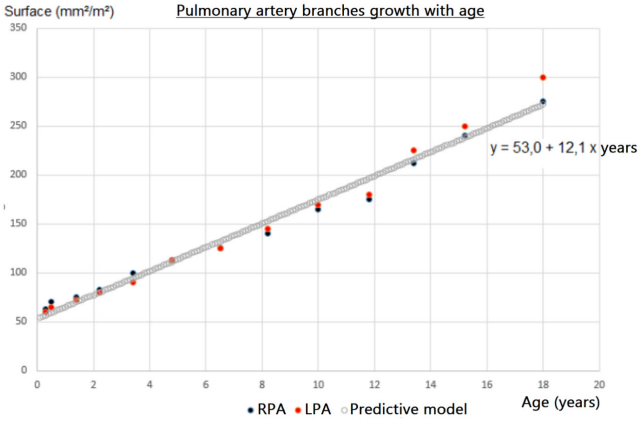

Figure-1 Pulmonary branches growth comparing to predictive model of pulmonary artery growth in normal population. LPA: left stented pulmonary artery, RPA: right stented pulmonary artery.

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P-313

Safety and efficacy of aortic coarctation stenting in children and adolescents: wondering the safest weight to perform percutaneous treatment

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Background and Aim: Percutaneous aortic coarctation treatment with primary stent implantation is the first choice in adult patients. However, current guidelines do not suggest a lower weight limit to perform this procedure safely.
The aim of this study was to retrospectively analyze the safety and the mid-term outcome of aortic coarctation stenting in pediatric age at different patients' weights.
Methods: We retrospectively enrolled 47 patients, 8 of them weighting lower than 25 kg, 10 with a weight between 25 and 30 kg and 29 patients with a weight >30 kg.
Results: Covered CP stent was used in 32 patients (68.1%), bare CP stent in 6 (12.8%), Andrastent XL in 8 (17.0%) and Palmaz Genesis in one case (2.1%). The balloon mean diameter was 13.8 ± 2.4 mm, range 10-18 mm. The procedure was effective in all patients. The aortic gradient dropped from 28.0 ± 7.8 mmHg to 2.2 ± 2.0 mmHg ($p<0.0001$). Hemostasis was achieved by a surgical cutdown in 20 (42.5%) patients, manual compression in 2 (4.3%) or by vascular closure devices (VCDs) in 25 (53.2%) patients. There was no difference in terms of efficacy, safety and complication rate among the three weight-based groups. We found a trend toward higher incidence of vascular complications following hemostasis with VCDs (4/24, 16.7%) vs surgical cutdown (1/21; 4.8%)
Conclusion: In conclusion, aortic coarctation stenting is a safe procedure in patients weighting less than 30 kg. Surgical arterial cutdown can minimize the risk of vascular injury by reducing the stress on the arterial wall in smaller patients or in case a large sheath is needed.

Keywords: aortic coarctation, stent, Interventional Cardiology

P-314

Percutaneous approach to residual pulmonary bifurcation stenosis in conotruncal diseases

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Background and Aim: Residual stenosis after RVOT surgery represents a difficult problem to manage in patients with conotruncal defects. Despite a detailed multimodality imaging, the anatomic characterization of distal pulmonary trunk and pulmonary artery bifurcation may be difficult in these patients.
The aim of this study was to retrospectively analyze the outcome of the percutaneous treatment in pediatric patients with post-surgical stenosis of pulmonary artery bifurcation.
Methods: We reviewed all the procedures performed on patients with conotruncal disease and post-surgical lesions involving the pulmonary artery bifurcation and the origin of one or both the pulmonary arteries between January 2014 and March 2021.
Among the 263 patients treated in our center for RVOT and pulmonary artery angioplasty and/or stenting, were identified 32 eligible patients.
Results: The median age of 5.9 years. Standard high pressure balloon approach was attempted in 26 patients, effective in 5. A standard pulmonary branch stenting was performed in 10 patients, effective in 6. A kissing balloon approach was chosen in 9 patients (6 after angioplasty or stenting failure), this technique was effective in 8 cases. Finally, a bifurcation stenting was performed in 11 patients (second step in 9 cases), effective in all the cases. In no patient approached by kissing balloon a bifurcation stenting was needed.
Compared to bifurcation stenting, patients treated with a kissing balloon approach were younger (4.6 ± 5.0 yrs vs 15.2 ± 9.0 yrs). In addition, 10/11 of bifurcation stenting group were ToF and only one had a TGA, while in kissing balloon group 5 patients had TGA, 3 ToF and one truncus arteriosus.
The mean follow-up was 25 ± 18 months. Between the 11 patients treated by bifurcation stenting, echocardiography showed a RV pressure of 31.3 ± 8.8 mmHg and a RVOT peak gradient of 25.4 ± 13.1 mmHg. Between the eight patients treated by kissing balloon, the RV pressure was 44.2 ± 8.8 mmHg, RVOT peak gradient by echo was 29.0 ± 11.5 mmHg.
Conclusion: Standard balloon angioplasty and standard stenting may be ineffective in post-surgical stenoses involving pulmonary artery bifurcation. In these patients, a kissing balloon or (when a percutaneous pulmonary valve replacement is indicated) a bifurcation stenting followed by side branch de-jailing are indicated to effectively relief the gradient.

Keywords: Tetralogy of Fallot, pulmonary artery stenosis, pulmonary branches stenosis, stent, kissing balloon

P-315

Assessment of right ventricular mechanics in pediatric patients with valvular pulmonary stenosis before and after balloon valvuloplasty

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Background and Aim: Percutaneous balloon pulmonary valvuloplasty (BPV) is the treatment of choice in severe pulmonary valve stenosis. However, little is known regarding right ventricular mechanics immediate changes after percutaneous balloon dilation. This study sought to explore right ventricle (RV) mechanics in pediatric patients early after percutaneous BPV
Methods: 43 pediatric patients (19 males), mean age 3,2 ± 4,9 years with severe pulmonary valve stenosis and indication for percutaneous BPV were recruited. All patients underwent standard transthoracic echocardiography (TTE), and speckle-tracking echocardiography (STE) with analysis of right ventricle free wall longitudinal strain and strain rate (RVFWLS) one day before and one day after the procedure. For each patient were collected invasive parameters during the interventional procedure.
Results: After the procedure, there was an immediate significant reduction of both peak-to-peak transpulmonary gradient (peak-to-peak PG) and ratio between right ventricle and aortic systolic pressure (RV/AoP) with a drop of 29,3 ± 14,67 mmHg and 0,43 ± 0,03, respectively. Post-procedural echocardiography showed peak and mean transvalvar pressure gradient drop (50 ± 32,23 and 31 ± 17,97, respectively). The degree of pulmonary valve regurgitation was mild in 8% of patients before the procedure, following the intervention it reached 29% with a statistically significant increase ($p = 0,007$). However, the incidence of pulmonary valve moderate and severe regurgitation remained stable after

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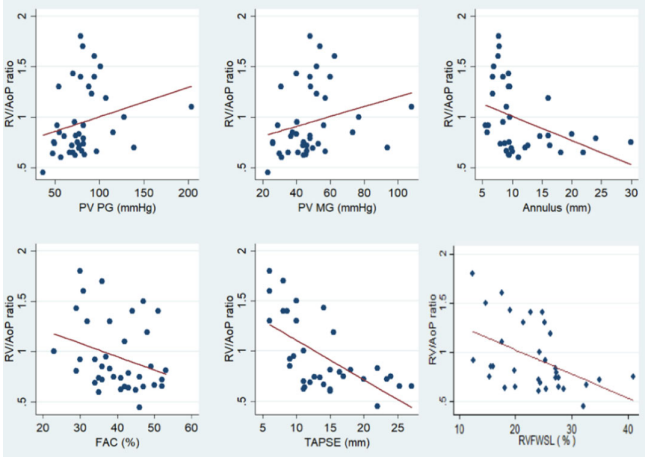
the procedure. The analysis of right ventricular function and mechanics showed a significant improvement of Fractional Area Change (FAC) immediately after the procedure (40,11% vs 44,42%, p = 0,01). On the other hand, right ventricular longitudinal systolic function parameters, TAPSE (p = 0,60) and RVFWLS (p = 0,31), did not improve significantly after intervention. Finally, pre-procedural invasive RV/LV ratio showed good correlation to echocardiographic transvalvular peak and mean pressure gradient (R = 0,375, p = 0,019 and R = 0,40, p = 0,012, respectively), as well as with FAC (R = 0,31p = 0,05), TAPSE (R = 0,62 p<0,001) and RVFWLS (R = 0,46 p = 0,01)
Conclusion: Percutaneous BPV represents an efficient and safe procedure to relieve severe pulmonary valve stenosis. RV showed an immediate improvement of global systolic function following afterload reduction. Conversely, longitudinal systolic function did not show signficant improvement. Finally, invasive preprocedural RV/AoP ratio demonstrated better correlation with echocardiographic evaluation of stenosis degree and right ventricular function compared to invasive peak-to-peak pressure gradient.

Keywords: Pulmonary valve stenosis; Balloon pulmonary valvuloplasty, Right ventricle mechanics, RV function, longitudinal strain

RV/Ao pressure ratio correlation with pulmonary stenosis degree



P-316
Treatment of 12-year-old girl with midaortic syndrome by using cook formula stent

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Background and Aim: Middle aortic syndrome (MAS) is a severe form of aortic coarctation and mostly involves the thoracic and abdominal part of the aorta. Traditional surgical approach has some potential risks and repetitive operations. Percutaneous stent implantation is a feasible and safe method for the palliation of MAS in childhood. Herein, a child with MAS was presented who has been treated by transcatheter stent implantation
Methods: After CT angiography showed stenosis in the distal thoracic and proximal abdominal aorta, cardiac catheterization was performed due to persistent hypertension despite maximum medical therapy. Before stent implantation, to safely advance the long sheath, a predilatation was performed using a 5 x 2 cm Tyshak II balloon. After then, a 40x9 mm cook formula stent was gradually inflated. Later, a 40 x 9 mm second cook formula stent was partially overlapped with the first and gradually inflated. As it was noticed that there was a pressure gradient in the distal part of the stents, a 7 x 58 mm stent was inflated gradually in overlap. Control angiography demonstrated good position of the stents with aortic final diameter of 9 mm and maximum gradient of 15 mmHg remained between the ascending and descending aorta.
Results: After the cardiac catheterization, the patient's hypertension regressed. CT scan revealed (Fig 1b and Fig 2b) a short part of the distal abdominal aorta still hypoplastic. One year after the procedure, the patient did not complain of any symptoms, and her blood pressure remained normal.
Conclusion: In conclusion, MAS is utterly a severe form of aortic coarctation and leads intractable hypertension and end organ failure. Because surgical approach has several limitations and risks of reoperation, transcatheter endovascular stent implantation should be kept in mind as a
feasible alternative treatment. But on the other hand, percutaneous interventions have been reported by other investigators, that there is a high incidence of restenosis and reintervention. This could be anyway a good example of effective stenting implantation with a relatively new hybrid stent family that provides the opportunity to perform stent redilatation over time.

Keywords: Balloon-expandable stents (BES), Cook formula stent, Endovascular procedures, Midaortic Syndrome, Stent implantation

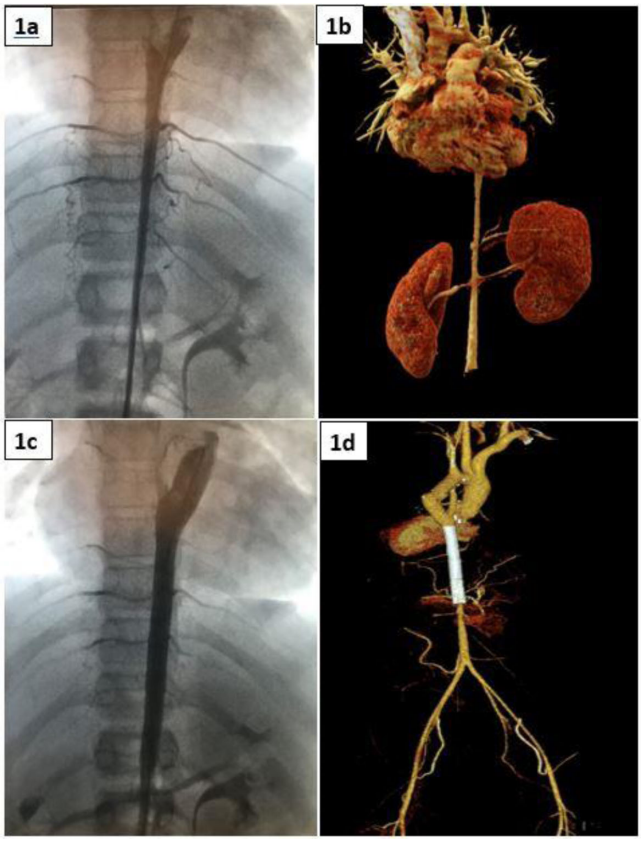

Figure 1. Angiography images before and after stent implantation (Fig1a and 1c). CT angiography images before and after cardiac catheterization (Fig 1b and 1d).

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P-317

The role of endomyocardial biopsy in children with left ventricular dysfunction

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Background and Aim: Endomyocardial biopsy (EMB) is a well-known diagnostic tool for the investigation and treatment of myocardial diseases and so far remains the gold standard for the diagnosis of myocarditis. Due to its invasivity with a complication rate ranging from 1% to 15%, its role in the diagnostic work-up of pediatric heart failure is not well established. Aim of this study is to define the clinical role and the safety of EMB in children presenting with severe left ventricular dysfunction.
Methods: We performed a retrospective review of the data of children who underwent endomyocardial biopsy in our Center from 2011 to 2021. Patients with heart transplantation were excluded from this analysis.
Results: During the study period 29 children underwent 30 EMBs for systolic LV dysfunction. Mean age was 7.3 year, 5 children (17%) were < 1 years old and 2 of them had a body weight < 8 Kg. Seventeen patients (57%) had a diagnosis of myocarditis, 9 patients received a diagnosis of primary dilated cardiomyopathy, 1 child had Carnitine deficiency, 1 a laminopathy and the patient whose presentation was an isolated right ventricular dysfunction had a Uhl disease (post-mortem finding). Viral PCR on myocardial specimen was positive in 12/17 patients (70%) and viral myocarditis was confirmed. 10 patients had PVB19 viral genome copies (83%), 1 patient had HHV6 viral genome copies and 1 patient had influenza A viral genome copies. EMB results led to a change in medical therapy in 12 patients (40%). Treatment changes were: antiviral therapy in 5 patients with viral myocarditis, immunoglobulin therapy in 6 patients, carnitine supplementation in 1 patient and immunosuppressive therapy in 1 patient with virus negative myocarditis. Regarding complications, cardiac perforation occurred in 3 patients (10%) weighting less than 20 Kg. All these patients survived except one
Conclusion: In the diagnostic work up of children with systolic heart failure, EMB-derived information are still essential prerequisites to establish an accurate diagnosis of myocarditis and a successful management of these patients. In our experience EMB results changed the clinical management of children with cardiomyopathy in 40% of the cases with a 10% rate of major adverse advents.

Keywords: Endomyocardial biopsy (EMB), myocarditis, LV dysfunction, cardiac perforation.

P-318

Interventional treatment of severe haemoptysis due to aortopulmonary collaterals in fontan patients

Felix Von Criegern, Jan Hansen, Gunther Fischer, Uebing Anselm; Department of pediatric cardiology, UKSH, Campus Kiel, Germany

Background and Aim: Bronchial bleeding with haemoptysis is a rare complication in Fontan patients. The bleeding usually occurs from aorto-pulmonary collateral vessels, which are commonly present in Fontan patients.
Methods: We present three cases with bronchial bleeding and haemoptysis from aorta-pulmonary collaterals and their successful treatment by endovascular embolization.
Results: All patients presented with significant haemoptysis resulting in respiratory compromise and dropping haemoglobin levels. Chest-X-ray, computer tomography (CT), bronchoscopy or a combination of these modalities localised the site of bleeding within the lung. Thereafter, aorto-pulmonary collateral vessels were identified by selective angiography and successfully embolised using microcoils or vascular plugs. In all three patients haemoptysis subsided after embolization so that the patients recovered quickly to be discharged from hospital within 5 days. Preprocedural localisation of the likely bleeding site seemed paramount to identify the culprit collateral artery angiographically. Large convoluted collaterals but also fine reticular arteries running in close proximity alongside the bronchial tree were identified as the bleeding source.
Conclusion: Development of aortopulmonary collaterals is a well-known phenomenon in patients with univentricular hearts palliated with the Fontan operation. In rare cases, aorto-pulmonary collaterals of variable size and morphology can cause bronchial haemorrhage. The cases presented here illustrate the importance of preprocedural imaging including bronchoscopy to locate the likely bleeding site into the lung as culprit vessels might be small.

Keywords: Fontan, Haemoptysis, Collaterals,

CoilEmbolization

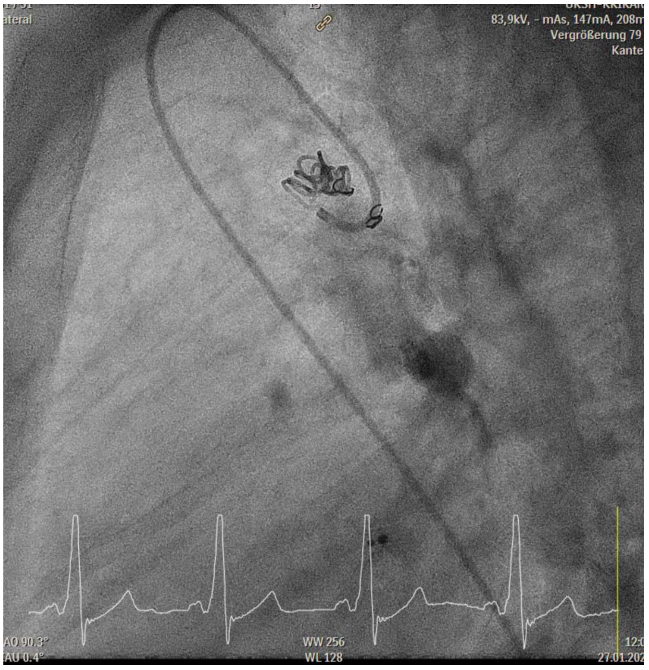

Figure 1. The picture shows the bleeding site with coils in situ right after embolization of the feeding vessel

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P-319

Stenting the arterial duct; a case of extended durability in the context of a duct-dependent functionally single ventricle circulation

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Background and Aim: Patent ductus arteriosus (PDA) stenting has evolved immensely since the 1990s. Indeed, it is a superior alternative to surgically constructed systemic to pulmonary shunts, especially in duct-dependent functionally single ventricle physiology where maintaining branch pulmonary integrity is of paramount important. Equally, it maintains superiority when comparing composite end-points (death from coronary steal and LCOS, over-circulation, hospital stay etc.). The technical nuance (deep learning curve) is acknowledged.
Drug-eluting coronary stents have been adopted from adults to modify the incidence of neo-intimal hyperplasia, albeit the pathogenesis in ductal tissue well distinguishable from atherosclerosis driven atheroma formation. Durability is particularly crucial in resource-limited environments with limited surgical capacity/timing. Herein, we describe a PDA stent that lasted 24 months unrevised.
Methods: A case narrative review. A primigravida, singleton pregnancy with no anomaly scan, delivering a 3kg term, male infant. Postnatal diagnosis was made during birth hospitalisation. Prostaglandins were instituted to maintain his convoluted PDA (sole source of pulmonary supply) in the setting of Tricuspid atresia-membranous pulmonary atresia. Branch pulmonary arteries were confluent but diminutive (< -2 z-scores). Rest of anatomy (including function) was favourable. Duct patency was maintained (saturations 80-90s) despite prostaglandins' deconstitution, with constriction observed from chronological 6 months (saturations 60s). Overlapping drug-eluting 5 mm coronary stents for length coverage were implanted. Anti-platelet therapy was constituted.
Results: Post-procedural saturation was 86%. Minimal neo-intima proliferation was observed serially. With somatic growth, he desaturated to 60-70s with rising haemotocrit. Pre-stage II palliation's assessment was performed before a bidirectional cavopulmonary shunt (+ take-down stent), at 2 years, 7 months. Baseline Nakata index was 263, mPAP 12mmHg, mTPG 2-3mmHg, PVRi 1.06iWu, PVRi/SVRi 0.05, LVEDP 8mmHg and no venous or systemic-pulmonary collaterals. Stage II palliation uneventfully achieved saturations > 82%.
Conclusion: While limited by numbers and single-centre experience, our case potentially demonstrates that drug-eluting PDA stents could last > 6 months. More reports are needed but the other conondrum that merits work is the description of potential inherent host factors attributable to this phenomenon, beyond drug-eluting stents. A tailored for purpose stent with ideal biologic properties (self-expandable, radial strength, low surface energy etc) to prolong durability, will be a landmark discovery.

Keywords: Staged single ventricle palliation, PDA stenting, drug-eluting stents, durability

P-320

Percutaneous closure of perimembranous VSD with the lifetech KONAR-MF device: mid–term results including the multifenestrated and gerbode type defects

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Background and Aim: The objective of this study is to report one center experience in VSD closure using the Lifetech multifunctional device including multifenestrated and Gerbode type perimembranous (Pm)VSD.
Methods: From March to October 2021, 27 consecutive patients were included in the study: 24 with PmVSD and 3 with muscular VSD. Eight had multifenestrated Pm VSD including 4 Gerbode-type VSDs.
Results: There were 15 males and 12 females, and the mean age at implantation was 21 years Implantation succeed in all under general anesthesia. A retrograde approach was used in 22 patients and antegrade approach in 5 patients. All but one received only one device. Nor embolization, neither complete AV block or device dislocation was observed. Retrograde approach was associated with lower dose of radiation. After release, immediate full occlusion was noticed in 30 % of the patients but rose to 2/3 at one month and 77% at 6 months. Six of the 8 patients with multifenestrated defect has no shunt. Persistent shunt were all tiny intraprosthetic leak.
Conclusion: MFO is effective and safe in VSD closure using both antegrade and retrograde approach with no AV block or valvular leak observed. The retrograde approach seems more convenient and was associated with lower dose of radiation. Longer follow-up with inclusion of more patients are mandatory to establish the safety of this technique.

Keywords: Perimembranous VSD, Konar MFO, VSD closure, Gerbode type VSD

P-321

Mid-term follow-up using lifetech KONAR-MF device for perimembranous and muscular ventricular septal defects

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Background and Aim: Treatment of hemodynamically relevant muscular ventricular septal defects (mVSDs) is still challenging for pediatric cardiac surgeons and interventionalists. The use of VSD closure devices has developed since 1988 and complication rates as particularly early and late-onset complete heart block have decreased to levels at or below those documented in surgical VSD closure. We evaluated the efficacy, safety, and outcome of transcatheter perimembranous (p) and mVSD closure using the novel KONAR-MF™ VSD occluder.

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Methods: Between July 2020 and August 2021, 6 patients with hemodynamically significant p and m VSDs underwent transcatheter closure using the KONAR-MF™ VSD occluder. 2 of these patients had a complex morphology, one swiss cheese VSDs and the other one a midmuscular to apical mVSD. All implantations were performed retrogradely under sedation, transoesophageal echocardiography, and fluoroscopic guidance. Prospective follow-up using transthoracic echocardiography and electrocardiogram were performed until December 2021.
Results: Median age and weight of all 5 pediatric patients was 179 days (6 months) and 7,3 kg, respectively. One adult patient (66 years) with a pVSD also underwent the procedure. Median follow-up time was 227 days. All devices were successfully and rapidly implanted. Median procedure duration was 123 minutes in all patients, 78 minutes in 4 uncomplicated cases and 281 minutes in the 2 cases with complex morphology. Sheath size was 4 or 6 French. In all patients, VSD closure was achieved. Mild residual shunting was detected by echocardiography in 3 patients directly after the intervention and in 4 patients at latest follow up. Only 1 patient revealed regurgitation of the atrioventricular and semilunar valves after intervention. 4 patients showed significant improvements regarding signs of heart failure. There was no complete heart block, major complications or death.
Conclusion: Transcatheter VSD closure by KONAR-MF™ is a promising tool in adult and pediatric patients even with complex morphology. Advantages are a softer design, the use of both an antegrade and retrograde approach, a small sheath size, and the lower risk of heart block. Further studies will be needed for evaluation of mid- and long-term outcome.

Keywords: Transcatheter VSD closure, KONAR-MF™, mid-term follow-up, infants

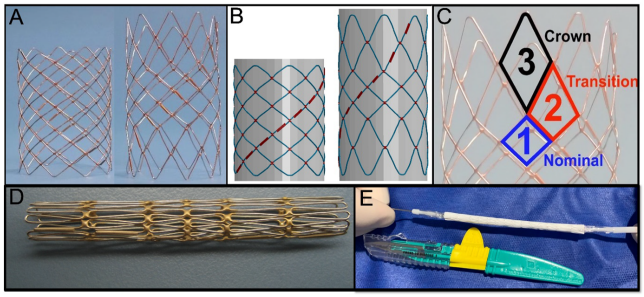
P-322/Moderated Poster
First in man use of a Novel FDA approved stent design for congenital heart disease- the G-ARMOR stent

Gareth Morgan, Jenny Zablah; The Heart Centre, University of Colorado

Background and Aim: We present the first-in-man clinical uses of a new hybrid cell structure covered stent, designed for Congenital Heart Disease applications, and approved by the US FDA. It represents a significant redesign of the Cheatham Platinum (CP) Stent (Numed Inc, Hopkinton NY), maintaining the traditional benefits of the covered CP whilst significantly decreasing shortening and allowing controlled flaring at the ends through its combination of larger and standard sized cells.
Methods: The stent was designed using a a combination of feedback from physician questionnaires. We then followed an iterative process using a combination of physical prototypes, CAD design simulation software and FEA analysis of the later prototypes. Stringent bench testing of the new design was performed to US FDA ISO standards. We implanted the stent in a 40yr old patient with a large superior sinus venosus defect involving the right upper and middle lobe pulmonary veins. The second case was that of a 55yr old man with complete atresia at the aortic isthmus. We used virtual reality systems for case planning and a 3D printed model for patient education and preparation. The procedures were guided using a combination of Vessel Navigator (Philips Medical Systems, The Netherlands) fusion imaging, 3D transesophageal echocardiography and traditional angiography and fluoroscopy.
Results: The clinical results were excellent in both cases. In the SVASD cases, there was no obstruction to pulmonary venous return and no visible L-R shunt on the transthoracic echo on 24hr and 2week follow-up. The patient with aortic atresia had an excellent angiographic, electrocardiographic and CT follow-up result with no angiographic stenosis and no significant gradient by any modality.
These are the first uses of this stent in clinical subjects.
Conclusion: The design is specifically aimed towards procedures where stent shortening is undesirable. Hence, coarctation of the aorta as well as stent implantation in preparation for percutaneous pulmonary valve placement are obvious use areas, as well as the growing body of evidence supporting percutaneous treatment of sinus venosus defects.

Keywords: aortic coarctation, covered stent, percutaneous pulmonary valve placement; conduit stenting, large vessel stent, sinus venosus atrial septal defects.

Schematic showing Design MOdifications with the New G-ARMOR Stent



By changing the wire length used to create some of the cells of the CP we were able to significantly alter the stent design and function without changing the mechanical properties, leading us to the G-ARMOR stent.

P-323
Single-centre experience of balloon valvuloplasty for severe aortic stenosis in childhood: could horizontal aortic root angulation predict outcomes?

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Background and Aim: Initial management of severe paediatric aortic valve stenosis involves surgical commissurotomy or transcatheter balloon valvuloplasty (BAV), often based on institutional preferences. Horizontal aortic root angulation measured via computer tomography (CT) has been implicated in procedure-related cardiac catheter complications in adults. As CT is not routinely performed in children prior to BAV, we hypothesised that aortic root angiography from left anterior oblique and 20° cranial angulation should enable robust angle measurements of the aortic root compared to surrounding structures, hence enabling risk stratification regarding BAV-complications.
Methods: We performed retrospective review of BAV cases between 2011-2021 at a Swedish tertiary paediatric CHD centre. There were no institutional changes to case selection and no changes to interventional team. Moderate/severe aortic regurgitation and death caused by BAV were defined as BAV-complications. Angle measurements between diaphragm, ascending aorta and aortic root were compared. Two blinded investigators analysed BAV-angiographies. Computational modelling of

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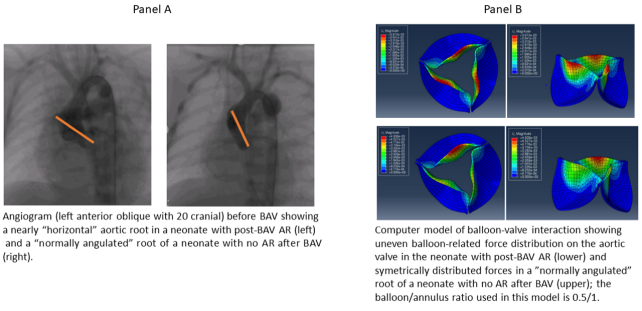
balloon-valve interaction (BVI) was designed to simulate outcomes for different angles in selected cases. Descriptive statistics (median/IQR), including measurement of interobserver variability are summarized.
Results: There were 17 BAV cases (males:16), age: 20/16-32 days; weight: 5.9/4.4-7.9 kg. 3/17 (18%) children had undergone prior heart surgery or BAV at other institutions. Follow-up was 40/25-95 months. Pressure gradient prior to BAV was 62/50-86 and afterwards 20/16-32 mmHg. Elective reinterventions occurred in 2/17 (12%). Aortic root to ascending aorta angulation could not reliably measured from angiography. Angles between diaphragm and aortic root showed good agreement (Cronbach's alpha = 0.82). BAV-complication in terms of AR occurred in 1/17 (6%). This neonate had the sharpest diaphragm to aortic root angle (<10th percentile; Figure/Panel A) and required balloon upsizing (balloon/annulus ratio (BAR) = 0.9/1) after initially unsatisfactory gradient reduction with mini-Tyshak balloon (BAR = 0.8/1). BVI simulation suggested that higher angulation during balloon inflation would lead to more uneven distribution of forces on the valve making associated AR more likely (Figure/Panel B).
Conclusion: The low incidence of BAV-complications in this small cohort necessitates further research assessing extreme aortic root angulations measurements on angiography to identify low versus high risk for BAV- complications. BVI modelling of these novel imaging markers could be used to guide future interventions.

Keywords: aortic valve stenosis, balloon valvuloplasty

Illustration of angle measurement from aortic angiography prior to balloon valvuloplasty (BAV) & simulation modelling of force distribution during BAV



Angiogram (left anterior oblique with 20 cranial) before BAV showing a nearly "horizontal" aortic root in a neonate with post-BAV AR (left) and a "normally angulated" root of a neonate with no AR after BAV (right).

Computer model of balloon-valve interaction showing uneven balloon-related force distribution on the aortic valve in the neonate with post-BAV AR (lower) and symetrically distributed forces in a "normally angulated" root of a neonate with no AR after BAV (upper); the balloon/annulus ratio used in this model is 0.5/1.

P-324

Cocoon devices for patent ductus arteriosus closure in children - our experience

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Background and Aim: Patent ductus arteriosus (PDA) is a frequent congenital heart disease being present in 5-10% of all. The interventional approach for PDA closure can have many possibilities and methods.
Complex PDAs are challenging especially when children are younger than 6 months old, the weight is inappropriate for the age (less than 6 kg), the diameter is large, the aorta is small, or there is disproportionated pulmonary hypertension.
Methods: Between 2015-2021, 135 cases of PDA were evaluated for PVR study or PDA closure in our laboratory.
90 of patients (pts) were female (66.6%) and 45 pts male (33.3%). Children were between 4 months old and 18-year-old.
Results: From all 135 pts (37.8% of our activity), 123 cases beneficiated of interventional closure (91.1%). We used both devices (Occlutech, Amplatzer, Cocoon) and coils (PFM and Cook). In 110 cases (89.4%) we used Cocoon devices. Of all the complex pts, age less than 6 months was present in 2 pts (1.6%), weight less than 6 kg in 6 pts (4.87%), complexity (Krichenko types C, D, E) in 45 pts (36.58%), small isthmic/descending aorta (<7 mm) 1 pts (0.81%), large diameter (>3 mm at angiography) in 41 pts (33.3%). We closed almost all Krichenko types of PDA by using the Cocoon devices, except type B. Type A (conical) was present in 86 cases (69.9%), type C (tubular) in 21 cases (17.07%), type D (complex) in 7 cases (5.69%), type E (elongated) in 15 cases (12.19%). There were 2 PDAs type B (window): 1 of them was closed with an ASD device, and another one had irreversible pulmonary hypertension.
Of the 110 patients who benefited from closure, the success rate was 94.5% (104 patients). 1 patient developed ductal spasm and the PDA could not be crossed, 2 patients had a significant gradient in the isthmus aorta - the procedure was discontinued and they were referred for surgery; 3 patients had a diameter of PDA≤1 mm and the duct could not be crossed.
Conclusion: PDA closure for different anatomical types and different ages is a safe method. Tips and tricks for anatomical difficult cases are essential for a positive result.

Keywords: patent ductus arteriosus, Cocoon devices, closure

P-325

Early results and short-term follow-up in transcatheter closure of patent ductus arteriosus in extremely preterm infants

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Background and Aim: The aim of this study was to inform early results and short term follow-up in transcatheter closure of patent ductus arteriosus (PDA) in the extremely preterm infants (EPIs) in our center.
Methods: The prospectively collected data of 14 EPIs who underwent transcatheter closure of PDA (TCCP) between December 2019 and October 2021 were evaluated. The transvenous antegrade approach was performed in 10 patients, guided by fluoroscopy, venous angiography, and transthoracic echocardiography (TTE) with the placement of the entire device within an intraductal position. The retrograde approach via the femoral artery was performed in 4 patients.
Results: Transcatheter closure was attempted in 14 EPIs with median gestational age 27 weeks (interquartile range (IQR) 25-28 weeks), median birth weight 913 g (IQR 793-1186 g) and was successfully performed in 12 EPIs (86%). The median

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procedural age was 31 days (IQR 26-48 days), median procedural weight was 1230 g (IQR 1120-1700 g). The median procedure and fluoroscopic time were 35 min (IQR 29.25-67.25 min) and 11.5 min (IQR 7-24.75 min) respectively (Table 1).
Three procedural complications were device embolization, pericardial effusion/cardiac tamponade, and femoral artery thrombosis. The embolized device was successfully retrieved, and the PDA closed spontaneously in a short-term follow-up. Pericardiocentesis was performed successfully and the effusion didn't accumulate again in the catheter room and intensive care follow-up. After the procedure, PDA closed spontaneously in this patient. Femoral artery thrombosis resolved with unfractionated heparin. The two procedural failures were related to the underestimation of ductal diameter due to ductal spasm and malposition of the device to the duct.
There were no procedural deaths, cases of post-ligation syndrome, residual PDA. Survival to discharge was 100%. After a median follow-up period of 9months (IQR 7-11 months), the device partially protruded into the LPA from the PDA with mild LPA stenosis in one patient. The gradient of LPA stenosis was reduced by pulmonary balloon angioplasty at 6 months of follow-up.
Conclusion: In our hospital, TCCP was performed with a high degree of success and an acceptable complication rate in EPIs. Based on our initial experience, early and short-term follow-up is encouraging.

Keywords: patent ductus arteriosus, preterm, transcatheter

Table 1. Patient demographics and details of the procedure

Patients	Gestational Age (weeks)	Birth Weight (g)	Procedure Weight (g)	Procedure Age (days)	Minimal PDA Diameter (mm)	Minimal PDA Length (mm)	Ventilator Dependent	Femoral Vascular Access	Procedure Time (min)	Fluoroscopy Time (min)	Device Size (waist diameter/ length mm)	Complications
1	27	920	1000	23	1.7	5	IMV	Vein	90	24	4/2[a]	None
2	28	1180	1280	24	2.2	5.5	IMV	Vein	75	27	4/2[b]	None
3	24	870	870	36	2	7.5	IMV	Vein	45	12	4/2[b]	None
4	28	770	1155	29	2.3	7	NIMV	Vein	25	10	4/2[b]	LPA stenosis at follow-up
5	29	1290	1885	52	2.8	8	NIMV	Artery	35	8	5/2[b]	None
6	24	600	1430	75	2	8	IMV	Vein	20	13	4/2[b]	None
7*	27	920	1140	30	2.9	8	IMV	Vein	95	32	4/2[b]-5/2[b]	Device embolization
8	25	750	1060	32	2.5	7.5	IMV	Vein	35	11	4/2[b]	None
9	26	860	1180	28	2.8	8	IMV	Artery	30	7	4/2[b]	None
10	32	1900	1990	22	3	8	-	Artery	30	6.5	5/2[b]	None
11	27	1205	1170	27	2.1	6.1	NIMV	Vein	55	16.5	4/2[b]	None
12	26	905	1500	47	2.3	9.1	IMV	Artery	32	5.5	4/2[b]	femoral artery thrombosis
13	27	1050	1900	41	2	6	IMV	Vein	27	7	4/2[b]	None
14*	26	800	1640	60	3.1	7	IMV	Vein	65	39	5/2[b]	pericardial efusion/ cardiac tamponade
Median	27	913	1230	31	2.3	7.5			35	11.5		

[a]=The Amplatzer Piccolo Occluder (Abbott Structural Heart, Plymouth, MN), [b] =The Amplatzer Duct Occluder II Additional sizes (ADO II AS), PDA = patent ductus arteriosus, * =procedural failures

P-326

Early outcomes of the treatment of aortic coarctation with begraft aortic stent in children and young adults

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Background and Aim: In this study, we reported early outcomes of using BeGraft aortic stent in the treatment of aortic coarctation (CoA) in our center.
Methods: This is a single center retrospective analysis of 11 implanted BeGraft Aortic stents (Bentley InnoMed, Hechingen, Germany) in CoA between July 2020 and November 2021. Advantages of this stent are deployement through a smaller sheath and its low profile.
Results: The BeGraft aortic stent was used in 11 patients (10 males, 1 females) with a median age of 13.7 years (IQR (interquartile range) 12–16 years) and a median weight of 43 kg (IQR 35–62 kg). Coarctation was native in eight patients and recurrent in three patients. The BeGraft Aortic stent was successfully implanted in all patients (12/29 mm (stent diameter/the length of the stent) in 5 patients, 14/29 mm in 3 patients, 12/39 mm in 2 patients, 14/39 mm in 1 patient). In five patients, after the stents were opened completely by the first balloon, they were exchanged with a balloon, 1-3 mm larger in diameter and the stents were redilated. The median catheter derived systolic peak-to-peak pressure gradient was 23 mmHg (IQR 16–37 mmHg) before the procedure and 3 mmHg (IQR 1–5 mmHg) after the procedure. The median procedure and fluoroscopic time was 60 min (IQR 50-67 min) and 18 min (IQR 15-25 min) respectively. The median follow-up duration was 5 months (IQR 2-12 months). During follow-up, only one patient (9%) had stent narrowing that required dilation.
Conclusion: According to our results the BeGraft aortic stent is considered to be safe and effective in the treatment of CoA from childhood to young adult and short term follow up, however more experience and medium- to long-term follow-up are required.

Keywords: aortic coarctation, children, congenital heart disease, covered stent, young adults

Table 1. The details of 11 patients treated with BeGraft aortic stent

Patient No	Gender	Age (years)	Weight (kg)	Implantation site	Stent diameter/ length (mm)	Sheath size (French)	Peak-to-peak gradient (mmHg)		Coarctation diameter (mm)	
							Pre	Post	Pre	Post
1	Male	12	43	Native CoA	14/39	12	16	1	9	14.5
2	Male	13.7	53	Recurrent CoA (post-balloon angioplasty)	14/29	12	15	0	8.5	14
3	Male	11.3	35	Native CoA	12/29	10	37	5	5.5	13
4	Male	15	34	Recurrent CoA (post-op)	14/29	12	20	0	8	13.5
5	Male	16	96	Native CoA	12/39	10	23	3	6.5	10
6	Male	11	39	Native CoA	12/29	10	35	7	2.5	10
7*	Male	22	68	Recurrent CoA (post-stenting)	14/29	12	15	8	11	14
8	Male	13	37	Native CoA	12/29	9	30	2	2	11
9	Male	17.9	62	Native CoA	12/39	10	38	2	5.0	13.5
10	Female	12.7	35	Native CoA	12/29	9	22	3	3.5	10.5
11	Male	15	53	Native CoA	12/29	10	42	5	3.5	11.5
Median		13.7	43				23	3	5.5	13

* = Stent narrowing during follow-up

P-327

The results of cardiac catheterization low birth weight infants less than 2.5 kg with acute and mid-term outcomes

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Background and Aim: Due to the increased survival rate of low birth weight infants with congenital heart disease, the incidence of cardiac catheterization intervention in these infants is increasing. Limited data are available on cardiac catheterization experience in this group. With this study, we aimed to share our 2-year experience in cardiac catheterization cases in low birth weight infants in a single center.

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Methods: Cardiac catheterizations performed on infants weighing <2500 g between January 2020 and December 2021 were retrospectively reviewed. Demographic data, procedure, outcomes, and follow-up data were collected.
Results: Total 53 cardiac catheterizations performed on 48 patients. Interventional procedures were performed in 46 and diagnostic catheterizations in 7 infants. The mean age at catheterization was 21.2 days (range 1 to 60 days) and the average weight 1865,6 g (range 870 to 2500 g). 34 (%73,9) of the interventional procedures were successful, 4 were partially successful and 8 were unsuccessful. Premature PDA closure (n = 16), balloon coarctation angioplasty (14), pulmonary balloon valvuloplasty/angioplasty (8), stenting of the ductus arteriosus (7), RVOT stenting (3), pulmonary valve perforation (2), balloon atrial septostomy (2) were performed. There were also cases when two procedures were performed at the same time. Complications during the procedure; arrhythmia was observed in 5 patients, stent/device embolization in 6 patients, and cardiac arrest in 1 patient. One patient died during the procedure due to AV block and hemodynamic disorder developed during the procedure. Two patients died on the day of the procedure due to post-procedure hemodynamic instability. Doppler USG was performed in only 8 patients due to the suspicion of thrombosis and thrombosis was detected in 6 of them. Re-intervention was required in 9 patients and 2 of them were operated during the follow-up. 15 patients required surgery in the post-procedure follow-up. In the mid-term follow-up, 5 of the patients died of cardiac origin and 8 of them due to secondary reasons.
Conclusion: Surgical intervention is associated with high mortality and morbidity in infants <2500 g. Therefore, although there are complications; interventional catheterization is an important procedure as definitive therapy or palliative therapy in low birth weight infants.

Keywords: congenital heart disease; cardiac catheterization; low birth weight; <2.5 kg; complications

Surgery and Intensive Care

P-329
Coagulation profile of single ventricle patients prior to first cardiac intervention in comparison to healthy controls

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Background and Aim: Thrombotic and thromboembolic events remain challenging complications in single ventricle (SV) patients. A high frequency of clotting factor abnormalities leading to a prothrombotic state seems to play an important role. Whether the coagulation imbalance precedes unphysiological circulation or is a consequence of univentricular palliation, as well as the clinical impact is still a matter of debate.
Methods: This is a multicentre, prospective, longitudinal, controlled cohort study with consecutive inclusion of 29 SV patients and 24 healthy controls (HC). Blood samples for coagulation profile were taken prior to any surgical or interventional procedure. The median (IQR) age at blood sample collection was 5.5 (1.75-11.25) days in the SV group and 7.8 (2.95-23.1) days ($p = 0.22$) in the HC with a non-significant male predominance in both groups ($p = 0.09$).
Results: SV patients had significantly reduced levels of protein C (PC) ($p<0.001$), free-protein-S (PS) ($p<0.001$), and antithrombin (AT) ($p<0.001$) compared to HC. Coagulation factor (F) II ($p = 0.03$), V ($p<0.001$), VII ($p<0.001$), X ($p = 0.03$), XI ($p = 0.004$), and fibrinogen ($p<0.001$) as well as plasminogen ($p<0.001$) were also reduced in SV patients. Von Willebrand factor (vWF) and FVIII were similar within the groups. Patients with low AT ($p = 0.03$) or low PS ($p = 0.02$) had a longer ICU-stay. Thromboembolic events including stroke and ischemic colitis were associated with lower PS ($p = 0.05$), AT ($p = 0.007$), and FV ($p = 0.02$). Incidence of shunt thrombosis was increased in patients with low PC ($p = 0.03$), but incidence of peripheral vessel thrombosis did not correlate with coagulation inhibitors or FV. No patients showed relevant increased bleeding prior/after surgery.
Conclusion: Abnormalities in coagulation profile are present directly after birth prior to any intervention in SV patients. These abnormalities do not seem to be caused by inflammatory processes, as acute phase proteins (vWF and FVIII) are normally distributed between the two groups. Significantly decreased FV suggest that coagulation abnormalities are likely due to decreased liver synthetic capacity. The disbalance of coagulation parameters is associated with increased ischemic thrombotic risk, shunt thrombosis and longer ICU-stay.

Keywords: Coagulation, thromboembolic events, single ventricle

P-330
Reverse differential and differential cyanosis in a TGA newborn

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Background and Aim: Reverse differential cyanosis is well recognized in transposition of great arteries(TGA) with persistence pulmonary hypertension of the newborn (PPHN). With critical and invasive management supported by extracorporeal membrane oxygenation (ECMO), the hemodynamic changes in TGA-PPHN reversed to differential cyanosis.
Methods: A term male newborn was detected clinical cyanosis at 1 hour after delivery. His pre-ductal and post- ductal saturation was 88% and 94% while he was breathing in room air. He became hypotensive during echocardiogram evaluation. He was referred to our institute due to TGA with severe PPHN. Upon our NICU arrival, the infant was cardiac arrested which required resuscitation. The infant was intensively supported by high dosage of inotropic medication, fluid-electrolyte adjustment, pulmonary vasodilators and high frequency ventilators. His condition was transiently stabilized but severe hypoxia and multi-organ impairment was progressed. Decision was made to activate veno-arterial ECMO support to bypass heart and lung during grievous period.
Results: During the ECMO support, the clinical cyanosis was improved but there was a significant differential cyanosis with pre-ductal and post- ductal saturation was 95% and 75%. Hemodynamic explanation confirmed by echocardiogram as Figure. With ECMO support along with intense pulmonary vasodilators, PPHN was improved with diminished difference between the pre-ductal and post-ductal saturation. The ECMO was decanulated on day 12.
Conclusion: Significant hemodynamic reversal flow could be observed in TGA-PPHN while the infant being support on

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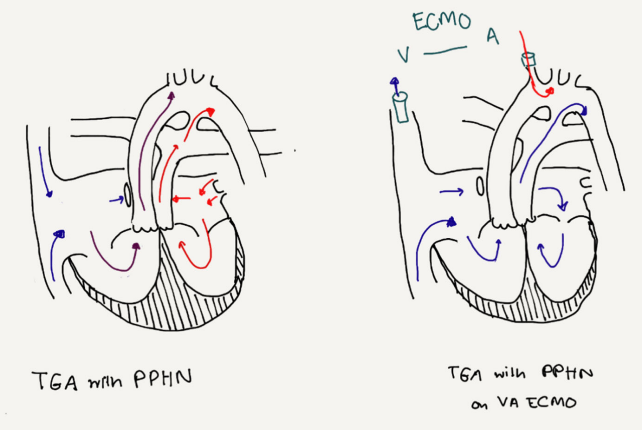
ECMO. Understanding the hemodynamic changes would be benefit for management and objective key result.

Keywords: Transposition of great arteries, Persistent pulmonary hypertension of the newborn, reverse difference cyanosis, difference cyanosis, ECMO

Reverse differential vs differential cyanosis



P-331

Neo-pulmonary valve creation technique using right atrial appendage

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Background and Aim: The use of the right atrial appendage (RAA) as a neo-pulmonary valve (PV) in the repair of Tetralogy of Fallot (TOF) has been used as a new and unique treatment option in our clinic since 2019 in suitable patients. We also have expanded the use of this method to other pathologies that require a neo-pulmonary valve. Herein we want to share our increased experience.
Methods: From October 2019 to October 2021, neo-pulmonary valve with RAA applied to 31 patients. 27 patients with TOF, TOF with pulmonary atresia and TOF with the absence of Pulmonary valve had an RAA-PV replacement, the other four patients had a right ventricular outflow tract reconstruction with transannular patch for other anomalies. All patients underwent postoperative echocardiographic examination on discharge and at the 6th month.
Results: The postoperative course was uneventful for most of the patients only one patient required mechanical support (ECMO) on the postoperative third day due to pulmonary reperfusion injury and this patient weaned successfully and was discharged on postoperative day 34. No mortality was detected. Median ventilation time, intensive care unit stay and the length of hospital stay were 8 hours (2-250), 1 day (1-25), and 7 days (4-34), respectively. Longer perfusion time was not correlated with postoperative times. At 6 months echocardiography showed none/trivial PI in 26 patients and mild PI in 4 patients, mild to severe in 1 patient.
Conclusion: Early results showed that neo-pulmonary valve reconstruction using the RAA tissue may provide a reasonable alternative for RVOT reconstruction with the transannular patch But long-term results are needed.

Keywords: Neo-pulmonary valve; pulmonary valve reconstruction; right atrial appendage.

P-332

Surgical results for aortic coarctation in children

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Background and Aim: Isolated aortic coarctation repair (AC) is performed by left thoracotomy with resection and extended end-to-end anastomosis (REEA) technique with low mortality and morbidity rates. Long-term re-coarctation and late hypertension are the most important complications. In this study, we aimed to present the surgical treatment results of children who underwent surgery with LT due to isolated AC in our clinic.
Methods: 90 consecutive patients who underwent isolated coarctation repair in our clinic between 2011 and 2021 were retrospectively reviewed. The preoperative characteristics, operative data, and postoperative early and long-term results of the patients were analyzed.
Results: Thirty (33.3%) of the patients were newborns, 46 (51.1%) were infants, and 14 (15.5%) were children. Extended end-to-end anastomosis was applied to all patients. Pulmonary artery band was applied to 3 (3.3%) patients simultaneously, and aberrant right subclavian artery division was applied to 1 (1.1%) patient. The mean cross-clamp time was 29.13 ± 6.97 minutes. Early reoperation was required in 2 (2.2%) patients. In the early period, mortality was observed in only 1 (1.1%) patient. Recoarctation developed in 8 (8.8%) patients, reoperation was performed in 4 (4.4%) patients and reintervention was performed in 4 (4.4%) patients. 22 (26.8%) patients are being followed up under antihypertensive treatment due to hypertension. Average follow-up time of the patients 41,3±22,8 was the months. There was no mortality in the late period.
Conclusion: Isolated coarctation is successfully treated with the left thoracotomy REEA technique with l
ow mortality, morbidity and low long-term re-coarctation rates. Preoperative surgical planning and approach are important in these patients. Treatment of patients with aortic arch hypoplasia and additional anomalies with median sternotomy reduces long-term interventions.

Keywords: Aortic coarctation, Recoarctation, Congenital heart disease, surgery

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P-333

Coronary artery stent implantation in an infant with williams syndrome on ECMO support

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Background and Aim: Percutaneous coronary artery stenting is rarely performed in pediatric population and considered for rescue intervention in selected cases. Williams Syndrome (WS) is one of a kind that may be complicated by its coronary artery involvement throughout the course of the arteriopathy. We report a rare case of an infant with WS who successfully had percutaneous left main coronary artery (LMCA) stenting under extracorporeal membrane oxygenation (ECMO) support following extended peripheral pulmonary arterioplasty surgery.
Methods: A 6 month-old male patient with diagnosis of WS related severe peripheral pulmonary stenosis had extended pulmonary artery reconstruction at our institution. During surgery, ischemic parts on the left ventricular myocardial surfaces were noted and the surgical team had difficulty weaning the patient from circulatory support. Catheterization of the coronary arteries was urgently performed on the postoperative first day. Angiography confirmed severe LMCA stenosis with diminished distal perfusion and diffuse narrowing of supraaortic region through its course to ascending and descending aorta. A 2.5x9 mm bare-metal stent was deployed in LMCA proximal to its bifurcation. Stent dilation was achieved by utilizing 1mm and 1.5 mm coronary balloons. Control angiography showed improved LMCA calibration and restoration of sufficient antegrade flow. Heparin infusion was administered for the next 24 hours before maintanence of thromboprophylaxis with aspirin and clopidogrel on the next day. The patient was decannulated from ECMO support 7 days following the stent implantation. Remarkably elevated cardiac troponin levels were normalized eventually with appropriate ECG changes and residual mild ventricular systolic disfunction.
Results: Coronary artery involvement should be a particular concern in patients with Williams Syndrome, which may manifest as ostial/diffuse stenosis, dilation of coronary arteries or obstruction to coronary artery inflow by the aortic valve, the sinotubular ridge, or a combination of both.
Conclusion: Percutaneous coronary artery stenting is a relatively safe option in infants and young children with coronary artery stenosis in urgent situations. These procedures are technically performed with a low complication rate. Readily available ECMO support throughout the procedure is advantageous for infants exhibiting symptomatic heart failure. Aggressive thromboprophylaxis with aspirin and clopidogrel should be initiated and maintained to delay the risk of stent thrombosis.

Keywords: coronary artery stenosis, stent implantation, Williams syndrome, extracorporeal membrane oxygenation

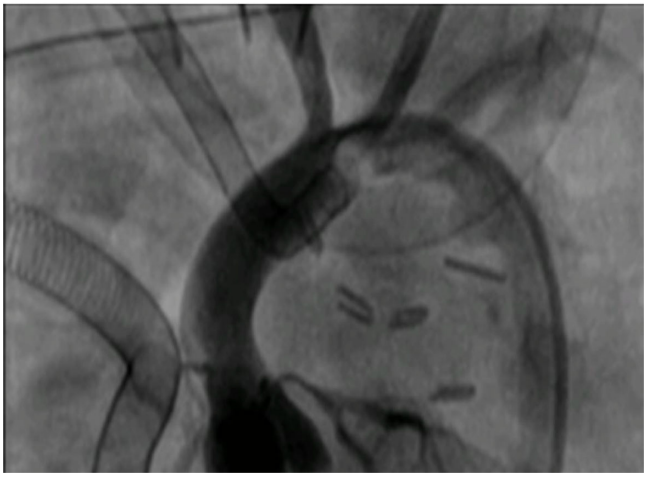

Figure (a) Aortic root angiogram shows diminished caliper of LMCA (b) Repeat angiography after placement of 2.5 x 9 mm coronary stent

P-334

Percutaneous and surgical pulmonary valve replacements results

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Background and Aim: Pulmonary valve replacement (PVR) is especially necessary in long-term follow-up of TOF patients, but also in congenital heart surgery for non-TOF reasons. In our study, we aimed to present the results of percutaneous and surgical PVR performed in our clinic between the years 2016-2021 in a comparative manner.
Methods: The data and statistics of our patients were obtained using the database of our hospital and were evaluated retrospectively.
Results: During the period included in the study, a total of 155 patients underwent pulmonary valve replacement. 127 of these patients are from the group that underwent PVR percutaneously and 28 of them surgically. Of the patients who underwent percutaneous PVR, 72 were male and 55 were female; of those who underwent surgery, 17 were male and 11 were female. The mean age of the percutaneously administered patients was 18.5 (5.8-63), and the mean weight was 55,2 (17-100). Those who underwent

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surgical PVR were 12.5 (0-37), 33.5 (1-84), respectively. Of the patients who underwent percutaneous PVR, 100 (78%) were previously operated for TOF, and the other patients were pulmonary stenosis (n = 12), pulmonary atresia (n = 6), TGA (n = 4), post-Ross procedure (n = 3) and truncus arteriosus (n = 2). The proportion of patients who underwent surgery for reasons other than TOF was 25%, and VSD-PA (n = 2), ASD-PS (n = 1), c-TGA (n = 1), DIRV-DORV (n = 1), Ebstein Anomaly (n) = 1) and one patient was operated for endocarditis. There were no early or follow-up deaths in either group. Biological valve was used in all surgical and percutaneus PVRs, and conduit valve was replaced for 4 patients due to size mismatch. In addition, cryoablation was applied to two patients.

Conclusion: The most important reason why the number of percutaneous valve replacements is significantly higher than the number of surgical replacements is that the priority of our center is the percutaneous approach. Surgical method is preferred for reasons such as residual cardiac defect, arrhythmia, tricuspid valve insufficiency, pulmonary artery stenosis, pulmonary annulus size mismatch, any complications that may develop in the percutaneous approach. Although our early results are satisfactory, it would be more appropriate to evaluate our patients with their long-term results.

Keywords: Pulmonary valve replacement, Percutaneous pulmonary valve implantation, Congenital heart disease

P-335
Ozaki procedure in pediatric congenital aortic valve pathologies

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Background and Aim: Surgical treatment for pediatric aortic valves (AV) pathologies is still controversial. The Ozaki procedure is defined as a successful technique with its results in recent years. We evaluated the early results in pediatric patients who underwent the Ozaki procedure in our clinic.

Methods: Between June 2019 and August 2021, 22 patients underwent the Ozaki procedure. The mean age was 13.3 years (1.5-18), and the mean body weight was 53.2 kg (9-90 kg).13 patients had significant aortic insufficiency, 7 patients had significant aortic stenosis, 1 patient had AV endocarditis, 1 patient had Shone complex. The preoperative mean aortic annulus of the patients was 21.6 mm (8.5-26). The aortic valve was unicuspid in 6 patients, bicuspid in 11 patients, and tricuspid in 4 patients. Single leaflet Ozaki technique was implanted in 3 patients. Standard Ozaki procedure was applied to 19 patients. The mean Cardiopulmonary bypass time was 153.6 minutes, and the cross-clamp time was 135.6 minutes.

Results: The mean ICU stay was 2.8 days, and the hospital stay was 8.5 days. Reintubation and inotropic support was required in one patient due to low cardiac output on postoperative day 0, tachycardia and low CF developed in one patient on postoperative day 7, and a patient who was operated due to Shone complex underwent permanent pacemaker implantation due to postoperative AV complete block. As an additional intervention, mitral repair in 4 patients, tricuspid repair in 1 patient, LVOTO resection in 2 patients, CRT implantation in 1 patient, and ascending aorta replacement in 1 patient were performed. No mortality was observed. Three patients required reoperation, 1 patient developed advanced AI at 1 month postoperatively, and 3 developed AI after 1 year postoperatively. Two patient had reoperation due to leaflet detachment and patient had aortic valve replacement.

Conclusion: The early results of AV neocuspidization performed with the Ozaki method are satisfactory. The stability of the glutaraldehyde-treated autologous pericardium at the aortic leaflet location should be monitored closely. The results of the method should be followed up with larger series in the medium and long term.

Keywords: Ozaki procedure, Aortic valve neocuspidization, Aortic valve

P-336
Predictors of prolonged pleural effusion after fontan operation

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Background and Aim: Even though mortality after Fontan procedure has improved, non-physiological flow dynamics may cause significant morbidity such as prolonged pleural effusion(PPE), the most common postoperative complication. This study aims to enlighten the factors affecting PPE after Fontan operation.

Methods: The medical data of 69 patients, who underwent Fontan operation between June 2018- December 2020 and still alive, was reviewed. Baseline characteristics, preoperative clinical and hemodynamic data from cardiac catheterization, operative/postoperative variables of patients were analyzed.PPE was defined as chest tube drainage lasting more than 7 days (group 1: PPE -, group 2: PPE +). Among 69 patients, 40 were female (58%). The most frequent diagnosis was tricuspid atresia (n: 13, 19%). Twenty-eight (40%) patients had PPE, 11 (16%) had effusion lasting longer than 14 days. Ten patients with PPE (35%) had pulmonary atresia coexistent with the primary diagnosis. The left ventricle was dominant in 39 (56%) patients. AV valve regurgitation was mild in 40% of the patients. The median age at Fontan operation was 4.9 years (2.5-26). The median mean pulmonary artery pressure (mPAP) was 11 mmHg (range 6-19). Extracardiac Fontan procedure was applied to all except seven patients who underwent intra-extracardiac conduit. Fenestration was performed in 6 (8%) patients, three of whom had PPE postoperatively. Fontan surgery was performed in 6 patients (8.7%) over the age of 10. Four of them had PPE, while in 2 patients, PPE lasted over 14 days. The median follow-up time was 1.2 years (0-2.2). Preoperative median mPAP (11 mmHg vs 12 mmHg, p:0.015), postoperative early (0-3 days) CRP (40 gr/L vs 70 gr/L, p:0.04), late (3 days-discharge) albumin (3.4 gr/dL vs 3.1 gr/dL, p:0.014) and GGT levels (27 IU/L vs 33 IU/L, p:0.04) showed significant differences in PPE+ group.

Results: Etiology of PPE after Fontan procedure is obscure and likely multifactorial. According to our study, tricuspid atresia is less likely to be a risk factor for PPE, whereas pulmonary atresia and older age at Fontan operation were predictors of PPE though this data is not statistically proven due to limited number of patients.

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Conclusion: Preoperative risk-stratifying and implementing consistent postoperative care targeting the unique physiology of Fontan circulation are crucial. Preoperative PAP and elevated early postoperative CRP levels may be good predictors for PPE.

Keywords: effusion, Fontan, prolonged, single ventricle

P-337

Late atrial thrombus formation after surgical closure of atrial septal defect- a case series

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Background and Aim: Ostium secundum atrial septal defect (ASD) is considered a simple heart defect that is now 'easy to treat', either by transcatheter intervention or by surgical approach, with a very low rate of complications. Although rare, thrombotic complications due to right atrial thrombus formation are reported in the literature. We report 3 cases of late thrombus formation in the right atrium after surgical closure of ostium secundum ASD.
Methods: All of these cases are children operated over a period of 17 years (from 2004 until 2021). The three patients received the surgical correction between the age of 2.5 and 5 years. Two of the children are boys and they both have Down syndrome. They underwent surgical correction of large ASDs with bovine pericardial patches. The postoperative course was uneventful.
In all of the cases the thrombus was an incidental late finding on a routine check-up more than one year after the surgical correction. The children were asymptomatic and the thrombus was detected upon regular transthoracic echocardiography. It was a medium size, hyperechoic mass, attached to the free wall of the right atrium and did not cause obstruction of the systemic venous return.
Results: All the children had a negative thrombophilia screening and anticoagulation therapy was started upon the diagnosis. In two of the cases the thrombus was surgically removed because the echocardiographic aspect was considered at high risk of embolization and the pathology report confirmed the clinical diagnosis. Oral anticoagulant therapy was administered for the first 6 months following surgery. The third child, whose mass was sessile, received a conservative approach with 6 months of anticoagulation therapy followed by 2 years of antiplatelet treatment without significant modification of the dimensions of the atrial mass.
All the children are doing well and no other thrombotic event was detected until now.
Conclusion: Due to the small number of cases reported in the literature, there is no consensus on the management and prevention of such complication, taking into consideration the risks and benefits of surgical versus conservative therapies. In our cases, the indication of surgery was based on the echocardiographic characteristics of the mass.

Keywords: right atrial thrombus, surgical closure of ASD, complication

P-338

Management and outcomes in partial anomalous pulmonary venous drainage (papvd) in paediatric patients: a single tertiary centre's experience

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Background and Aim: Partial Anomalous Pulmonary Venous Drainage (PAPVD) presents with variable anatomical patterns involving drainage of one or more pulmonary veins directly into the Right Atrium (RA) or via one of its systemic venous connections rather than into the Left Atrium (LA). It may be associated with other Congenital Heart Disease (CHD) or can be an isolated anomaly. The resulting increase in the pre-tricuspid left to right shunt can lead to Right Ventricular Volume Overload (RVVO) and increased flow to the lungs. Surgical repair involves redirecting pulmonary venous flow to the LA directly or via a baffle. We aim to review management and outcomes of PAPVD in paediatric patients at a single centre.
Methods: Our database was searched for patients with a diagnosis of 'PAPVD', 'sinus venosus atrial septal defect', 'scimitar' and 'dextroposition' between 1st Jan 2002- 1st Jan 2021. Patients between 0-18 years were included for analysis. Patients with total anomalous pulmonary venous drainage and uni-ventricular circulations were excluded. Anatomy of PAPVD and diagnostic modality were documented. Primary outcome was conservative or surgical management including indications for each. Secondary outcomes were mortality, complications and symptomatic disease.
Results: A total of 36 patients were included. The median age was 109.5 months (IQR 58-155 months). 4/36 (11%) had isolated PAPVD and others (89%) had associated CHD. An Atrial Septal Defect was present in 22/36 (61%) of which the most common type was a superior sinus venosus defect 11/22 (50%). Indications for surgery included presence of RVVO, pulmonary hypertension (PH), multiple vein involvement or established increased QP:QS ratio. 24/36 (67%) patients underwent surgical repair. Within the operated group, there were no deaths and no immediate complications requiring return to theatre. On follow up, obstruction to the pathways were noted in 4/24 (17%). 1 child required re-intervention by cardiac catheter at 3 months post-surgery to address stenosis of the baffle. 3/36 children (8%) died but all of them were in the non-operated group.
Conclusion: There is no agreed consensus for when and in whom to intervene for paediatric PAPVD however locally, patients with a clear indication undergo surgical repair. Surgery has a low risk of complications and mortality.

Keywords: Paediatric PAPVD, Scimitar, PAPVD surgery

P-339

Event-free interval of 50 years after surgery for congenital pulmonary stenosis - was dacron the key to success?

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Background and Aim: Two patients who underwent surgery for pulmonary stenosis in childhood attracted attention by similar disease causes (Tetralogy of Fallot and Alagille Syndrome) and an event-free interval of 50 years. Therefore we examined the synthetic material (Dacron) implanted in these patients histologically to assess its performance.
Methods: Specimen were obtained during re-operation (pulmonary valve replacement). After histologic preparation, tissue analysis was carried out in a standardized fashion. Patient history was examined in depth.
Results: Tissue analysis revealed variable thickening and massive, cockade-like and coarse calcifications, excessive fat depositions, swirls and irregular fibers of collagens and ossifications. Characteristic fibrous structures causing double refraction in polarization led to the identification of formerly fibrous synthetic material. An extensive inflammatory response was present. With

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no apparent vascular wall ultrastructure, fibrotic structures in various layers could be identified on the luminal side. An extensive shrinkage of the native pulmonary valves was observed.
Conclusion: Severe transformations of the surrounding prosthetic material led to dysfunction in the long-run. From the immunological point of view, Dacron was not completely inert but even triggered extensive destructive and reconstructive processes. Pulmonary stenosis corrected early in childhood (with the technique of 1967 and implantation of Dacron) had a good long-term prognosis in our two cases.

Keywords: congenital heart surgery, history, histology, pathology

P-342

Medium-term outcomes and risk factors in single ventricle congenital heart disease - systemic left versus systemic right ventricle

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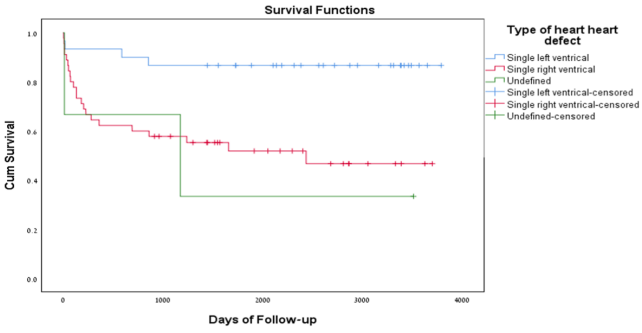
Background and Aim: Single ventricle congenital heart defects (SV-CHD) are the most severe type of congenital cardiac anomalies. Substantial developments in surgical and medical treatments over recent decades improved survival rates amongst SV-CHD patients. The aim of this study was to analyze current survival rates of patients with SV-CHD and compare the outcomes for single left ventricle (LV) versus single right ventricle (RV). In addition, we examined whether lactate level prior to and during first cardiac surgery could serve as a predictor of survival following first cardiac surgery.
Methods: We conducted a retrospective observational study based on patients' surgical and medical records. We included all infants diagnosed with SV-CHD who had their first cardiac surgery at our hospital before one year of age, between the years 2011-2018. The hospital's Ethics Committee approved the study protocol.
Results: 78 patients (43 males, 55%) met inclusion criteria. 45 (57.7%) had single RV. 30 (38.4%) had single LV and 3 (3.7%) had two ventricles not amenable for surgical septation. Median follow up was 6.95 years. Overall mortality during follow up was 36%. Survival rates were significantly higher for patients with single LV compared to patients with single RV (86.7% versus 51.1% $p = 0.002$, Fig 1).
Higher lactate level prior to first cardiac surgery significantly correlated with higher mortality ($p = 0.011$). Higher maximal lactate level during first cardiac surgery significantly correlated with higher mortality ($p = 0.001$). Utilizing receiver operating characteristic (ROC) curve statistical analysis we found lactate cutoff level of 7.05 mmol/l during first cardiac surgery as a predictor of mortality (specificity of 61.8%, sensitivity of 81.8%). Among demographic characteristics, we found statistically significance correlation between higher gestational age at birth, higher birth weight and shorter duration of the first operation with higher survival rates ($p = 0.04$, 0.01 and 0.005, respectively).
Conclusion: Patients with a single RV heart defect have significantly lower survival rates compared to patients with a single LV. The data may assist health care providers and patients' families during prenatal counseling. Higher lactate levels prior to and during the first cardiac surgery significantly correlate with higher post-surgical and overall mortality rates.

Keywords: Congenital heart defect, Hypoplastic left ventricle. Hypoplastic right ventricle, Lactate

Figure 1: Survival



P-343

Effects of cardiac surgery on anthropometric growth measures of children with down syndrome and failure to thrive. observational control study

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Background and Aim: to assess the effect of cardiac surgery on growth catch-up of DS children with failure to thrive and CHD and investigate other causes of FTT in DS children.
Methods: We conducted a retrospective observational study in tertiary cardiac center from 2015 to 2018. We included all cases of DS diagnosed with CHD and FTT who completed a one-year follow-up after cardiac surgery. We divided the cases into two groups; "control group" includes children who normalized their growth parameters and "underweight group" includes those who remained in FTT category during the follow up period. We compared both groups for multiple risk factors
Results: most of DS had FTT upon surgery. 50% of cases completed one-year follow-up including 29 (60%) in the control group and 19 (40%) in underweight group. Within 6 months post-surgery, the control group though did not reach yet normalization of growth parameters, demonstrated statically significant improvement in weight for age, weight Z-score in compared to underweight group. Within 12 months post-surgery, the control group achieved normalization of growth parameters and continue to show more statistically significant differences in growth parameters. Both groups had comparable post operation course. Univariate analysis of possible peri-operative risk factors showed no difference between both groups except for presence of untreated subclinical hypothyroidism in 58% of the underweight group versus 17% in control group ($p = 0.005$)
Conclusion: FTT in DS patient is multifactorial which needs thorough investigation and work up by multidisciplinary team. Cardiac surgery may not guarantee the improvement of growth parameters.

Keywords: Down Syndrome, post cardiac surgery, Failure to thrive, Growth velocity

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P-344
First-in-man use of the EXCOR® venous cannula for combined cavopulmonary and systemic ventricular support in fontan circulation failure

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Background and Aim: The Berlin Heart EXCOR® system has been developed for mechanical circulatory support (MCS) of paediatric patients with terminal heart failure. A recently introduced iteration of the system (EXCOR® Venous Cannula, Berlin Heart GmbH, Berlin, Germany) is dedicated to support patients with univentricular physiologies by facilitating implantation of the EXCOR® device into the Fontan pathway. We report the first-in-man use of this system for combined cavopulmonary and systemic ventricular support in a patient with Fontan circulation failure.
Methods: A 12-year-old boy (140cm, 42.7kg, BSA 1.29m2, Intermacs 2) with hypoplastic left heart syndrome developed therapy refractory systemic ventricular dysfunction and Fontan circulatory failure after surgery for aneurysmal degeneration of the DKS anastomosis. Given the critical hemodynamic situation, MCS of the systemic ventricle and cavopulmonary pathway was indicated. Surgery comprised of standard Berlin Heart EXCOR® implantation to support the failing ventricle (12mm apex/staged 12/9 mm arterial cannula/50ml ventricle) and implantation of the novel Fontan inflow cannula to support the cavopulmonary circulation. The latter was achieved by performing a TCPC takedown with subsequent anastomosis of a staged 12/9 mm outflow cannula to the pulmonary artery and implantation of a 14/18mm EXCOR® Venous Cannula as subpulmonary inflow graft, which was connected to the superior vena cava and Fontan tunnel using GORE-TEX® grafts.
Results: Implantation of two EXCOR® VAD systems utilizing the novel Fontan cannula and postoperative recovery were uneventful. End-organ functions continuously improved to age- and condition-specific norm values at one-month post-implant. At five weeks post-implant, the patient is gradually mobilized on the EXCOR® Active Unit (Berlin Heart GmbH, Berlin, Germany), and transplant listing is pending.
Conclusion: The Berlin Heart EXCOR® Venous Cannula is the first dedicated system for mechanical support of Fontan circulatory failure. In our patient, combined subpulmonary and subaortic support normalized hemodynamics and reversed end-organ dysfunction.

Keywords: congenital heart surgery, mechanical circulatory support, Fontan, single ventricle, ventricular assist device

P-346
Outcome of late complete repair of tetralogy of fallot: A 13-year single-center experience

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Background and Aim: Currently, the surgical strategy of Tetralogy of Fallot (ToF) is a one-time surgical complete repair performed during the first months of life. Pulmonary valve-sparing repair (PVSR) is promoted for the best long-term result, with a pulmonary annulus Z-score cut-off value around -3 in infants reported in the literature. For many years, late surgical repair of ToF has been performed at our institution for children coming from emerging countries. We aimed to describe the preoperative anatomy in this population and analyze the outcomes and feasibility of PVSR.
Methods: Single-center retrospective observational study of humanitarian patients, older than one year, with a confirmed diagnosis of ToF and complete surgical repair between 2005 and 2018 at our institution.
Results: One hundred sixty-one patients met the inclusion criteria. The median age at repair was 4.5 [3.0 – 6.3] years with a sex ratio (M/F) of 1.75/1. Median preoperative peripheral pulse oximetry was 78 [70 - 85] %, hemoglobin 163 [142 - 190] g/L and hematocrit 50.5 [44.0 - 59.1] %. A modified Blalock-Taussig shunt was performed in 11.2% before complete surgical repair. Preoperative echocardiographic measurements showed the most relevant hypoplasia at the level of the main pulmonary artery and the pulmonary valve annulus level (Z-score of -3.41 [-4.93 - -2.00] and -1.84 [-3.42 - -0.85], respectively). The median CPB time was 125 [105 - 151] min with a median aortic cross-clamp of 76 [62 - 91] min. The PVSR was possible in 46.6% of patients with significant higher Z-score in the PVSR-group but without identifiable cut-off value. The median duration of invasive ventilation was 26.1 [21.3 – 71.7] hours after CPB weaning. The median ICU stay was 5.9 [4.7 - 7.9] days. There was no death. The surgical reintervention rate at 30 days was 14.9%, mainly due to pulmonary valve regurgitation.
Conclusion: Late complete surgical repair of ToF presents low short-term mortality and morbidity. PVSR is linked to pulmonary valve annulus size but not only, unlike in infants. Preservation of the pulmonary valve seems to be impacted by other parameters than z-scores of the pulmonary valve annulus.

Keywords: Tetralogy of Fallot, late repair, pulmonary valve-sparing repair, outcome

P-347
Prolonged mechanical ventilation after congenital heart surgery in pediatric patients: do we need an improved predictive model?

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Background and Aim: Prolonged mechanical ventilation (PMV) is a well-recognized factor as a quality metric for pediatric cardiac surgical programs. Most of the risk factors for PMV are described and analyzed. In the last years some of these factors are combined in specific scores. Even more, some authors had established predictive models to detect proactively patients in risk for PMV. This study aims to develop a new predictive model, based on vasoactive-ventilation-renal (VVR) score, for PMV after congenital heart surgery (CHS) in pediatric patients.
Methods: Medical files of patients 0-18y who underwent heart surgery in 2016 and 2017 were reviewed. Patients that met the

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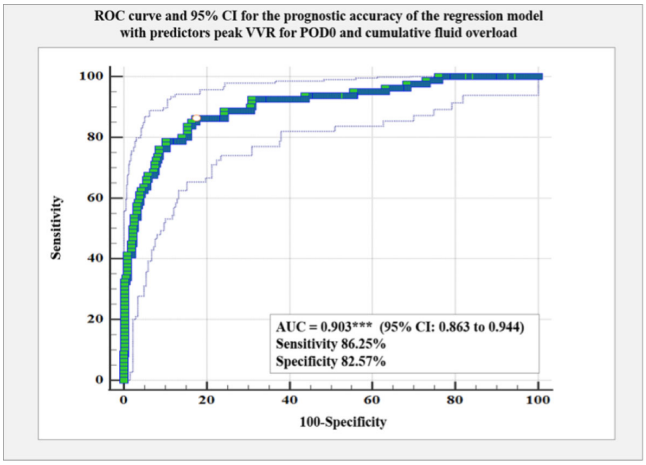
inclusion criteria were studied. PMV was defined as invasive mechanical ventilation ≥ 96h. The patients were divided in two groups according to duration of mechanical ventilation: group 1- patients with PMV, group 2 - patients without PMV. The focus was on VVR score and fluid overload in the first 48 hours after the operation. Data were presented as medians with IQR or as means ± standard deviation. A non-parametric Mann-Whitney U test, binary logistic regression test and ROC curve analysis integrated in the statistical software SPSS 24.0 were used. A value of $P < 0.05$ was considered significant.
Results: 438 patients were operated in 2016 and 2017 and 384 of them were included in the study. 80 patients (20.8%) needed PMV (group 1) and 304 (79.2%) did not need PMV (group 2). There was a statistical significance between group 1 and group 2 concerning the peak VVR for the day of operation 58,25 (33,48) vs. 25,65 (19,8) and cumulative fluid overload in % for the first 48hours +2,54(13,29) vs. – 1,19(3,4). After combining this two factors in a predictive model, the ROC curve analysis showed AUC 0,903 (95% CI 0,863-0,944) with sensitivity of 86.25% and specificity of 82,57%.
Conclusion: Combining VVR and cumulative fluid overload for the first 48 hours resulted in establishment of a new reliable predictive model for PMV after CHS in pediatric patients in our center.

Keywords: prolonged ventilation, VVR score, fluid overload, congenital heart surgery, children



P-348

Retrospective analysis regarding outcome of patients with tetralogy of fallot with pulmonary atresia with or without major aorto-pulmonary collaterals

Eva Sames Dolzer, Anna Fahrnberger, Michaela Kreuzer, Roland Mair, Gregor Gierlinger, Roland Gitter, Christoph Prandstetter, Andreas Tulzer, Gerald Tulzer, Rudolf Mair; Children's Heart Center Linz, Kepler University Hospital, Linz, Austria

Background and Aim: Tetralogy of Fallot patients with pulmonary atresia have a largely varying source of pulmonary perfusion with often hypoplastic or absent central pulmonary arteries. Therefore single stage full correction is not feasible in all patients. A retrospective single center study was undertaken to assess outcome of these patients regarding type of surgical procedures, long term mortality, achievement of VSD closure and analysis of postoperative interventions.
Methods: 76 patients with TOFPA operated between 1.1.2003 and 31.12.2019 are included in the study. Patients with ductus dependent pulmonary circulation underwent primary single stage full correction including VSD closure and RVPA conduit implantation or transannular patch reconstruction. Patients with hypoplastic pulmonary arteries and MAPCAs without double supply were treated by a unifocalization and RVPAC implantation. The follow up period ranges between 0 and 16,5 years (median 4,4 years).
Results: 31 patients (41%) underwent single stage full correction at a median age of 12 days, 16 patients could be treated by a transannular patch. 30 days mortality rate in this group was 6%. In the remaining 45 patients the VSD could not be closed during their first surgery at a median age of 89 days. A VSD closure was achieved later in 64 % of these patients after median 178 days. 30 days mortality rate after the first surgery was 13% in this group. Overall long term survival was 81,6% showing no significant difference between the groups with and without MAPCAs ($p > 0,999$). Cox regression revealed older age at first surgery and single stage full correction as positive influence on the occurence of reintervention after VSD closure. Median intervention-free interval (surgery and transcatheter intervention) after VSD closure was 1,7 ± 0,5 years [95%CI:0,7-2,8].
Conclusion: VSD closure was achieved in 79% and at a significant earlier age in patients without MAPCAs ($p < 0,01$). Although patients without MAPCAs predominantly underwent single stage full correction at newborn age, the overall mortality rate and the interval until reintervention after VSD closure did not show significant differences between the two groups with and without MAPCAs. The high rate of proven genetic abnormalities (40%) did also pay its tribute to impaired life expectancy.

Keywords: Tetralogy of Fallot, Pulmonary atresia, MAPCA's

P-349

Outcome of norwood patients with a birth weight less than 2,5 kg

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Background and Aim: Low birth weight is a known risk factor for mortality after the Norwood procedure. A bilateral pulmonary artery banding and open ductus arteriosus can postpone the Norwood procedure and might improve survival of premature or low birth weight neonates. A retrospective single center analysis was carried out investigating in hospital mortality and long-term survival of Norwood patients born with a body weight of less than 2,5 kg.
Methods: 7% of 434 Norwood patients of our center showed a birth weight below 2,5 kg. Hence 30 patients operated between 2002 and 2017 were included in the study. Group A consists of 22 low birth weight patients who had a primary Norwood procedure at a median age of 9 days (range 3-29 days).Their body weight at the Norwood procedure was mean 2,3 kg (range 1,9-2,47 kg). Group B consists of 8 patients who received a bilateral pulmonary artery banding due to low birth weight between 1,2-2,0 kg (mean 1,7 kg). The arterial duct was kept open with prostaglandines or stent implantation (2 cases).
Results: In hospital mortality of Group A (primary Norwood) was 27,3% (6/22 pts). Two more patients died at the age of 2 months (total mortality 36,4%). Group B (PAB) showed only one case of early mortality (12,5%) at the age of 19 days after PAB and

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Norwood and TAPVC repair 2 weeks later. Another child of Group B died at the age of 2,4 years (total mortality 25%). The median follow up period of all survivors is 10,2 years (range 4,2-17,3 years). The Norwood procedure in Group B was done at a median age of 57 days (range 19-180 days) with a mean body weight of 2,76 kg.
Conclusion: Bilateral pulmonary artery banding seems to warrant an advantage on outcome compared to a primary Norwood procedure in children with a birth weight of less than 2,5 kg. Long-term survival after PAB might be comparable to heavier patient groups with primary Norwood. Due to different weight groups and small sample size in our study a direct comparison between primary or staged Norwood in the small weight group is difficult.

Keywords: Norwood procedure, HLHS, pulmonary artery banding, low birth weight

P-350
Pediatric resuscitation outcome in congenital heart disease (PRO-CHD registry study)

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Background and Aim: Children with congenital heart defects, especially those after heart surgery, have an up to ten times higher risk for an in-hospital cardiac arrest, compared to children without heart disease. Although this patient cohort is particularly vulnerable, these events and outcomes are not systematically monitored in Germany. We designed a prospective multicenter-registry study aiming to record in-hospital cardiac arrests of children with congenital - (CHD) and acquired heart disease in Germany.
Methods: A web-based registry was designed, prospectively collecting data (patient´s demographics, CPR data, post-resuscitation care and clinical course, neurological outcome and follow-up), according to the Utstein template. All children (0-18 years) with heart disease suffering a cardiac arrest requiring resuscitation of ≥ two minutes will be included. The primary outcome is survival to discharge, the secondary outcome is morbidity, with a particular focus on neurological morbidity. All German pediatric cardiology centers will be encouraged to participate.
Results: This registry will provide data on the mortality, early and mid-term neurologic outcome, and quality of life after cardiac arrest of children with heart disease in Germany. Correlations between patients´ characteristics resuscitation characteristics and post-resuscitation care with primary and secondary outcomes will be analyzed.
Conclusion: By systematically recording and analyzing the outcome after in-hospital cardiac arrest in children with CHD, this study is an important first step to close knowledge gaps regarding the risk factors and outcome of cardiac arrest in this patient group, and aims to improve care and outcomes of these vulnerable patients (figure 1).

Keywords: resuscitation, neurologic outcome, quality of life, registry study

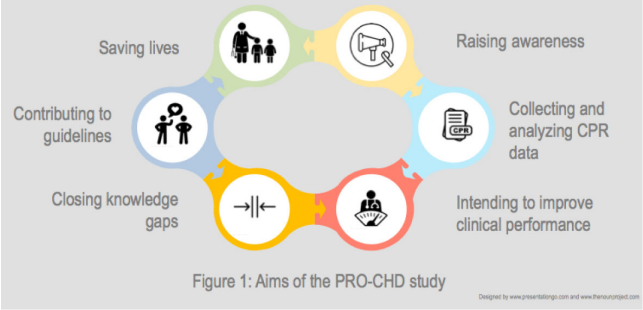

Figure 1: Aims of the PRO-CHD study

P-351
Extra-cardiac, beating heart surgical preparation for subsequent percutaneous fontan completion: a valuable alternative?

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Background and Aim: The Fontan operation is considered as the final palliation for complex heart diseases with univentricular physiology. To reduce the number of surgical procedures several hybrid approaches have been developed. A simple alternative that involve extra-cardiac, beating heart surgical preparation for a patient to receive transcatheter Fontan completion (TFC) at a later stage is presented.
Methods: A 5-year-old,18kg boy, was found to have atrio-ventricular discordance, ventriculoarterial concordance and multiple ventricular septal defects (VSD). At 14-months of age, a bidirectional-Glen shunt and pulmonary artery (PA) banding were performed. At 3-years, he underwent a one-and-a-half ventricle repair (1.5VR): hemi-Mustard procedure, main PA was debanded and VSDs closed. One month later, as a result of circulatory failure, reoperation was undertaken to take down of the 1.5VR and to prepare for subsequent TFC.
Results: 1. Extra-cardiac, beating heart surgical preparation: the roof of the right atrial (RA) appendage was attached to the unopened lower part of the right PA. A 20mm Gore-Tex tube was anastomosed, in an extracardiac fashion, to the base of the RA appendage and to the lower part of the right PA. A 20mm ring of Gore-Tex was placed around the inferior vena cava above the diaphragm (fig. 1A). 2. TFC (26-month after surgical preconditioning): access from the right internal jugular vein (8Fr) allowed needle perforation of the lower part of the right PA and passage into the RA. Through the left femoral vein (20Fr), Andra XXL 57 and 30 mm stents, followed by a covered CP10Z60 stent mounted on a 20mm BIB® and fenestrated in the middle, were deployed to connect the IVC to the PA. A 9mm Bentley stent was then expanded with a 10mm Conquest balloon across the left PA to exclude pulmonary forward flow. Shortly after, severe desaturation developed related to a too large fenestration (fig. 1B). Two CP8Z39 stents mounted on a 24mm BIB®, allowed complete obliteration of the right-to-left shunt. No other significant adverse events occurred. Patient was discharged home on day eight.
Conclusion: TFC following an extra-cardiac, beating heart surgical preparation seems to be a feasible practice, however, further research will be needed to substantiate its clinical applications.

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Keywords: Complex Congenial Heart Disease, Univentricular Physiology, Hybrid Procedures, Transcatheter Fontan completion

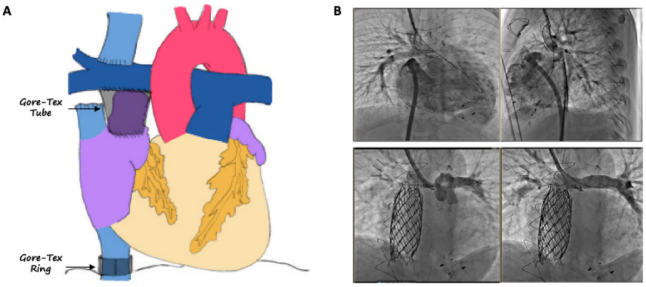

Figure 1 A. Sketch of the operative product. B. Angiograms before and after transcatheter Fontan completion.

P-352
Intraoperative flow study in tetralogy of fallot with pulmonary atresia and major aortopulmonary collaterals: a novel approach

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Background and Aim: Ventricular septum defect (VSD) closure in tetralogy of Fallot (TOF) with pulmonary atresia (PA) and major aortopulmonary collateral arteries (MAPCAs) and hypoplastic native pulmonary arteries can lead to fatal suprasystemic right ventricular pressures postoperatively. Therefore, pre- and intraoperative assessment of pulmonary resistance is essential for choosing the right surgical management strategy. An intraoperative flow study after unifocalization was introduced in order to facilitate the decision for primary or staged VSD closure. Our novel approach is using a temporary right heart bypass to measure pulmonary resistance at almost physiologic parameters without cardiac arrest.
Methods: Five patients underwent TOF repair with right ventricle to pulmonary artery conduit (RVPAC) implantation and unifocalization (n = 2), PA patch plasty (n = 1) or without pulmonary artery reconstruction (n = 2) at our institution from January to November 2021 at a median age of 6.6 month. After pulmonary artery reconstruction the intraoperative flow study was conducted to decide whether VSD closure can be performed or not. Therefore, patients were intraoperatively ventilated and the arterial cannula was switched from the aorta to the RVPAC. This right heart bypass flow was increased up to 3l/min/m^2 and pulmonary artery pressure was invasively measured.
Results: This technique was feasible without problems. Pulmonary artery pressure on right heart bypass was below 25mmHg in four patients and 31mmHg in one patient allowing successful primary VSD closure in four patients and successful staged closure in one patient. Median bypass time was 196 min, median aortic cross clamp time 64 min. 3 Patients were extubated within 24 hours, one at post operative day 3 and one patient had a prolonged weaning course of 13 days. Median ICU stay was 3 days and median hospital stay 14 days. One patient suffered an intermittent acute kidney failure post operatively but regained normal kidney function.
Conclusion: The intraoperative flow study using a temporary right heart bypass is simple to establish and pulmonary vascular resistance can be measured at almost physiologic parameters. Decision making for or against VSD closure can be supported by this method to achieve a complete repair without the risk for increased right ventricular pressures.

Keywords: tetralogy of Fallot, pulmonary atresia, major aortopulmonary collateral arteries, ventricular septum defect closure, intraoperative flow study

Intraoperative flow study

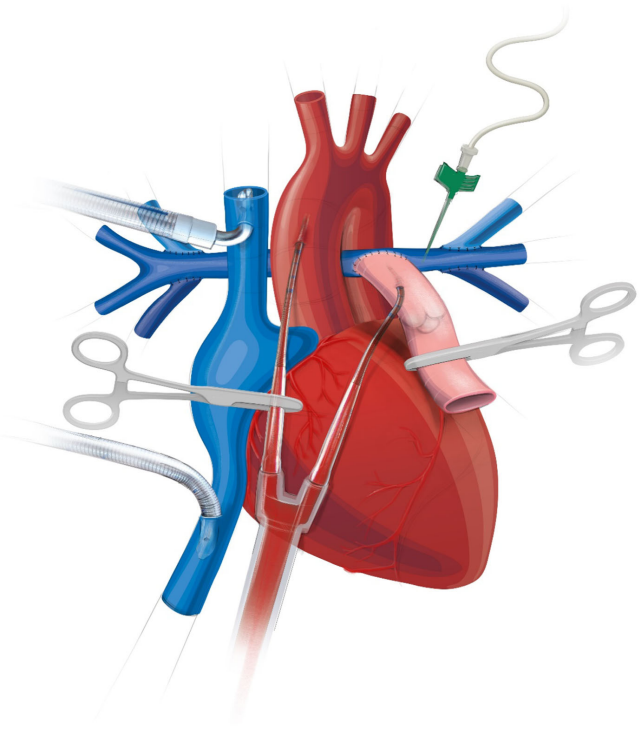

Figure Temporary right heart bypass to measure pulmonary vascular resistance intraoperativley: flow 3l/min/m^2

P-353/Moderated Poster
Surgical treatment of the aortic coarctation depending on the anatomical characteristics

Hyungtae Kim, Kang Ho Choi, Si Chan Sung; Department of Thoracic and Cardiovascular Surgery, Pusan National University Yangsan Hosptial, Yangsan, South Korea

Background and Aim: To evaluate early and long-term results of surgical treatment of the aortic coarctation (CoA) depending on the anatomical characteristics.
Methods: A retrospective clinical review of the patients with CoA undergoing surgery between March 2011 and August 2020 was performed. We devise a surgical classification to separate various entities in the spectrum of CoA: type I = tightly packed all three head vessels; type II = left subclavian artery separated from two other head vessels; type III = all three head vessels separated from one another. Each of these types has subtypes: a = without long isthmic portion and b = with long isthmic portion.

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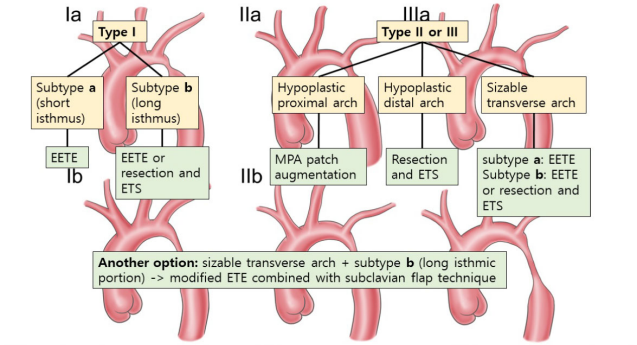
Results: One hundred eleven consecutive patients were included. The median age and weight at operation were 8 days (range, 1-1490 days) and 3.2 kg (range, 1.9-18.5 kg). Several operative techniques have been performed depending on the anatomical characteristics: extended end-to-end anastomosis (EETE) via sternotomy in 54 (48.6%); EETE via thoracotomy in 12 (10.8%); resection and end-to-side anastomosis in 31 (27.9%); main pulmonary artery patch augmentation in 12 (10.8%); and modified end-to-end anastomosis combined with subclavian artery flap technique in 2 patients (1.8%). Early mortality was one case (0.9%), and postoperative complications occurred in 12 patients (10.8%). The mean follow-up duration was 43.9 ± 29.8 months. During follow-up, there were two late deaths (1.8%). Five patients (4.5%) have undergone re-interventions, and three patients (2.7%) have had reoperations for the restenosis of CoA. All type Ia patients (21 patients, 18.9%) have undergone EETE via sternotomy or thoracotomy.
Conclusion: Surgical treatment of the CoA depending on the anatomical characteristics is a safe and low-risk procedure concerning either early and late outcomes.

Keywords: Coarctation of the aorta, Anatomical classification, Congenital heart disease

figure_01



EETE = extended end-to-end anastomosis; ETE = end-to-end anastomosis; ETS = end-to-side anastomosis

Surgical strategy depending on the anatomical characteristics

P-354

Salvage mitral valve replacement with a home-made gore-tex tricuspid valve for an infant

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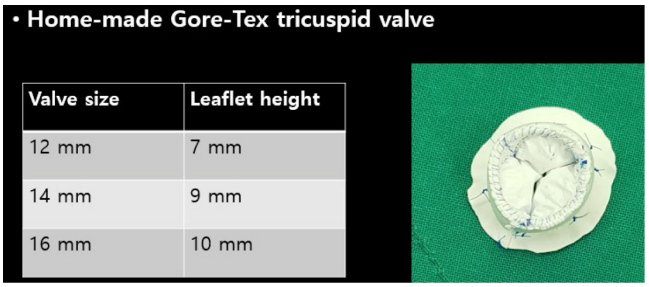
Background and Aim: Mitral valve replacement (MVR) in small infants is associated with high morbidity and mortality. There are no ideal prostheses available for those patients. We report our initial experience on MVR using a home-made Gore-Tex tricuspid valve for an infant patient.
Methods: A home-made Gore-Tex tricuspid valve was prepared and used for a 7-month patient requiring MVR.
Results: Preoperatively, the patient showed unstable vital status and ventilator dependency. After MVR with a 12mm home-made Gore-Tex tricuspid valve, the patient could be extubated from the ventilator. However, a 3.6-month follow-up echocardiography demonstrated a sustained severe mitral stenosis, and we performed redo-MVR with a 16mm home-made Gore-Tex tricuspid valve at the supra-annular position. The patient is still doing well for a 3-month out-patient follow-up.
Conclusion: Our home-made Gore-Tex tricuspid valve could be feasible for the small infant requiring MVR.

Keywords: Mitral valve stenosis, mitral valve replacement, artificial mitral valve

figure_01



P-355

Early and mid-term results after hybrid-surgical strategies in mitral and pulmonary melody valve implantation in children – multi-institutional MID-TE

Ireneusz Haponiuk[1], Maciej Chojnicki[2], Konrad Paczkowski[2], Julia Haponiuk Skwarlińska[3], Mariusz Steffens[2], Anna Romanowicz Sołtyszewska[2], Marta Paśko Majewska[2], Monika Opacian Bojanowska[2], Paweł Macko[2], Katarzyna Gierat Haponiuk[4], Joanna Sawicka[5]
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Background and Aim: Stented bovine jugular vein graft (Melody valve), originally designed for percutaneous implantations in pulmonary position in adult patients, is one of very few available valvular prostheses adjusted to the pediatric valvular implantations, with reported promising mid-term results. We present early and mid-term results after 4 consecutive unique hybrid-surgical strategies in Melody valve pediatric implantations.
Methods: The first two patients presented with a rapid deterioration of acute endocarditis. The children aged 23 and 15 months after preoperative antibiotic therapy, were operated because of severe mitral incompetence, presented as leaflets disruption and chordae discontinuation in echocardiography. After initial attempts for mitral valvuloplasty, the hybrid methods of self-modified Melody valves implantation into the mitral positions (Melody-MVR) were used. The third patient presented with a previous multi-institutional history of neonatal persistent truncus arteriosus(PTA) type II surgical repair and a percutaneous implantation of 2 stents into the pulmonary trunk and a Melody valve into the pulmonary position. Due to an incidental obstruction of the right pulmonary artery, the patient was reoperated in the emergency settings including impending stent removal with perfect

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Melody valve function preservation and hybrid intraoperative balloon plasty of both pulmonary arteries. The fourth patient was 6 months-old boy with genetical disorders and unfavorable common atrioventricular (AV) defect, with critical AV-valve incompetence, severe heart and respiratory failure. The boy underwent an alternative correction of cardiac defect with single PTFE-patch technique and an initial implantation of double-folded inverted Melody valve into the mitral position (CAVSD-Melody-MVR).
Results: The intraoperative epicardial echocardiography(EE) in all the patients confirmed good Melodies function without any signs of left ventricular outflow tract obstruction(LVOTO). Short term transesophageal echocardiography(TTE) and angiography control in all the patients and mid-term control in the first two patients (2 vs 36 months) showed constantly competent mitral valve Melody prosthesis, while the last patient presents slow improvement of genetically-defined multiorgan failure.
Conclusion: To conclude, pediatric Melody implantation appeared to be safe and effective treatment for pediatric patients with mitral or pulmonary non-reparable valve disfunction, with good early and mid-term results, despite the endocarditis and generalized non-cardiac problems, as well as unfavorable percutaneous treatment.

Keywords: Melody valve, pediatric mitral valve replacement, Melody-MVR, CAVSD-Melody-MVR, acute endocarditis, pediatric cardiac surgery.

P-356
Pulmonary atresia with interventricular defect and no intrapericardial pulmonary arteries - what to expect?

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Background and Aim: Pulmonary atresia with an interventricular defect (PA+VSD+MAPAs) is a disease entity with a wide spectrum of anatomic variants and clinical manifestations. The form of pulmonary vascularization has the main influence on the management and prognosis. The aim of the presentation is to present the treatment results of this difficult group of patients- - observation performed by one cardiologist.
Methods: Data from 53 patients operated on for PA + VSD + MAPCAs aged 1.8 years (0.4-11) were analyzed. None of the patients had intrapericardial pulmonary arteries. The observation period was between 2 months and 14 years (3,4 ± 3,2 years). The operations were performed in 5 different centers. In the period between 6 and 12 months after the procedure, a control cardiac catheterization was performed.
Results: Complete one-stage correction was performed in 45/53 patients. In 13/53 patients, systemic-pulmonary shunt was performed previously, and in 5 of them 1 - 3 MAPCAs were additionally ligated. In 8/53 pts, unifocalization with shunt (5/8 most often – Laks shunt) or with reconstruction of the outflow tract from the right ventricle (3/8) was performed. 4/8 patients underwent complete correction within 0.8 years on average.
During FU the right ventricular/aortic (RV/Ao) pressure ratio was 0.43 ± 0,18 in control cardiac catheterization. Balloon angiopathy of pulmonary artery stenosis was performed in 33 pts (including 24 with cutting balloons). In this group, a decrease in the RV/Ao pressure ratio from 0.71 ± 0,21 to 0.42 ± 0,28 was achieved.
2 pts died during observation. Cardiac surgery procedures were performed in 12 pts: homograft was replaced in 7, pulmonary arteries were dilated in 7 (including 3 with "ghost vassels" unifocalisation). 3 pts were disqualified from further surgical treatment.
Conclusion: In recent decades, the treatment outcomes of patients with PA + VSD + MAPCAs have improved significantly, but they still require constant monitoring, including cardiac catheterization and interventional management.

Keywords: pulmonary atresia, unifocalisation, angioplasty

P-357
Minimally invasive access for pulmonary valve replacement, 2 years of experience with a novel approach

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Background and Aim: The population of patients requiring pulmonary valve replacement (PVR) following surgery during infancy, such as tetralogy of Fallot repair, is growing. Despite advances in transcatheter pulmonary valve implantation, surgery remains indicated in patients with large regurgitant pulmonary valves or stenotic right ventricle-to-pulmonary artery conduits. Full conventional sternotomy is widely used for PVR, while left anterior minithoracotomy has been described only in selected cases.
Our aim is to describe our early experience with upper left ministernotomy as a systematic approach for PVR and redo PVR in teenagers and adult congenital patients.
Methods: From December 2019 to December 2021, 13 patients underwent PVR via ministernotomy at our institution. The indications were severe pulmonary regurgitation in 11 patients and severe conduit stenosis in 2 patients. Ten patients (76.9%) already had at least 1 sternotomy, including 6 patients who previously underwent tetralogy of Fallot repair. Of note, PVR was performed in a teenager with a history of 4 sternotomies. Pulmonary homografts, stentless porcine bioprostheses or stented pericardial bioprostheses were implanted. Median age at operation was 24.6 years old (17.6 – 38.4).
Results: There were no deaths, no cardiac injuries and no conversions to full sternotomy. The 3rd intercostal space was the most frequently used for the leftward extension of the ministernotomy. In 6 patients (46.1%), a residual intracardiac shunt was present and aortic cross-clamping was necessary. Median cardiopulmonary bypass and cross-clamping times were 70 (64 – 105.5) and 50 (40 – 75) minutes respectively. Median intensive care and hospital stays were 2 (1 – 2.5) and 7 (6 – 11) days respectively. Three patients required transfusion with blood products. All patients are doing well after a median follow-up of 13.1 months (10.3 – 16).
Conclusion: Ministernotomy is a safe and feasible approach for PVR and redo PVR in teenagers and adults. This approach combines the excellent outcomes of minimally invasive surgery (minimal blood loss and expedited recovery), which are even more obvious in patients with previous sternotomies, and the advantages of a

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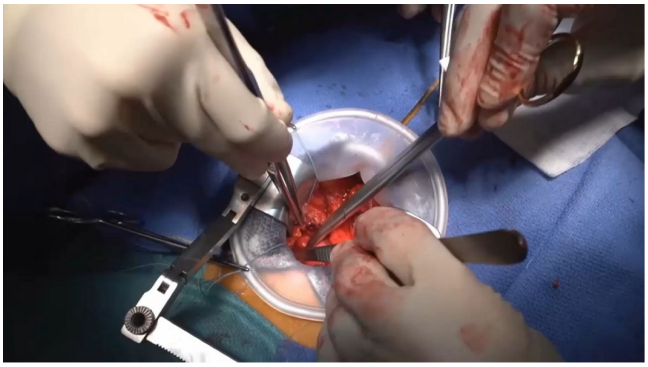
midline incision such as the access to the ascending aorta and the absence of additional scar in cases of redo PVR.

Keywords: Pulmonary valve replacement, minimally invasive cardiac surgery, ministernotomy, tetralogy of Fallot, pulmonary regurgitation

Video still of a redo pulmonary valve replacement



P-358

Initiating a pediatric cardiac surgery program in europe during the COVID-19 pandemic: methods, challenges and outcomes

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Background and Aim: Before 2020, no pediatric cardiac surgery program was available at our institution, despite being a university hospital providing tertiary care for 6 million inhabitants.
Our goal is to describe the preparation and the first year of experience of our pediatric cardiac surgery program, which will eventually cater for 300 patients annually.
Methods: The project was supported by European funds (INTERREG program). Medical and nursing staff training was performed via a transborder collaboration. Significant investments were necessary to reach the required standards for the facilities (operating rooms, pediatric intensive care beds) and equipment (cardiopulmonary bypass and ECMO machines, ultrasound systems etc.). A multidisciplinary team was built over 3 years. The pediatric ECMO program was started a year prior to the surgical program. In parallel, a program dedicated to the study and care of neurological impact of congenital heart diseases and interventions was set up. Importantly, a progressive upscale was devised: only children with a weight > 5 kg requiring non-complex surgeries were operated on during the first year.
Results: The first year of experience was marked by challenges caused by the successive COVID-19 waves, such as restricted access to the operating room and a subsequent slow-down in the progression of the schedule. Fifty-nine patients constituted the cohort of the first year (October 2020 – October 2021). In addition to low-risk procedures (left-to-right shunts closures etc.), cases included 6 tetralogy of Fallot repairs, 1 Ross procedure and 2 bilateral cavopulmonary connections. There were no early or late deaths. Median age was 6.3 years old (1.8–9.8) and median weight was 18.5 kg (10.0–32.0). Fourteen patients (23.7%) were operated on with a weight < 10 kg. Bypass cases represented 72.9% (43 patients) of all cases. Median cardiopulmonary bypass and cross-clamping times were 88 (52–153) and 51 (26–98) minutes respectively. Median intensive care and hospital stays were 3 (2.0–6.7) and 6 days (5–11) respectively.
Conclusion: Despite COVID-19-related difficulties, our pediatric cardiac surgery program achieved excellent outcomes in selected patients. Institutional support, meticulous planning, team cohesion and perseverance are keys for successful initiation of a program requiring such high technicality.

Keywords: pediatric cardiac surgery; regional public health; European Union; new surgical program; COVID-19

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Results of surgical versus balloon angioplasty treatment for critical coarctation of aorta in neonates and young infants

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Background and Aim: Coarctation of aorta (CoA) is a common congenital heart defect that accounts for 5-8% of all congenital heart diseases. Neonates and young infants with critical CoA very often present with congestive heart failure and need urgent treatment. Treatment could be surgical or by balloon angioplasty (BA). Surgery is the standard treatment for native coarctation in neonates, hovewer BA may be preferred as an alternative treatment in selected patients. Our objective was to compare our results of surgical versus balloon angioplasty treatment for critical coarctation of aorta in neonates and young infants.
Methods: Between 2018 and 2021, a total of 18 procedures, includidng 8 BA and 10 surgical treatments, were performed in neonates and young infants less than 2 years of age. Mean age for BA group was 5.3 ± 7.2 months (range 5 days to 22 months) and mean age for surgical group was 7.1 ± 8.4 months (range 6 days to 24 months). The hospital records, echocardiography reports, catheterization data, angiography images and operative data were reviewed.
Results: 3 patients of 8 in BA group were presented re-coarctation during follow-up period and were referred to surgical correction. Surgical correction of these patients went uneventfully. In surgical group also 3 patients presented re-coarctation during follow-up period. 2 patients were corrected successfully by BA. The 3rd patient has also a ventricular septal defect (VSD) and the re-coarctation was re-operated together with the VSD surgery successfully. There was no mortality in both groups during follow-up period. There was no significant difference in hospital stay and immediate transcoarctation residual gradient between the groups. However, the ICU stay and ventilation duration is significantly higher in surgery group.

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Conclusion: In our small group of patients, we observed that both BA and surgical treatment are efficient and safe in the treatment of critical CoA in neonates and young infants.

Keywords: Coarctation of aorta, balloon angioplasty.

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Congenital left ventricular aneurysm repair:a case report on a rare condition

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Background and Aim: Left ventricular (LV) aneurysms can be asymptomatic or may be found when patients present with heart failure, rhythm disorders, or peripheral arterial embolisms. LV aneurysms are caused by myocardial infarction, hypertrophic cardiomyopathy, Chagas disease, tuberculosis, trauma, and surgery. Congenital LV aneurysms are extremely rare and are generally isolated (1). Although there are various clinical presentations, most cases are asymptomatic. We present the case of a newborn pediatric patient who underwent an LV aneurysm repair.
Methods: A 1-year-old male patient underwent an aneurysmectomy and plication to correct an LV aneurysm. Routine controls revealed the presence of an LV aneurysm, so he was hospitalized for further investigation and treatment. He was completely asymptomatic: an electrocardiogram indicated that his sinus rhythm was normal, but a preoperative chest x-ray showed cardiomegaly, especially in the left ventricle. Blood tests indicated that his pro-BNP was elevated. A preoperative echocardiogram (echo) revealed a left ventricular aneurysm with a diameter of 15 × 23 mm on the lateral wall and decreased LV systolic function. The ejection fraction of the LV was 38%. CT angiography showed a 20 × 24 mm saccular aneurysm originating from the LV lateral wall at the midventricular level (Fig. 1). A multi-disciplinary pediatric heart team discussed the case and recommended a surgical resection. Intraoperatively a saccular mass of approximately 2 × 2.5 cm was detected in the LV wall (Fig. 2) and an aneurysmectomy was performed. The aneurysm sac was found to be filled with a thrombus (Figs. 3, 4). The defect was closed with a Dacron patch and 5-0 Prolene sutures (Fig. 5). The patient had an uneventful post-surgical course and was discharged on post-operative day 6.
Results: No recurrence was detected on a follow-up echo conducted 1 year after the surgery and the patient's LV function improved dramatically in the postoperative period.
Conclusion: LV aneurysms are extremely rare in the neonatal period, and often accompany other pathologies. Diagnosis is difficult because the condition is usually asymptomatic. Although there are few cases reported in the literature, we believe that using a Dacron patch to repair LV aneurysms is a safe and lifesaving surgical choice.

Keywords: LV aneurysms, congenital heart surgery, aneurysm repair.

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Inducing labour does not reduce preoperative morbi-mortality in infant with prenatal diagnosis of tranposition of great arteries

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Background and Aim: Management during first hours of life in infant with prenatal diagnosis of transposition of great arteries (TGA) are highly predictive of preoperative morbimortality. The aim of this study is to assess the effect of induction of labor on fetuses with transposition of great arteries.
Methods: Retrospective study in newborn infants with prenatal diagnosis of TGA after an induced or a spontaneous delivery. We evaluated hour at birth and pre-operative morbidity defined by use of inotropic drug, mechanical ventilation, metabolic acidosis and necrotizing enterocolitis. Working hours were defined from 8am to 5pm.
Results: 93 infants were included in the study (61% male). Labor was induced in 54 infants, delivery was spontaneous in 39 infants. The 2 groups were similar regarding gestational age (38weeks, $p = 0.58$), sex, and birth weight (3300 vs 3200g, $p = 0.36$). 38 infants were born at working hours in the induce delivery group compared to 20 in the spontaneous delivery group ($p = 0.06$). Balloon atrial septostomy was performed at 5.1 ± 7.4 hours in the induce delivery group and 2.8 ± 3.5 hours in the spontaneous delivery group ($p = 0.78$). Pre-operative morbidity was reported for 25 infants in the induce delivery group and 22 infants in the spontaneous delivery group ($p = 0.27$). 3 infants died before surgery and 2 after surgery in the induce delivery group, versus 1 died before surgery in the spontaneous delivery group ($p<0.05$). The 2 groups were similar regarding the postnatal age at surgery (8.8 days ± 5.7 vs 8.2 days ± 3, $p = 0.88$).
Conclusion: Induction of labor in infant with prenatal diagnosis of TGA does not reduce pre-operative morbidity and mortality. Our study does not support the usefulness of induction of labor after a prenatal diagnosis of TGA.

Keywords: NICU, TGA, Induce labor

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Prophylactic use of PGE1 improves the outcome in infants with moderate or severe congenital diaphragmatic hernia (CDH)

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Background and Aim: Suprasystemic pulmonary hypertension and functional left ventricle hypoplasia are highly predictive for pulmonary morbidity and death in infants with Congenital Diaphragmatic Hernia (CDH). Aim of the study is to report the effects of prophylactic Prostaglandin E1 (PGE1) infusion in newborns with moderate or severe CDH and pulmonary hypertension.

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Methods: All the infants with moderate or severe CDH with PPHN and born in the Nord-Pas de Calais area, France, between January 2010 and December 2018, were included in the study. Moderate and severe CDH were respectively defined by LHR o/e between 26 and 35% or between 36 and 45% with the liver up and LHR o/e between 15 and 25% for the left CDH or under 45% for right CDH. The outcome of the newborn infants treated prophylactically with PGE1 to maintain patent the ductus arteriosus or not, were compared.
Results: 139 infants with isolated CDH were included in the Nord-Pas de Calais CDH cohort during the study period. 13 infants with moderate or severe left CDH were treated with prophylactic PGE1 (mean duration of infusion: 14 days) and compared to 267 infants. Infants were comparable regarding sex (male: 39% vs 50%, p = 0.5), gestational age (39 ± 1 GW vs 39 ± 2 GW, p = 0.99), birth weight (3000 ± 600 gr vs 3100 ± 700 gr, p = 0.43) and Fetal Lung Volume o/e (29 ± 9 % vs 32 ± 5 % p = 038).
Mortality rate at 1 year was 0/13 in the prophylactic PGE1 group and 6/26 in the infants not treated prophylactically by PGE1 (p = 0.02). No significant difference was found on duration of mechanical ventilation (85 hours [82-86] vs 84 hours [75-87], p = 0.78), duration of O2 supplementation (51 days [0-78] vs 59days [0-81], p = 0.75), and days without hospital at 90 days (91 ± 49 days vs 83 ± 81 days, p = 0.75).
Conclusion: Prophylactic use of PGE1 in infants with CDH and moderate to severe lung hypoplasia is associated with a decrease risk of death during the first year of life. We speculate that patent ductus arteriosus may support cardiorespiratory function during the early life in infants with moderate or severe CDH.

Keywords: Congenital Diaphragmatic Hernia, NICU, PGE1, ductus arteriosus, PPHN

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Initial experience with the fontan procedure in latvia

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Background and Aim: The first Fontan procedure performed in the Children's clinical university hospital in Riga was commenced in 2012. Since then, the perioperative care and surgical skills have improved significantly, although in a population of 1,8 million it is a challenge to keep good quality with a low volume of Fontan procedures.
Methods: We reviewed the medical records of all the patients undergoing a Fontan procedure between 2012 and 2021 that was performed in the Children's Clinical University Hospital in Latvia.
Results: Between 2012 and 2021, a total of 19 patients underwent Fontan procedure in our hospital. Since November 2020 we have performed the Fontan procedure without the assistance of a mentor surgeon from a more experienced center. At the time of surgery, the mean age was 6.1 ± 2.0 (4-12) years. The mean weight at the time of surgery was 18.3 ± 3.9 (12-25) kg. Mean pulmonary artery pressure before the surgery was 9.3 ± 3.5 (4-17) mmHg. For three patients the main ventricle was the right ventricle, for 14 patients – the left ventricle, and for two patients both ventricles. Aortic occlusion was necessary for 5 patients to perform additional surgical interventions besides the Fontan procedure. Four patients did not have Fontan tunnel fenestration for several reasons. The mean cardiopulmonary bypass time was 142 minutes. 11 out of 19 patients were extubated in the operating theatre. The mean time for chest drains removal was 7.2 ± 2.8 (3–15) postoperative day (POD). For those extubated in the operating theatre, drains were removed significantly earlier (before POD7) compared with those extubated later (p 0.02). The mean length of stay in the intensive care unit was 7.1 ± 2.8 (3–16) days.
Conclusion: Completing the Fontan surgery in a country with a small population is feasible with acceptable results. Since 2018 most of the patients underwent fast-track extubation which has resulted in an earlier chest drain removal time.

Keywords: Fontan

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Aortic valve reconstruction with ozaki technique: our initial experience

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Background and Aim: Congenital aortic valve (AoV) disease affects young patients with an active lifestyle and still growing up. When conventional surgical repair techniques are insufficient to solve the problem, valve explant is mandatory. In these cases Ozaki technique offers neo-valve reconstruction with excellent hemodynamic without needing long-life anticoagulant therapy. Ozaki is a good alternative to other AoV surgical replacement like mechanical/biological prosthesis, homograft and autograft with Ross/Ross-Konno techniques We present our initial experience with Ozaki AoV reconstruction in congenital patients.
Methods: Retrospective study of 12 patients with congenital AoV pathology operated from October-2019 to October-2021. All received Ozaki procedure: Neo-leaflets are constructed using a set of sizers and templates leading the technique to be highly reproducible. Autologous pericardium is the gold standard to create the neo-leaflets. Finally we obtain a neo-AoV with a wide coaptation surface. Adequacy of the Ozaki was assessed with echocardiogram. Statistic analysis was performed with SPSS.20
Results: Median age: 14 years [IQR:8,8-28,3]: from 4,9 to 40 years, 7(58%) children (<18 years). Previous aortic valvuloplasty: 5(42%) percutaneous, 1(8%) surgical. AoV Diagnosis: 7(58%) double lesion, 3(25%) severe insufficiency, 2(17%) severe stenosis. AoV anatomy: 7 bicuspid, 4 unicuspid, 1 cuadricuspid. Ozaki was done with autologous pericardium in 11 patients to create the neo-leaflets, and heterologous pericardium patch was used in the child with previous surgery that also required Nicks annular enlargement (Figure-1). Mean extracorporeal and cross-clamp time: 174 ± 49, and 146 ± 36 minutes respectively. We had one technical failure needing reconversion to prosthetic valve replacement. In-hospital mortality was null. Postoperative median hospital stay was 7,5 days [IQR:7-9]. Follow up was complete, mean 10 ± 6 months: All patients are alive and well with normal neo-AoV (peak AoV gradient: 24 ± 12 mmHg, with AoV insufficiency ≤I) except one with progression to moderate insufficiency. We had no endocarditis or reinterventions until now.

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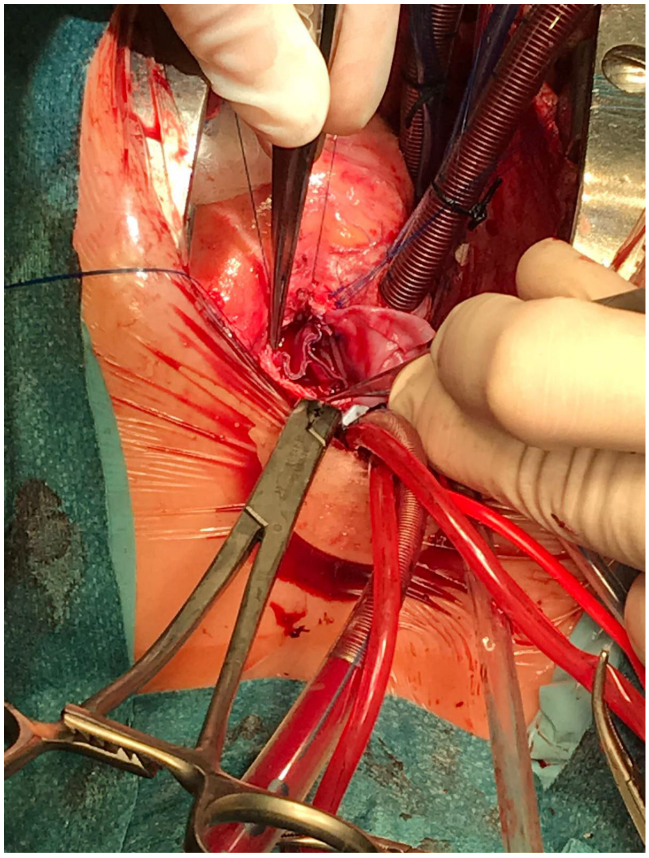
Conclusion: Ozaki technique is reproducible, and allows the creation of a neo-AoV with excellent hemodynamics and good initial results. Ozaki AoV reconstruction is promising in congenital patients with aortic valvulopathy not amenable to conventional repair techniques. We need time to confirm the advantages of the Ozaki respect to other traditional AoV replacements.

Keywords: Ozaki technique, congenital aortic valvulopathy



Ozaki with Nicks annular enlargement and heterologous pericardial leaflets in a 8 years old girl with previous aortic valve repair.

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Mitral prosthesis in children: the show must go on

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Background and Aim: Surgery of mitral valvulopathy in children not amenable to repair is a challenge. In these cases the only option is mitral valve replacement (MVR), to recover a competent mitral function. We present our experience with mitral prosthesis in paediatric age, focusing in our short and medium term surgical results, survival and freedom of reoperation.

Methods: Retrospective study: 41 MVR in 39 children (<18 years old), operated from 2002-2020. Statistical analysis performed with SPSS.20

Results: Median age: 5,2 years [IQR:1,7-13], with 15% <1 year. Median weight: 14 kg [IQR:8,5-41], with 32% ≤10 kg. Previous surgery in 61%; 81% had ≥1 valvuloplasty before/during MVR time. NYHA preoperative status III-IV: 75%. Preoperative pulmonary hypertension: 19%-moderate, 49%-severe. Mitral surgical indication: 11(27%) stenosis, 20(49%) regurgitation, 10(24%) double lesion. All patients were operated and receive a mitral prosthesis (Table-1) Inhospital mortality: 2(4,9%). Postoperative median intubation: 17 [IQR:3-87] hours, intensive care unit stay 7 days [IQR:4-13] and hospital stay 15 days [IQR:9-27]. 10(24%) patients needed reoperation, most frequently an additional pacemaker (5), bleeding revision (4) and mitral re-replacement indicated by prosthesis thrombosis (2). Follow-up was complete in 95%, median 50 months [IQR:20-99]. Late mortality: 5(13%). Kaplan-Meyer survival curve was 92%, 85%, and 79% at 1, 5, and 10 years respectively. Inhospital and late mortality risk factors: preoperative NYHA status (p<0,001), mitral stenosis (p:0,05), low weight (p:0,003), small prosthesis size (p:0,015). Late mitral reoperation required in 7(19%), mostly related to patient overgrowth/mismatch and prosthesis thrombosis. Late mitral reoperation risk factors: Low age (p<0,001), low weight (p< 0,001), previous surgeries (p:0,025), mitral stenosis (p:0,05), small mitral annulus (p:0,002), small prosthesis size (p<0,001). Nowadays 97% are in NYHA status I-II; 15% and 7% patients have moderate and severe pulmonary hypertension respectively.

Conclusion: MVR is the last technical option in children with not repairable valves. We observed NYHA status and pulmonary hypertension improve after mitral prosthesis, with 4,9% inhospital and 13% late mortality. Morbidity is still a matter of concern, with 24% needing early reoperation and 19% in the mid term follow-up. Low weight, mitral stenosis and small prosthesis size are risk factors associated to mortality and reoperation.

Keywords: Mitral prosthesis children, congenital mitral valvulopathy

Table-1: Mitral valve prosthesis: Surgical details

Surgical MV approach	Transeptal 11 (27%), Left atriotomy 27 (66 %), superior septal 3 (7%)
Prosthesis type	Mechanical 36 (88%), bioprosthesis 5 (12%)
Prosthesis size	Median 21 mm (IQR: 17-26)
Prosthesis implantation	• Supraanular 17 (41%): 6 of them supported with a conduit Annular 24 (59%) • Papillary muscle conservation 19 (46%)
ECC time	168 minutes (IQR : 129-220)
Aortic cross-clamp time	115 minutes (IQR : 94-140)
Associated surgery in 17 (41,5%)	Subaortic membrane resection 4 (10%), Foramen ovale closure 4 (10%), Aortic valve replacement 3 (7,3%), ECMO 2 (5%), Pulmonary valve bioprosthesis 1 (2,4%), Interventricular comunication closure 1 (2,4%), definitive pacemaker 1 (2,4%), Tricuspid annuloplasty 1 (2,4%),

MV: Mitral valve, IQR: interquartile range, ECC: Extracorporeal circulation, ECMO: Extracorporeal circulation with membrane oxigenator

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Risk factors for early pulmonary homograft dysfunction in congenital heart disease

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Background and Aim: Pulmonary homografts (PH) are used as a first-line treatment for surgical right ventricular outflow tract (RVOT) reconstruction in patients with congenital heart disease (CHD). Despite a better freedom from reintervention than prosthetic conduits, PH are not spared from failure and cases of early dysfunction are regularly described. The aim of this study was to assess the rate of early PH dysfunction in patients of the Bordeaux University Hospital and to identify associated risk factors.
Methods: A monocentric retrospective study was conducted in children and adults with CHD and PH implantation for RVOT reconstruction. Clinical and echocardiographic data were collected during follow-up. PH dysfunction was defined as a peak of gradient greater than 50 mmHg and/or as pulmonary regurgitation greater than moderate. Early dysfunction was defined as occurring within two years postoperatively. Primary endpoint was the early PH dysfunction rate at 2 years. The dysfunction-free survival curve was calculated according to the Kaplan-Meier method. A logistic regression with univariate then multivariate analysis was performed to identify risk factors for early dysfunction.
Results: Between January 2002 and November 2020, 112 PH were implanted in 110 patients and 11 cases of homograft dysfunction were reported during the follow-up, including 9 cases of early dysfunction. The rate of early dysfunction was 9.4 [3.3 - 15.1] % and freedom from reintervention was 94.6 [90.0 - 99.0] % at two years. The only independent risk factor identified by the multivariate analysis was duration of extracorporeal circulation ($p = 0.007$) but the length of stay in intensive care unit ($p = 0.088$) and the initial maximum pulmonary transvalvular gradient ($p = 0.06$) were also close to significance in the multivariate analysis.
Conclusion: Although PH provide a durable substitute for RVOT reconstruction, a significant proportion of patients presents early PH dysfunction and requires premature reintervention. An inflammatory mechanism is suspected but dedicated studies should be conducted to validate this hypothesis.

Keywords: Pulmonary homograft, early dysfunction, congenital heart disease

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Low fibrinogen is associated with early adverse outcomes after cardiac surgery in children

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Background and Aim: Bleeding complications are common after cardiopulmonary bypass (CPB) surgery for congenital heart disease (CHD). While lack of consensus exists on effective therapies, fibrinogen has been identified as a major candidate marker for target-oriented perioperative management of bleeding. AIM: To study the incidence of hypofibrinogenemia after paediatric cardiac surgery and its association with early adverse outcomes.
Methods: Retrospective single-centre study of children undergoing surgery for CHD over a 2-year period. Data was collected from electronic medical records. Postoperative hypofibrinogenemia was defined as a serum level below 2 g/L in the 24 hours immediately after surgery. Main outcomes were mortality in the first 24 hours, postoperative need for any blood product, re-intervention, low output cardiac syndrome (LOCS), prolonged mechanical ventilation and ICU length-of-stay.
Results: We included 200 patients; 50.5% male; mean (SD) age, 3.9 years (3.3); weight, 15.3 kg (15.2); 76% ($n = 152$) undergone CPB. Mean (SD) RACHS was 2.3 (0.8), with no difference between CPB and non-CPB surgeries ($p = .13$). Fibrinogen levels were significantly lower after CPB (1.8 ± 0.73 vs 2.1 ± 0.53 g/L;$p = .02$), regardless of CPB duration (Pearson's $r = -.07$;$p = .48$). On multiple regression analysis, postoperative hypofibrinogenemia was independently associated with increased need for blood transfusion ($p = .04$), early re-intervention for haemostasis control ($p = .04$), LOCS ($p<.001$) and death ($p = .016$), after adjusting for age, weight, RACHS and postoperative serum levels of haemoglobin and platelets. Early postoperative mortality was 3.0% ($n = 6$) and was highly associated with greater surgical risk (RACHS 3.8 vs 2.2;$p<.001$).
Conclusion: Low fibrinogen levels are associated with increased risk of death and early adverse outcomes after cardiac surgery in children with congenital heart disease. Cardiopulmonary bypass is an important risk factor for postoperative hypofibrinogenemia and should warrant close monitoring of this key element in haemostasis. Intraoperative monitorization and early correction of low fibrinogen may be an effective strategy to improve early survival and outcomes in paediatric congenital heart surgery. More studies are needed to prospectively address the clinical impact of fibrinogen as a therapeutic target after cardiopulmonary bypass surgery.

Keywords: Fibrinogen, bleeding, paediatric cardiac surgery, cardiopulmonary bypass, congenital heart disease

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Pulmonary arteries growth after bidirectional cavopulmonary connection in functionally univentricular heart program

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Background and Aim: The optimization of the pulmonary arteries (PA) size is an important concern after the bidirectional cavopulmonary connection (BDCPC) in patients with functionally univentricular heart as it precedes the completion of the Fontan program. We analyzed the PA growth between the time of the BDCPC and the time of the total cavopulmonary connection (TCPC) in a large monocentric cohort of children, and we explored the factors associated with differential growth profiles.
Methods: 118 patients (median age at the BDCPC 8 months (IQR 5-18)) were included. Diameters of the right and the left PA were measured proximally to the first lobar bifurcation on the pulmonary artery angiography obtained before BDCPC and compared to angiographies obtained before TCPC. Nakata index was also calculated using these two measurements. Associations of pertinent covariables with the PA growth were explored.

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Results: Both pulmonary artery branches grew between BDCPC and TCPC (9.1mm (IQR 7.2-12.2) to 12.2mm (IQR 9.9-14.4) for right PA diameter; p<0.0001, 8.5mm (IQR 6.5-7.3) to 9.6 mm (IQR 8.0-12.4) for left PA diameter; p = 0.0001). However, PA branches growth was not proportional to child's growth as we noted a decrease of the Nakata index over this period. The main factor associated with a better PA branch growth was the presence of an additional pulmonary blood flow through a pulsatile antegrade flow (+28.2mm2 for right PA section area; p = 0.020, +16.8mm2 for left PA section area; p = 0.043). No other factor preceding BDCPC or associated procedure performed simultaneously had an impact on PA branch growth.
Conclusion: PA branches growth is reduced after BDCPC and this may hamper the functioning of the future TCPC. Maintaining a pulsatile antegrade blood flow whenever possible could optimize PA growth and prevent deleterious early remodeling of the pulmonary vascular bed.

Keywords: functionally univentricular heart, bidirectional cavopulmonary anastomosis, pulmonary arteries growth, Nakata index, pulsatile additional pulmonary blood flow

P-369
The influence of the coronary reimplantation site on the proximal coronary artery diameter after the arterial switch operation

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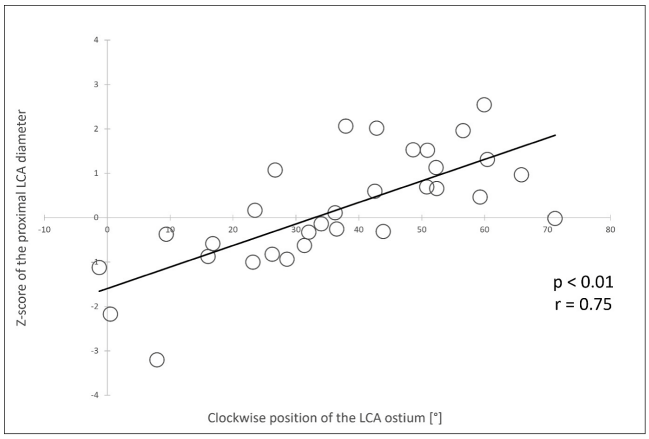
Background and Aim: Coronary events are life-threatening long-term complications of the arterial switch operation for transposition of the great arteries. The positive effect of a more lateral left coronary artery reimplantation on reducing coronary risk is widely accepted. The aim of this study was to evaluate the association between the parameters of the reimplantation and the diameters of the coronary arteries to help understand the development of coronary events more precisely.
Methods: The position of the ostium, the branching angle and the diameters of the coronary arteries were determined in 35 asymptomatic pediatric patients (age: 10.8 ± 3.1 years) with multiplanar reconstruction using GE Revolution CT scanner in a retrospective study.
Results: The reimplantation site of the left coronary arteries (LCA; left main coronary artery or left anterior descending artery arising from sinus #1 facing sinus) was closer to the main pulmonary artery (MPA) and to the plane of the neoaortic annulus compared to the right coronary arteries (RCA; arising from sinus #2 facing sinus) (p<0.01). The Z-score of proximal RCA diameters (1.3 ± 1.2) was significantly larger compared to the LCA (0.2 ± 1.3) (p<0.01). A closer position of the LCA ostium to the MPA caused a more acute angulation (p<0.01; r = 0.81). The Z-score of the proximal LCA diameter had a positive correlation with the distance from the MPA (p<0.01; r = 0.75) and the branching angle (p<0.01; r = 0.70). The Z-score of proximal LCA diameter and the Z-score of proximal left anterior descending artery diameter (0.5 ± 1.2) showed a significant, but weak correlation (p = 0.03; r = 0.39). A more anterior reimplantation of the RCA caused a greater branching angle (p<0.01; r = 0.83), but neither correlated with the diameter. The height of the reimplantations showed no correlation with the diameters.
Conclusion: Taking into consideration that only the LCA diameter correlated with the parameters of the reimplantation, and it was reimplanted closer to the MPA compared to the RCA, we assume that the proximity of the reimplantation to the MPA – especially an interarterial origin – has a greater influence on the LCA diameter than the branching angle. Besides the acute angulation, the reduced proximal LCA diameter could cause the alteration of blood flow leading to early coronary events.

Keywords: transposition of the great arteries, arterial switch operation, coronary computed tomography angiography

The association between the position of the LCA ostium and the proximal LCA diameter



The clockwise position of the LCA ostium is defined as the angle between the line connecting the center of the aorta and the MPA and the line connecting the center of the aorta and the LCA ostium. LCA: left coronary artery, MPA: main pulmonary artery

Other

P-370/Moderated Poster
Factors associated with acute kidney injury after cardiopulmonary bypass in children

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Background and Aim: Acute kidney injury (AKI) is a common complication after cardiovascular surgery in children, noted in ~40% in children undergoing cardiopulmonary bypass. We sought to determine clinical factors and inflammatory and vascular endothelial markers associated with AKI.
Methods: A prospective observational cohort study of pediatric patients with a cardiac defect (acquired or congenital) requiring cardiac surgery with pulmonary bypass (CPB) and >2.5 kg was performed. AKI was defined as a 1.5 times increase in serum creatinine or absolute increase by ≥0.3 mg/dL (≥26.5 umol/L). Plasma inflammatory markers (IL-1a, IL-1b, IL-2, IL-4, IL-6, IL-8, IL-10, IL- 12p70, TNFa) and vascular endothelial markers (VEGF, vWF, RANTES, GM-CSF, MCP-1, platelet derived growth factor, microparticles) were assessed at 6 perioperative timepoints (induction, 5 minutes into CPB, end of CPB, 1 day post-op, 3 days

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post-op and 5-10 days post-op), and associations with AKI were determined using generalized linear regression models adjusted for repeated measures.
Results: n = 207 patients, 56% (n = 116) male, were included, of whom 31% (n = 65) developed AKI. The median age in the AKI group was 0.5 years (IQR 0.2-1.2) vs. 0.4 years (IQR 0.1-2.7) in the non-AKI group. The mean CPB time in the AKI group was 157 +/- 74 minutes vs. 110 +/- 56 minutes in the non-AKI group. Paradoxically, lower pre-operative creatinine was significantly associated with AKI (30 vs 37 umol/L; p<0.001). In univariable analyses, adverse outcomes significantly related to the presence of AKI included increased length of an open chest (p = 0.04), increased ventilation time (p = 0.003), increased intensive care unit stay (p = 0.007) and increased overall hospital stay (p = 0.003). In multivariable analysis, independent factors that were significantly associated with AKI included increased number of previous catheterizations (p = 0.002), no pre-operative prostaglandins use (p = 0.01), longer cardiopulmonary bypass time (p = 0.002), higher initial post-operative aspartate transaminase (p<0.001), post-operative platelet-derived growth factor (p<0.05), post-operative chylothorax (p = 0.02) and neurologic complications (p = 0.003). Inflammatory and vascular markers were not significantly associated with AKI.
Conclusion: AKI remains a prevalent problem associated with increased complexity and injury in other organ systems and was not associated with inflammation or vascular markers. The presence of AKI was associated with worse clinical outcomes.

Keywords: Pediatrics, Cardiology, Nephrology, Acute Kidney Injury, Outcomes

Surgery and Intensive Care

P-371
Correction of tetralogy of fallot in an infant with cystic fibrosis, a case report

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Background and Aim: Tetralogy of Fallot (TF) is the most common cyanotic congenital heart disease (CHD). Sometimes it's hemodynamics is ductal-dependent. Cystic fibrosis (CF) is a rare, progressive genetic multisystem disorder, being the lung the most severely affected. The combination of TF and CF is extremely rare. Our aim is to demonstrate unique clinical case of such combination and successful correction of CHD. To our knowledge this is only the third such case reported.
Methods: 1,8 kg boy who was prenatally diagnosed with TOF was born on the 34 week of gestation at the local hospital. He became desaturated up to 60 % on the 2 day of life and was put on ventilator and on prostaglandins with saturation increasing to mid-80 %. Patient was transferred to CICU, where diagnosis of TOF with severe pulmonary stenosis was confirmed. He was weaned from ventilator, but stayed dependent from supplemental oxygen. The surgery was denied because of low weight. The combination of multiple infiltrates in the lung fields with late meconium led us to the assumption that the reason of hypoxemia was not only CHD. It was revealed that he passed his first meconium only on the fifth day of life, and the result of CF screening was positive. CF was diagnosed by measuring level of immunoreactive trypsinogen in patient's blood. Prostaglandins were cancelled with no desaturation. Genetic test revealed the CFTR gene mutation Del508, which confirmed CF. Patient received appropriate treatment with per oral pancreatic enzymes combined with preemie formula.
Results: Patient underwent successful correction of TF with transannular patch at the age 14 weeks after achieving weight of 3 kg. He was discharged from the hospital on the 34 day after surgery.
Conclusion: The reasons of hypoxemia in newborn babies with cyanotic CHD might be due not only to the heart, but to the lung problems as well. One must remain aware of possible combination of CHD with inheritable pulmonary diseases, which may alter the clinical management of patients with congenitally malformed hearts.

Keywords: Tetralogy of Fallot, cystic fibrosis, clinical case

P-372/Moderated Poster
Health-related quality of life of ugandan children following valve replacement surgery for rheumatic heart disease

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Background and Aim: Valve replacement surgery (VRS) is performed to reduce morbidity and mortality among patients with severe rheumatic heart disease. However, lifelong anticoagulation and frequent monitoring is required, which potentially impacts on health-related quality of life [HRQoL]. In this study, we assessed the HRQoL of children with rheumatic heart disease (RHD) in Uganda following VRS.
Methods: Between April and August 2021, we cross-sectionally recruited consecutive participants who had had VRS before the age of 18 years. A pre-tested, semi-structured questionnaire was used to collect data from the study participants. The Pediatric Quality of Life Inventory –Cardiac Module (PedsQL-Cardiac module) was used to evaluate HRQoL of the participants. The HRQoL score ranged from 0 – 100, with higher scores signifying better quality of life. We considered total mean of ≥ 80% as optimal HRQoL.
Results: A total of 83 eligible participants were analyzed. Of these, 52 (60.5%) were female, with a median age of 18 (IQR: 14-22) years. Most (n = 71, 89%) of the VRS were performed in Salam-Centre for Cardiac Surgery in Khartoum, Sudan. Sixty-one (72.6%) participants had a single mechanical valve replacement. Just over half of the participants (n = 45, 54%) reported they do not mind being on life-long warfarin therapy. Forty-one (50%) reported they could not afford regular lab tests. Seventy (87.5%) participants had full understanding of warfarin use, however, (29.3%) feared bleeding. The optimal mean score of cardiac-specific HRQoL was achieved in 50 (60.2%) participants. Almost all participants had class 1 NYHA functional classification, Optimal HRQoL was associated with normal body mass Index (BMI) (p = 0.001), and acceptance of the presence of an artificial valve in their body (p = 0.001).
Conclusion: In this study, over two-third of the participants had optimal HRQoL and almost all had class 1 NYHA functional status. Normal BMI and acceptance of the presence of an artificial valve in the body was significantly associated with optimal

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HRQoL. We identified the need for support in regular blood testing and monitoring by making free "self-test kits", available and introducing self-management system.

Keywords: Rheumatic Heart Disease, Valve Replacement Surgery, Health-Related Quality of Life, Uganda

P-373

Truncus arteriosus: address to truncal valve insufficiency

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Background and Aim: In truncus arteriosus, truncal valve (TV) insufficiency (regurgitation [TvR] and stenosis [TvS]) is one of the most important factors influencing the prognosis. Neonatal cases with severely dysplastic leaflets may still result in death. We have performed actively three leaflets reconstruction with autologous pericardium in addition to conventional procedures even in neonates with severely dysplastic leaflets. Here, we focus on our result of TV repair.

Methods: The data of 10 patients who underwent TV repair for TV insufficiency in our hospital between January 2000 and March 2021 were retrospectively investigated in terms of survival, surgical reintervention, and echocardiography findings.

Results: TV repair at primary total repair (TR) in neonate was performed in 7 patients (age, 2.0-12 days [median: 6]; weight, 2.1-3.6 kg [median: 2.5]). Three patients (1 with PAPVC and 2 with low body weight) underwent bilateral pulmonary banding; all patients received TV repair at TR (age, 4-17.0 months [median: 16]; body weight, 7.2-8.3 kg [median: 7.7]). In preoperative TV findings, quadricuspid was in 7, tricuspid in 2 and bicuspid in 1. The reason for the intervention was TvR in 6 patients, TvS in 1 patient, and TvR/TvS in 3 patients. Sever TV insufficiency with circulatory failure was observed in 7 cases (neonate: 6). The surgical technique used for valvuloplasty was: tricuspidization; 7 (Mee method; 2, leaflet extension; 2, commissuroplasty/annulus reduction; 1, commissurotomy; 1, homograft replacement; 1) and bicuspidizition; 1. Three leaflets reconstruction with autologous pericardium was performed for two neonates with sever dysplastic leaflets. The length of follow-up after TV repair was 0.2-116 months (median: 9). Two patients died perioperatively (Mee procedure and homograft in neonate). Nine patients had moderate to severe TvR, and no patient had TvS. Reintervention on TV was performed in 6 patients. The time from TV repair to the reintervention was 0.2-42 months (median: 7). A second TV repair was performed in all of the reintervention cases, but two of them later required mechanical valve replacement.

Conclusion: Advanced TV repair in neonate yielded favorable outcomes as a lifesaving approach. Three leaflets reconstruction for neonates might be effective as a life-saving procedure.

Keywords: truncus arteriosus truncal valve insufficiency truncal valve repair neonate three leaflets reconstruction

Characteristics of TV surgery patients

✓ Characteristics of concomitant TV surgery patients (n =10)

Patient	Reason for TV surgery	TV anatomy	TV size, mm	Timing of TV surgery	Age at repair , day	BW at repair , kg	TV surgery type	Time to TV reoperation , month	TV reoperation	Follow up time
1	TvSR	Quadricuspid	12.9	Primary TR	12	2.1	Homograft replacement	1	Konnno procedure, ATS16mm	Death (early)
2	TvR	Tricuspid	14.4	Primary TR	6	2.5	One leaflet extension	42	Leaflet extension	82
3	TvR	Quadricuspid	13	Primary TR	10	2.2	Mee method	–	none	116
4	TvSR	Quadricuspid	12	Primary TR	6	2.9	Mee method	0.2	Patch closure for leaflet dehiscence	Death (early)
5	TvS	Tricuspid	16.9	After BPAB	120	7.2	Commissuroplasty, annulus reduction	–	none	24
6	TvSR	Quadricuspid	16.2	After BPAB	480	7.7	Bicuspidiztion; Leaflet slicing, commisurotomy	9	Two leaflet extension	12
7	TvS	Bicuspid	17.2	After BPAB	510	8.3	Commisurotomy	–	none	4
8	TvR	Quadricuspid	9.2	Primary TR	2	2.6	Tricuspidization; two leaflet excision, one leaflet reconstruction	14	Two leaflet extension, one leaflet reconstruction	111
9	TvR	Quadricuspid	11.1	Primary TR	3	2.3	Three leaflet reconstruction	5	Re three leaflet reconstruction --Konno procedure, ATS 16mm	13
10	TvSR	Quadricuspid	12.1	Primary TR	2	3.6	Three leaflet reconstruction	–	none	8

— with sever circulatory failure

P-374

Effect of restenosis after coarctation repair on exercise performance and cardiac function in children

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Background and Aim: Restenosis is a common complication after surgical repair of aortic coarctation, often requiring additional treatment by balloon dilation. The effect of such event on later clinical performance of these children is poorly understood. This study aims to investigate the effect of post-coarctation restenosis on exercise and cardiac performance in children at teenager age.

Methods: Exercise testing and LV function and mass (echocardiography) were assessed in children after coarctation repair comparing a group who developed restenosis, treated successfully by a single balloon dilation (ReCoa-BD), and a group without restenosis (Coa). The groups were matched at a 3/1 ratio for age, gender and associated cardiac anomalies.

Results: Out of 210 children undergoing prior simple coarctation repair, and in whom a reliable exercise test is obtainable (age 6-18y), 43 children without restenosis were matched to 14 children who had a single balloon dilation for restenosis. The mean age was 12.4 ± 2.6y at the time of evaluation. Children of ReCoA-BD group were younger at the time of surgical repair: median 7.0d (IQR 4.5 – 11.5) versus 31.0d (IQR 11.0 – 182.0)($p = 0.002$). The transaortic pressure gradient at the CoA zone was 26.5 ± 13.1 mmHg and 27.8 ± 9.1 mmHg ($p = 0.730$) for the CoA and ReCoA-BD group. Exercise test results were comparable between Coa and ReCoa-BD: VO2 max: 38 ± 9 vs. 35 ± 7 ($p = 0.122$); O2 uptake efficiency: 1760 ± 548 - 1688 ± 567 ($p = 0.953$); VE/VCO2: 27 ± 4 - 28 ± 5 ($p = 0.817$). Exercise-related hypertension was observed in respectively 21% and 14% of the CoA and ReCoA-BD group ($p = 0.741$). Echocardiography revealed

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comparable systolic and diastolic function parameters between Coa and ReCoA-BD, as well as LV volumes and hypertrophy (LVEDD: 44.1 ± 6.2mm vs. 43.5 ± 4mm, p = 0.753; LVESD: 26.5 ± 4.3mm vs. Study 25.5 ± 3.8mm, p = 0.477; LVMi: 82.5 ± 31.9g/m² vs. 76.5 ± 34.5g/m², p = 0.661).
Conclusion: This study shows that the development of a restenosis after coarctation repair has no detrimental effect on exercise performance nor cardiac function, if this complication is a purely early anastomotic constriction which is then effectively treated by a single balloon dilation. Further investigation is needed for the effect of restenosis due to residual morphological aortic arch lesions.

Keywords: Coarctation - Balloon dilation - exercise performance - cardiac function

P-375

Effectiveness of intraoperative heparinization in neonatal and pediatric patients with congenital heart diseases: focus in heparin resistance

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Background and Aim: The purpose of this study was to determine the prevalence of heparin resistance among cardiac surgical pediatric and neonatal patients and to identify associated risk factors.
Methods: The study included 306 patients undergoing on-pump cardiac surgery for period from January to December 2018. Patients whose activated clotting time (ACT) targets were achieved after the first administration of heparin formed the 1st group (n = 280); the second group (n-26) included patients with heparin resistance. The initial assessment of the haemostasiological profile included determining the PT, aPPT, FG, AT III activity, INR. Intraoperative control of heparinization was carried out with definition of ACT using a kaolin activator. A weight-associated protocol at the rate of 300 U/kg with target values of ACT >480 sec was used for intraoperative heparinization.
Results: The heparin resistance was verified in 8.5% of patients included in the study. Repeated administration of heparin at the maximum total dose 600 U/kg required in 80.77% cases. In spite of additional heparinization, 19.23% of patients had fresh frozen plasma infusion. There was reduced antithrombin activity in the heparin resistance group (p = 0.01) (Fig.1). The most patients with heparin resistance (57.7%) were treated with low molecular weight heparins at the preoperative period.
Conclusion: The determining the initial level of antithrombin activity can serve as a predictor of the risk of developing heparin resistance. The factor analysis made it possible to verify latent risk factors for heparin resistance to the form heparin pretreatment, chronic hypoxia, and chronic heart failure (Fig.2.)

Keywords: heparin resistance, neonatal, heparin pretreatment, chronic heart failure, chronic hypoxia

P-377

Risk factors for acute kidney injury after heart surgery in children

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Background and Aim: Acute kidney injury (AKI) is an important issue after cardiac surgery in children, ranging from 3 to 42% of this population, increasing morbity and mortality rates. We aimed to identify risk factors related to AKI after cardiac surgery in children, and also to analyse the role of AKI in death outcome.
Methods: We conducted a 4-years retrospective study of children aged under 18 years-old, submitted to cardiac surgery in a reference center in a northern city in Brazil. We collected clinical and demographic data from the medical records. For the diagnosis of AKI, we used the KDIGO AKI classification, calculated during the the first seven days after the procedure. We excluded children with death in the operating room, previous dyalitic chronic renal disease.
Results: A total of 305 patients were analyzed. Median age was 14.6 months (0.1- 209); median weight was 7.6 Kg (0.8- 52). 70% of the patients were RACHS 1 and 2. The frequency of AKI was 36.4%; 17% of patients were classified as AKI KDIGO 1; 8% KDIGO 2 and 10% KDIGO 3. Dialytic treatment ocurred in 5% of the cases. After multivariate logistic regression, low weight was a risk factor for AKI ($p<0.05$). Lenght of cardiopulmonary bypass was a risk factor for dialytic support and death ($p<0.05$)
Conclusion: Acute kidney injury frequency was 36.4%. After multivariate logistic regression, low weight was a risk factor for AKI. The lenght of cardiopulmonary bypass was a risk factor for the need of dialytic support and death.

Keywords: congenital heart disease, acute kidney injury

P-378

Neointimal hyperplasia in systemic-to-pulmonary shunts of children with complex cyanotic congenital heart disease

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Background and Aim: Neointimal hyperplasia might play a role in systemic-to-pulmonary shunt failure contributing to interstage morbidity in infants with single ventricle physiology after stage I Norwood procedure or children with other complex cyanotic congenital heart disease. The aim of this study was to elucidate histopathologic changes in systemic-to-pulmonary polytetrafluorethylene (PTFE) shunts and to determine whether increased neointimal hyperplasia is associated with early interventions such as balloon angioplasty, stenting or shunt revision. Further, we intended to identify risk factors for increased neointimal proliferation and shunt stenosis in affected infants.
Methods: Shunts removed at stage II palliation or other surgical procedures were processed for histopathological analysis. Slides were stained with Hematoxylin/Eosin, Richardson and immunohistochemistry was performed with alpha-smooth muscle-actin (α-SMA) and CD68 to identify vascular smooth muscle cells, myofibroblasts, and macrophages. Images were digitalized for quantification of histopathologic parameters. Univariate and multivariate analysis were performed to identify clinical risk factors contributing to neointimal hyperplasia and shunt stenosis.

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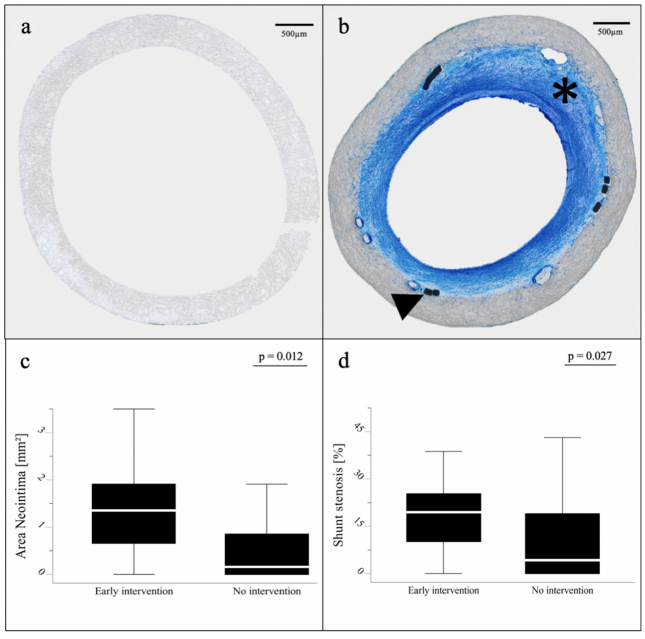
Results: Fifty-seven shunts from 55 patients (39 modified Blalock-Taussig anastomosis, 8 right-ventricle to pulmonary-artery anastomosis, 10 central shunts) were analyzed. The area of neointimal proliferation within the shunt was in median 0.75 mm2 [range, 0.03 - 3.65 mm2] and relative shunt stenosis in median 16.71 % [range, 0.17 - 65.82 %]. Thrombi, PTFE infiltration, α-SMA and CD68 were measured with 0.1 mm2 [range, 0.01-1.06 mm2], 1.86 mm2 [range, 0.14-6.56 mm2], 0.26 mm2 [range, 0.03-1.25 mm2] and 0.05 mm2 [range, 0.001-0.43 mm2], respectively. Neointimal hyperplasia and shunt stenosis correlated with each other and were significantly greater in the group that required early interventions comprising balloon dilatation, stent implantation and shunt revision. Multivariate regression identified shunt size and lower aspirin dosage as independent risk factors for greater neointimal proliferation and shunt stenosis.
Conclusion: Neointimal hyperplasia occurs frequently in systemic-to-pulmonary shunts and is associated with early interventions comprising balloon dilatation, stent implantation and shunt revision. Smaller shunt size and lower aspirin dosage are independent risk factors for increased neointimal proliferation. Pharmacological targeting could decrease interstage morbidity and the risk of shunt-failure in children with complex structural heart disease.

Keywords: Neointimal hyperplasia, Systemic-to-pulmonary-shunt, Shunt-Malfunction, Norwood Procedures, Hypoplastic-left-heart syndrome, Cyanotic heart defects

Neointimal hyperplasia and shunt stenosis in the early intervention group



A Histological comparison of cross sections from a shunt of the non-intervention group vs. a shunt of the early intervention group is shown in panel (a & b). Panel (a) shows a shunt without any neointimal formation in the lumen as well as no infiltration of cells inside the PTFE biomaterial. Panel (b) demonstrates a systemic-to-pulmonary shunt in which malfunction required a stent implantation. The star [⋆] depicts neointimal hyperplasia causing severe shunt stenosis; [triangle] points to a stent strut; staining: Richardson; scalebar: 500 µm. Panel (c & d) demonstrates boxplots showing the distribution of neointimal hyperplasia and relative shunt stenosis between the groups: Early intervention vs. No intervention. Early intervention included: Balloon angioplasty; stent implantation and shunt revision (Mann-Whitney-U test).

P-379

Long-term use of right atrial lines in postoperative pediatric cardiac patients

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Background and Aim: The postoperative management of neonatal and pediatric patients undergoing cardiac surgery has evolved considerably over the last few decades. Right atrial lines (RAL) are widely used in some institutions for limited periods of time as a routine part of postoperative care. In single ventricle patients, these lines play a significant role in preservation of vessels that are a part of palliative pathways. Despite their generalized use, there is limited data on factors associated with their complications. The aim of our study was to characterize the long-term use of RAL in pediatric cardiac patients and to identify factors associated with RAL complications in this population.
Methods: Observational retrospective cohort study in pediatric cardiac patients who underwent RAL placement during cardiac surgery in a children's hospital from January 2011 through June 2018.
Results: A total of 692 patients underwent 815 RAL placements during cardiothoracic surgery between January 2011 and June 2018. Median age and weight were 22 days (IQR 7-134) and 3.6 kg (IQR 3.1-5.3). Neonates accounted for 53.5% and single ventricle physiology for 35.4%. More than one third (38%) underwent palliation surgeries (shunts and cavo-pulmonary connections). Survival to hospital discharge reached 95.5%. Median RAL duration was 11 days (IQR 7-19) with a median RAL removal to hospital discharge time of 0 days (IQR 0-3). Thrombosis and migration were the most prevalent complications (1.7% each), followed by malfunction (1.4%) and infection (0.7%). Adverse events associated with these complications were seen in 12 (1.4%) of these RAL placements: anemia requiring transfusion (n = 1), tamponade requiring pericardiocentesis (n = 3), pleural effusion requiring chest tube placement (n = 2) and need for antimicrobials for bacteremia (n = 6). Multivariable logistic regression showed that RAL duration (OR 1.01, p 0.006) and palliation surgery (OR 2.38, p 0.015) were independently associated with complications.
Conclusion: The prolonged use of RAL seems to be feasible and safe. Our overall incidence of complications from long-term use remained similar to the reported with short-term use of these lines. While line duration and palliation surgeries were associated with complications, severity of illness could be a confounding factor. A prospective assessment of RAL complications may improve outcomes in this fragile population.

Keywords: right atrial line, neonates, children, pediatric cardiac surgery, complications

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Table: Multivariable logistic regression

Fully adjusted multivariable logistic regression to ascertain for factors associated with RAL complications

Predictor	OR (95% Confidence Interval)	p-value
Year	1.14 (0.95 - 1.37)	0.161
Age Child vs Neonate	1.67 (0.24 - 11.57)	0.768
Age Infant vs Neonate	1.64 (0.71 - 3.83)	0.611
Weight	0.99 (0.88 - 1.11)	0.822
Prematurity (No vs Yes)	0.54 (0.22 - 1.3)	0.166
Syndrome/association (No vs Yes)	1.48 (0.64 - 3.42)	0.361
Non-cardiac abnormalities (No vs Yes)	1.03 (0.44 - 2.42)	0.954
Genetic diagnosis (No vs Yes)	0.81 (0.29 - 2.27)	0.691
Surgical palliation (No vs Yes)	2.42 (0.82 - 7.14)	0.108
Shunt (No vs Yes)	0.61 (0.23 - 1.58)	0.304
RAL type double lumen vs triple lumen	3.99 (0.51 - 31.29)	0.595
RAL type single lumen vs triple lumen	7.06 (0.35 - 140.94)	0.303
Right atrial line duration (days)	**1.01 (1 - 1.02)**	**0.008**
Single ventricle physiology (No vs Yes)	0.86 (0.33 - 2.27)	0.765

Final model after backwards variable selection to identify factors associated with RAL complications

Predictor	OR (95% Confidence Interval)	p-value
Surgical palliation (Yes vs No)	2.38 (1.18-4.81)	**0.015**
Right atrial line duration (days)	1.01 (1-1.02)	**0.006**

*OR: odds ratio, RAL: right atrium line

P-380

Mortality and morbidity in patients with hypoplastic left heart syndrome (HLHS) related to risk factors

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Background and Aim: It was our aim to show whether well known risk factors for mortality after Norwood procedure have a predictive value for the morbidity and health related quality of life.
Methods: Retrospective analysis of 137 patients with HLHS who underwent Norwood procedure between 2000 and 2018 in our center. The statistical analysis was done with Kaplan-Meier survival analysis, Fisher's exact Test and Wilcoxon-Mann-Whitney-Test. The measurement of health related quality of life after Fontan-operation was done with PedsQL™Tool.
Results: We confirm the known preexisting risk factors for mortality after Norwood procedure (i.e. low birth weight, hypoplastic ascending aorta, syndromic disease, etc.). Overall survival of patients with risk factors is 40% in contrast to 69% in those without risk factors (p = 0,02). There is a significant difference of mortality after Norwood-1 procedure in patients with risk factors in comparison to those without (48% vs. 20%; p = 0,05). Interestingly we did not find differences in the need of mechanical ventilation (10,1 vs. 10,9 days), the need of inotropic support (12,5 vs. 12,4 days) or the development of a capillary leak syndrome (34% vs. 36%). Otherwise the need of ECMO in patients with risk was 22% in contrast to 5% in patients without risk. Fortunately there was no significant difference between the two groups after Norwood-2 in mortality and morbidity, for example concerning the number of cardiac catheterization (3,6 vs. 4,3), or total days of hospital stay until Norwood-3 operation (123 vs. 115) etc.The health related quality of life appears to be similar.
Conclusion: If the patient survives step 1 of Norwood procedure there is no relation between preexisting risk factors and further mortality, morbidity or quality of life.

Keywords: hypoplastic left heart syndrome

P-381

Injection-jet self-powered fontan circulation to effectively drop caval pressure in a failing fontan

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Background and Aim: The Fontan circulation is a fragile system in which imperfections at any one of multiple levels may compromise quality of life, produce secondary pathophysiology, and shorten life span. Elevated inferior vena caval (IVC) pressure plays a key role in "Fontan failure". We hypothesize that the Fontan circulation can be energized with an injection jet shunt (IJS) drawing flow directly from the aortic arch balanced by a fenestration. The IJS causes flow entrainment, leading to a clinically significant IVC pressure reduction of >3mmHg. We describe a tightly coupled multi-scale lumped parameter/computational fluids dynamics (LPM-CFD) model to validate this hypothesis.
Methods: A synthetic 3D-CAD model of the fenestrated total cavopulmonary connection (TCPC) was generated, with average dimensions matching those of a 2-4 yo patient. The prescribed cardiac output is of about 2.3L/min with a body surface area of 0.675m^2. Hemodynamics are modeled as unsteady, turbulent, constant density and blood is assumed non-Newtonian. Turbulence is approximated using a large eddy simulation (LES) approach. Potential optimal IJS configurations were determined through a parametric sweep of several geometric design parameters such as TCPC morphology, shunt and fenestration diameter and location (Figure 1). The effects of the IJS implementation on IVC pressure and systemic oxygen saturation were calculated.
Results: A set of baseline simulations representing a failing Fontan with elevated IVC pressure (+17.8 mmHg) is shown in Table 1 (cases 1-2 a). Fenestration enlargement to 7 mm (cases 1-2 b) results in a 3 mm Hg IVC pressure drop but also significant reduction in systemic oxygen saturation. Addition of an IJS (2mm nozzle) to this model preserves the IVC pressure drop of 3.2 mm Hg and improves systemic oxygen saturation with only a small additional volume load to the ventricle (CO/Qs = 1.2) (cases 3-4).
Conclusion: Our current models demonstrate the potential salutary effect of the IJS on the Fontan circulation. Further exploration of adjustable anatomic parameters is currently underway to find configurations that further reduce IVC pressure and improve systemic oxygen saturation. In-vitro simulations will also be considered to cross-validate the optimal outcome from the in-silico model.

Keywords: Congenital Heart Defect, Fontan, Computation Fluid Dynamics

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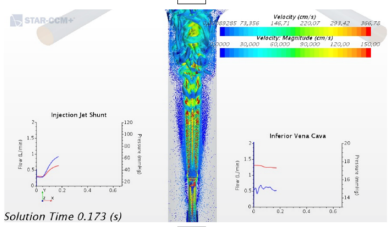
Sample Simulation Results

Case #	Model	Qp [L/min]	Qs [L/min]	CO [L/min]	Qp/Qs	CO/Qs	Pivc [mmHg]	Psvc [mmHg]	Plpa [mmHg]	Prpa [mmHg]	ΔPivc [mmHg]	sysO2 [%]	venO2 [%]
1a	4mmfen	2.049	2.268	2.268	0.904	1.000	17.827	17.792	17.012	17.008	-	93	73
2a	2Y-4mmfen	2.057	2.274	2.274	0.905	1.000	17.742	17.780	17.058	17.073	0.1	93	73
1b	7mmfen	1.305	2.431	2.431	0.537	1.000	14.693	14.662	14.221	14.234	3.1	76	57
2b	2Y-7mmfen	1.326	2.426	2.426	0.546	1.000	14.701	14.741	14.347	14.354	3.1	77	58
3	2Y-7cmivc-2mmijs-8mmfen	1.321	2.213	2.641	0.599	1.197	14.648	14.932	14.428	14.657	3.2	81	61
4	2Y-7cmivc-2mmijs-7mmfen	1.381	2.213	2.639	0.625	1.195	14.908	15.224	14.684	14.972	2.9	82	62

A



B

Figure 1 – (A) Summary table of baseline (4mm fenestration), enlarged fenestration (7mm fenestration) and fenestration with IJS configuration and (B) lambda 2 vorticity isosurfaces colored by velocity magnitude generated and propagating from the shear layers between the confined high-speed jet and the background IVC co-flow.

P-382
Outcomes of cardiac surgery patients requiring prolonged ICU admission

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Background and Aim: The average length of stay in the intensive care unit (ICU) following cardiac surgery is 4 days. A subset of these patients require prolonged ICU admission. This cohort require high levels of resources and have a higher risk of mortality. We record and analyse this group within our single tertiary ICU over a 6 year period.
Methods: From 2013 to 2019, 99 of 2937 (3.3%) congenital cardiac patients who underwent surgery required an ICU stay lasting >28 days. We examined factors associated with ICU mortality within this group.
Results: ICU mortality occurred in 22 of 99 patients (22.2%). A further 6 (6.1%) patients passed away in the hospital and 4 (4%) post discharge. Factors associated with mortality were moderate to extreme prematurity (gestation < 34 weeks) (odds ratio (OR), 3.75 [95% confidence interval (CI), 1.11 - 12.68], P = 0.03) and Trisomy 21 (T21) (OR 3.1, [CI, 1.07 - 8.98], P = 0.04). Other factors, including atrioventricular septal defect (AVSD) (OR, 2.57 [CI, 0.91 - 7.3], P = 0.08) and pulmonary hypertension (OR, 2.03 [CI, 0.66 - 6.24], P = 0.22) suggested increased mortality risk, however, these did not meet significance. Our data also suggests that AVSD patients with T21 (OR, 1.68 [CI, 0.46 - 6.08], P = 0.43) may perform better than those without T21 (OR, 3.2 [CI, 0.78 - 13.14], P = 0.11). Interestingly, our data indicates that single ventricle physiology (OR, 0.41 [CI, 0.09 - 1.97], P = 0.27), extracorporeal membrane oxygenation (ECMO) (OR, 0.6 [CI, 0.16 - 2.29], P = 0.46) and delayed sternal closure (DSC) (OR, 0.53 [CI, 0.19 - 1.5], P = 0.23) suggest reduced mortality risk, however, once again these did not meet significance and may be explained by the fact that T21 and premature patients are usually not candidates for single ventricle repair or ECMO.
Conclusion: Cardiac surgery patients who require prolonged ICU stay have high risk of mortality. Those who are at least moderately premature or have T21 are particularly at risk. Certain factors such as single ventricle physiology may counterintuitively predict more favourable outcomes in this group.

Keywords: cardiac, surgery, ICU, Prolonged, Prematurity, Trisomy21

P-383
A single centre review of management and outcomes after vascular ring surgery

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Background and Aim: Vascular rings are often associated with aortic arch abnormalities. In our series patients present more often with symptoms due to oesophageal compression than upper airway compression. Surgical intervention for vascular ring relief has a low complication rate and survival is greater than 99.7%. Whilst most patients will have resolution of their symptoms, in some symptoms persist. In about 5% to 10% recurrence of symptoms occurs with a small proportion requiring further surgery.
In our centre these patients have input from multiple disciplines including cardiology, SALT, ENT and gastroenterology.
Methods: This was a 5 year retrospective study of patients who have undergone vascular ring surgery in a single centre. Data was collected using Electronic Patient Records.
Results: Over a 5 year period (2016-2021) 32 patients underwent vascular ring surgery. One underwent repeat surgery having had a previous vascular ring relief 9 years earlier. 12 patients were diagnosed with a potential for vascular ring antenatally. preoperatively 6 patients had airway symptoms and the remainder feeding difficulties. Preoperative investigations included: CT scan (all patients), barium swallow (n = 5) and MLTB (n = 13). 5 patients had early post-operative complications: 4 pleural effusions/chylothorax and 1 postoperative cardiac arrest and required ECMO. At time of discharge over half of the patients (19/32) had ongoing symptoms: 10 had stridor, 14 had swallowing and feeding difficulties requiring ongoing SALT input; 5 had both swallowing difficulties and stridor. The outpatient follow up period varied considerably with 6 patients being discharged from follow up. The shortest post-operative follow up period was 3 months.
Conclusion: Vascular ring release surgery is a highly successful operation with a low complication rate. The management of patients who have undergone vascular ring surgery in our centre involves multiple teams. There is variation in practice in the current preoperative investigations and post-operative follow-up. We propose producing a guideline for uniform investigation practice before surgery and a structured multi-disciplinary follow-up.

Keywords: vascular ring, double aortic arch, right aortic arch

P-384
Perioperative viral pneumonia in infants with complex congenital heart disease after partial CAVO-pulmonary connection (PCPC)

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Background and Aim: The stage II correction in the univentricular approach is the PCPC. Due to the passive lung perfusion the oxygenation is dependent of a low resistance.

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Methods: Between 2015 and 2020 we included 79 infants after PCPC-surgery. We observed 6 cases of viral pneumonia with severe outcome. Aim was to compare these patients in two groups (n = 6/79) regarding perioperative and outcome parameters.
Results: The blood gas analysis, central venous pressure (mean 19 mm Hg, sd ± 5 und mean 19 mm Hg, sd ± 6; p = 0,733), CRP values (mean 37mg/dl, sd ± 21 und mean 42mg/dl, sd ± 24; p = 0,791) and ventilation parameters intraoperatively did not vary in both groups. In the control group the extubation took place after 9 hours (mean 522 min, sd ± 111 min). The invasive ventilation time in the pneumonia group continued 23 days in average (sd ± 26 d, p = 0,02). 4 infants died with their pneumonia, all patients in the control group survived (66,7 % vs. 0 %; p = 0,03).
Conclusion: The onset of a viral pneumonia in the early postoperative phase after PCPC is associated with a serious outcome. Further investigation should point out whether preventive measures could have an impact on risk improvement.

Keywords: univentricular, PCPC, pulmonary, ventilation

P-385
Outcomes of anomalous left coronary artery from pulmonary valve repair: impact of adjustable pulmonary artery band

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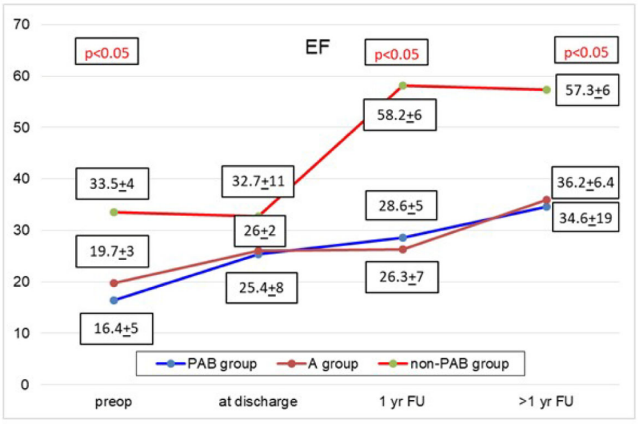
Background and Aim: Anomalous left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital anomaly resulting in myocardial ischemia, global left ventricular (LV) dysfunction, and mitral regurgitation (MR). Surgical placement of a pulmonary artery band (PAB) at ALCAPA repair may assist with recovery of select patients with severe left ventricular function (EF <25%). The purpose of this review was to assess the outcomes of these patients.
Methods: We performed a retrospective analysis of cases presenting to our center between January 2009 and December 2021. A pulmonary artery band was placed on patients with EF<25%. Baseline and follow-up data were collected from records, ventricular function was assessed by standardized transthoracic echo.
Results: 26 patients underwent ALCAPA repair at median age of 4 months (range, 19 days to 8 years). Surgery was performed by direct coronary reimplantation in all patients. Since 2017, we used an adjustable PAB in five patients with severe LV dysfunction (<25% EF). No patients required mitral valve repair at time of ALCAPA surgery. Two patients with EF<25% from the non-PAB group were supported by extracorporeal membrane oxygenation (ECMO) with median duration of 3.5 ECMO days. In patients with EF<25% that received PAB, none needed ECMO or transplant. The mean duration of postoperative follow-up was 5.6+3.6 years (range, 6 months to 11 years). There were no early or late deaths, and two patients, both from non-PAB group required reoperation: mitral valve repair with coronary artery augmentation (n = 1) and a heart transplant (n = 1). Figure 1 shows comparisons between patients with and without PAB and EF <25% compared to patient without PAB and EF>25%.
Conclusion: Surgical repair of ALCAPA has good outcomes with low mortality and reintervention rates. There is marked improvement of MR grade, return to normal LV function, and no mortality during a 10-year period in the group with EF>25%. Patients who present with EF<25% demonstrate a slower trajectory towards improvement with or without a PAB. Temporary adjustable PAB may be beneficial in patients with severe dysfunction as evidenced by no need for ECMO or transplantation in the PAB group despite severe dysfunction, however numbers are still small for major conclusions.

Keywords: ALCAPA, Pulmonary Artery Band, Heart Failure, Echo, Surgery

EF in ALCAPA



Timeline of EF recovery after surgical ALCAPA Repair

P-386
Pronostic factors of mortality and morbidity after aortopulmonary shunt anastomosis in pediatric patients with cyanotic congenital heart disease

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Background and Aim: The objective of this study was to assess the results of aortopulmonary shunt (APS) surgery and evaluate rates and risk factors for mortality and morbidity.
Methods: Retrospective analysis of all APS performed from 2009 to 2016 in patients with unrepaired cyanotic CHD. Demographics, clinical and biological data, surgical techniques, echocardiographic measurements, and outcomes were collected.
Results: Overall 91 patients (43 females) underwent surgery for APS at a median age of 9 days (1 to 744) and median weight of 3.15kg (2.2 to 8.8). Underlying CHD included: tetralogy of Fallot (30%), transposition of great arteries with ventricular septal defect and pulmonary stenosis (20%), single ventricle (25%), pulmonary atresia (14%) and double outlet right ventricle (6.5%). Preoperative prostaglandin infusion was required in 77% of the cases. Distal shunt was performed in 75 and central shunt in 16, In-hospital mortality was 13.2% and did not decrease over time. Shunt thrombosis occurred in 11% of the cases, stenosis in 8% and 60% experienced increased pulmonary blood flow and low cardiac ouput. Risk factors of in-hospital death were: metabolic acidosis at 4-hour postoperative (p = 0.02), with lactates > 3mmol/L (p = 0.043), occurrence of necrotizing enterocolitis (p = 0.04) and presence of pulmonary branch stenosis (left or right branch or both) in the early postoperative course after surgery (0.0392).
Conclusion: Metabolic acidosis at 4-hour postoperative, pulmonary branch stenosis and necrotizing enterocolitis are risk factors of mortality after aortopulmonary anastomosis. `

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Keywords: cyanotic congenital heart disease, neonate, prognosis, aortopulmonary anastomosis

P-387

The prevalence of tracheal oesophageal dysfunction associated with anomalies of the aortic arch: a retrospective review

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Background and Aim: Vascular rings cause tracheo-oesophageal compression presenting as aerodigestive symptoms. This study investigated whether therapeutic aortic arch surgical intervention alone is truly curative by detecting any persistence of aerodigestive sequelae.
Methods: Data were retrospectively collected for all aortic arch anomalies referred for surgery at our institution between 2005-2020. Patients were identified and categorised into a vascular ring or complex arch anomaly. Post- operative follow-up included a symptom-focused questionnaire and the most relevant outpatient clinic letters. Data were analysed with SPSS statistics.
Results: Our series included 117 patients; 80 (68.4%) had complex arch anomalies and no aero-digestive symptoms, 37 (31.6%) had a vascular ring and severe aero-digestive symptoms. We had an 81.2% response rate to the questionnaire. Minimal aerodigestive complications were observed in 5 complex arch anomalies (6.25%), but 16 patients with vascular rings (43.2%) had persistent aerodigestive symptoms at follow up. The mean length of follow up was 3.25 years.
Conclusion: Our questionnaire was highly successful, indicating a potential unmet clinical need and highlighted a previously unknown problem. Based on our questionnaire, we demonstrated that surgery on complex aortic arches is safe in patients with a regular tracheooesophageal bundle. However, vascular surgery alone does not seem to address all the abnormalities and ongoing symptoms in those with demonstrated preoperative tracheooesophageal problems associated with vascular rings. We conclude that the abnormalities of the aerodigestive structures may be unrelated to the technical validity of the surgical solutions but to coexisting tracheal oesophageal abnormalities. Interdisciplinary approaches on the trachea, oesophagus, and aorta rather than the aorta alone might be warranted at diagnosis to inform a more lesion-specific surgical approach.

Keywords: Vascular Rings Aerodigestive Complex aortic arches

P-388

Children who developed acute kidney injury after pediatric cardiac surgery have persistent markers of renal injury at mid- and long-term follow-up

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Background and Aim: Acute kidney injury (AKI) in the immediate postoperative period is common, occurring in 30-60% of children undergoing pediatric cardiac surgery for congenital heart disease (CHD). Longer-term outcomes and the incidence of chronic kidney disease (CKD) after AKI are not well known. This is an important issue, as CKD occurs in up to 30-50% of the growing ACHD population, being responsible for an excess burden in healthcare utilization. This prospective cohort study investigated the renal consequences of AKI at mid- and long-term follow-up.
Methods: All eligible children (<16 years) who had developed AKI following cardiac surgery at our tertiary referral hospital were prospectively invited for a formal renal assessment approximately 5 years after AKI, including measurements of estimated glomerular filtration rate (eGFR), proteinuria, alpha-1-microglobulin, blood pressure, and renal ultrasound. Longer-term follow-up data on renal function were collected at latest available visit.
Results: Among 571 patients operated over a 4-year period, AKI occurred in 113 (19.7%). Fifteen of these (13.3%) died at a median of 31 days (interquartile range, IQR 9-57) after surgery. A total of 66 patients participated in the renal assessment at a median of 4.8 years (IQR 3.9-5.7) after the index AKI episode. Thirty-nine patients (59.1%) had at least one marker of renal injury, including eGFR <90mL/min/1.73m^2 in 9 (13.6%), proteinuria in 27 (40.9%), alpha-1-microglobinuria in 5 (7.6%), hypertension in 13 (19.7%), and abnormalities on renal ultrasound in 9 (13.6%). CKD stages 1-5 was present in 18 (27.3%). CKD was associated with syndromes (55.6% vs 20.8%, $p = 0.015$). At 13.1 years (IQR 11.2-14.0) follow-up, eGFR <90mL/min/1.73m^2 was present in 18/49 patients (36.7%), suggesting an average eGFR decline rate of -1.81 mL/min/1.73m^2 per year.
Conclusion: Children who developed AKI after pediatric cardiac surgery show persistent markers of renal injury. Detection and treatment of CKD, proteinuria, and hypertension in children is critical because these are risk factors for cardiovascular diseases and progressive renal damage in adults. We therefore suggest a structured renal follow-up in this vulnerable population and recommend that prevention of renal disease should start in childhood to ensure optimal outcomes in the growing population of ACHD.

Keywords: acute kidney injury, cardiac surgery, children, chronic kidney disease, congenital heart disease, long-term outcomes

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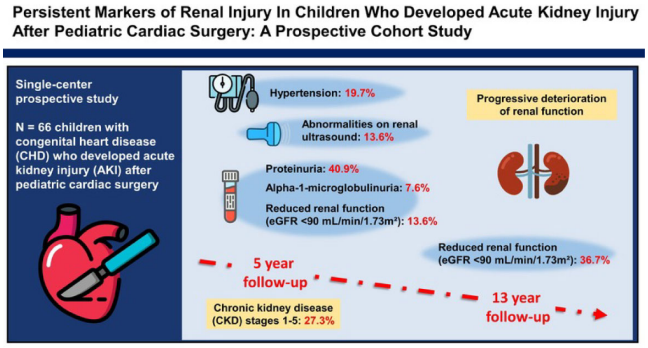
Central image summarizing the main findings of the study



P-389

Cone repair of tricuspid valve in ebstein's anomaly: 10 – years' experience of anatomical reconstruction

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[1]Ukrainian Children's Cardiac Center; [2]National Medical Academy of Postgraduate Education named after P.L Shupyk

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. Because of this, a part of the right ventricle (RV) becomes part of the atrium (atrialized) and dilates, and the leaflets and chordal apparatus of the TV become abnormal and dysplastic.
Objective. To analyze 10 – years' experience of successful cone reconstruction of TV in Ebstein's anomaly.
Methods: From 2011 to 2021 in Ukrainian Children's Cardiac Center 44 patients were operated with Ebstein's anomaly. The mean age of patients was 6.6 + 3.4 years. The smallest patient at the time of surgery was at least 4 days old. Preoperative diagnosis was made by the anatomy of the tricuspid valve: type "A" in 5 patients, type "B" in 15 patients, type "C" in 19 patients, type "D" in 5 patients. Electrophysiological study was performed in 28 patients, during which 23 additional atrioventricular connections were identified and radiofrequency catheter ablation was performed. MRI was performed in 42 patients. 10 patients had cone reconstruction combined with the Glenn shunt because of right ventricle failure.
Results: Early postoperative mortality was 6.8% (3 patients). Average postoperative follow-up was 7.3 + 1.8 years. During the follow-up visit the clinical condition of the patients was examined, tests with exercises stress were conducted to assess functional status, evaluated data of ECG, echocardiography, chest X-Ray and magnetic - resonance imaging of the heart. As a result of control there is a from mild to moderate insufficiency on the TV, contractile function of the left ventricle is good and mild decreased right ventricle's contractility, indicating good result of correction. There was not ATH late death. In late postoperative period reoperation was performed in 5 (12.1%) patients through a severe tricuspid insufficiency.
Conclusion: Methods of cone reconstruction of TV is a very effective technique of surgical anatomical treatment of Ebstein's anomaly with good early and late results.

Keywords: Ebstein's anomaly, Cone repair

P-391

Swiss evaluation registry for pediatric infective endocarditis (serpie) the right ventricle to pulmonary artery (RV-PA) conduit under fire!

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Background and Aim: In children, infective endocarditis (IE) is a rare but severe cardiac disease and information on etiology and management are scarce. This study aimed to analyse the epidemiology of pediatric IE in Switzerland.
Methods: Retrospective nationwide multicenter data analysis regarding frequency, microbiological spectrum, diagnostics, predisposing risk factors, clinical course, complications, therapy and outcome of cases of pediatric IE in children (<18 years) between 2011 and 2020 treated at the four main pediatric heart centers in Switzerland.
Results: Sixty-nine patients (27 male; 39%) were treated for IE. 37 patients (54%) fulfilled the modified Duke criteria (Li JS et al., 2000) for definite, 30 (43%) for possible, and 2 (3%) for suspicious IE. IE was diagnosed at median 6.39 years (IQR 11.79), with 19 patients (28%) during infancy (<1 year). Diagnosis was made at 6.50 days (10.75) after first symptom. Fifty-eight children (84%) had congenital heart defects (CHD), 42 (61%) qualified for antibiotic prophylaxis. IE primarily affected pulmonary (29;42%), mitral (10;14%), tricuspid (8;12%) and aortic valve (6;9%), ventricular septal defect (VSD) (4;6%), atrial septal defect (ASD) (1;1%), or non-localized (12;17%) Forty-eight patients (70%) had postoperative IE, with prosthetic valve IE in 33 (33/48;69%). Prosthetic valve IE affected frequently the right ventricular to pulmonary artery conduit (RV-PA conduit) (28/48;58%) with Contegra (21), Melody (5), Homograft (1), Shelhigh (1), or prosthetic material in VSD position (4/48;8%) VSD-Patch (3), Amplatzer-VSD-Device (1), and in ASD position (1/48;2) Amplatzer-ASD-Device (1/48;2%). Pathogens were staphylococci (27;39%) including staphylococcus aureus (19;28%), streptococci (14;20%), enterococci (8;12%), HACEK (6;9%), or otherwise culture negative (5;7%). Severe complications occurred in 35 patients (51%) including embolism (19;28%) to the lung (11), brain (9), spleen (3), kidney (2), extremities (2), or retina (1), sepsis (17;25%), and heart failure (16;23%). Thirty-two patients (46%) needed cardiac surgery with in twenty patients performed early (<28days). Five patients (7%) died.
Conclusion: IE is still a severe cardiac disease in childhood with relevant mortality and morbidity due to a high complication rate. Diagnosis is challenging with only about half of the patients

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meeting the Duke criteria for definitive IE. CHD represents the predisposing risk factor for IE, with a high number of IE associated with prosthetic pulmonary valve affection determining further valuation and therapeutic alternatives.

Keywords: Infective endocarditis, conduit, valve, sepsis, prosthesis, emboli

P-395

Mid-term outcome of valve-sparing repair for tetralogy of fallot

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Background and Aim: The mainstream of Tetralogy of Fallot (ToF) repair has changed to preserve own pulmonary valve from the transannular patch repair. However, the outcomes of this approach are not well known. In this study, we assessed mid-term pulmonary valve function in ToF patients who underwent valve-sparing repair(VSR).

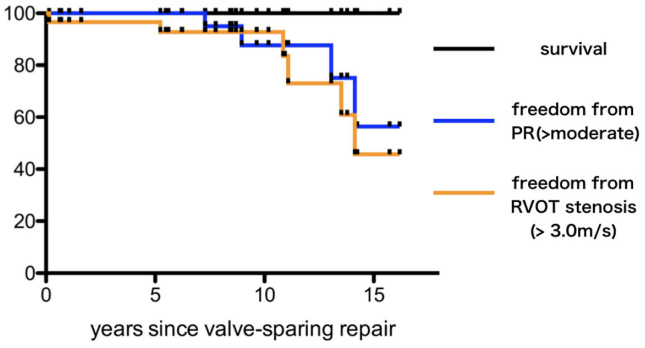
Methods: Consecutive 30 patients with ToF, who underwent VSR in our institute from September 2005 to November 2021, were included in this study. Regardless of pulmonary valve morphology, VSR was indicated when a Hagar dilator larger than 80% of the normal pulmonary annulus passes after performing commissurotomy and infundibular muscle resection. Incised pulmonary trunk and RVOT were closed with separate pericardial patches. All operations were supervised by only one surgeon. After VSR, pulmonary valve function was followed by echo cardiography.

Results: Eighteen patients underwent single stage correction, while staged correction following palliation was performed in 12. Before VSR, median pulmonary valve Z scores by echocardiography were -1.8 (-3.2 to 0.93). Median age and weight at VSR were 1.2(0.7-1.9) years and 8.4(6.4-9.9) kg. In macroscopic findings, valve morphology was tricuspid in 4 patients, bicuspid in 23 (transverse split:9, longitudinal split:14) and monocuspid in 1 (unknown: 2). As concomitant procedures, PA plasty beyond the bifurcation(2), pulmonary root reconstruction(1) and repair of complete AVSD(1) were performed.

Median operation time, cardiopulmonary bypass time and aorta cross clamp time was 314(237-458) min, 189(131-256) min and 132(86-166) min, respectively. Median follow-up period after VSR was 8.5 (0.14-16.2) years. There were no mortalities during the study period. Freedom from re-operations for the pulmonary valve was 100%, although the latest echocardiogram, which were performed 8.0(0.11-15.7) years after VSR, demonstrated significant pulmonary regurgitation (≧moderate) and RVOT stenosis (≧3.0 m/s) in 4(13%) and 6 (20%) patients, respectively.

Conclusion: Mid-term outcome of VSR for ToF was satisfactory. By this procedure, pulmonary valve function was maintained for more than 8 years in patients having small pulmonary annulus.

Keywords: Tetraloy of Fallot, pulmonary valve, valve sparing repair



P-397

Correlation of pulmonary arterial development and shortterm postoperative outcome in pediatric patients with univentricular heart based on angiography

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Background and Aim: During the second stage of the three-stage surgical palliation of patients with univentricular hearts the circulation of pulmonary arteries is replaced by a non-pulsating, low-pressure flow, which has a significant effect on the development of pulmonary arteries (PA). The aim of this study is to assess the effect of Glenn surgery on pulmonary arterial development and it's on short-term postoperative outcomes.

Methods: We examined the parameters obtained from catheter angiography scans of 40 children born between 2008 and 2018, 131 (1-505) days before Glenn surgery and 248 (63-929) days before the total cavopulmonary connection (TCPC) was completed, as well as their relationship with data of postoperative hospitalization.

Results: Z-score (0.8 ± 1.3) of the diameter of the left pulmonary artery (LPA) before Glenn was significantly higher than the Z-score of the right pulmonary artery (RPA) (-0.6 ± 2.0) ($p<0.01$). Between the two measurements, the LPA Z-score decreased by an average of 2.1 ± 1.0, while the RPA Z-score decreased by 0.6 ± 1.4, thus equalizing the difference in diameter. Glenn anastomosis position was not related to lack of LPA growth. For both PA's, negative correlation was found between the initial Z-score and the percentage increase in diameter ($p<0.01$). The increase in diameter is only significant in PA with a starting diameter of Z-score<0 ($p<0.01$). The Nakata index was 280 (101-699) before Glenn and 187 (75-506) before TCPC. Length of stay in postoperative intensive care unit and required circulatory and respiratory support, did not show any correlation with the PA parameters.

Conclusion: According to our data Glenn flow is not able to maintain the disproportionately larger pre-Glenn LPA diameter. However, it cannot be ruled out that the right location of

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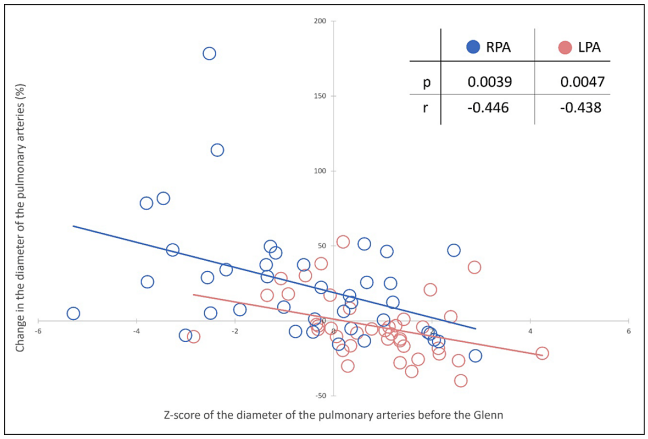
Glenn anastomosis also plays a role in the greater development of RPA. Based on our results, the large diameter difference between the two PA's, poor development of LPA and lower preoperative Nakata indices did not result in worse short-term outcomes. Further examination is needed to determine the cause of the difference in diameter between the two PA's prior to Glenn.

Keywords: univentricular heart, pediatric cardiology, congenital heart disease

The effect of Glenn surgery on pulmonary arterial development



The lower the Z-score of the diameter of both pulmonary arteries before Glenn the more significant their development after the Glenn procedure. RPA: right pulmonary artery, LPA: left pulmonary artery

Other

P-398

Early experience with myocarditis and pericarditis in adolescents following vaccination against COVID-19 in latvia

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Background and Aim: The European Medicines Agency has approved mRNA vaccines developed by Pfizer/BioNTech and Moderna for the vaccination of adolescents against the SARS-CoV-2 infection. Cases of myocarditis and pericarditis have been described as rare postvaccination complications. We describe the Latvian experience with adolescents suffering from myocarditis following COVID-19 vaccination.
Methods: From June to December 2021 four cases consistent with postvaccination myocarditis were admitted to the Children's Clinical University Hospital, which is the only centre with specialized paediatric cardiology care in Latvia. The Pfizer/BioNTech vaccine had been used in all. An ECG, Holter monitoring and ECHO was done, HS Troponin I levels checked, the most common infectious causes of myocarditis were excluded, and a cardiac MRI was performed in all cases.
Results: Case 1: 12-year-old girl, developed chest pain on postvaccination day Nr 4 (PVD4) after the 1st dose. Holter monitoring revealed rare non-sustained ventricular tachycardia (NSVT), ECHO showed moderate mitral insufficiency, and a hyperechogenic papillary muscle, troponin level peaked at 5339 pg/ml (PVD6), MRI (PVD 7) showed widespread myocardial oedema, transmural fibrosis. Symptoms resolved in 1 day, metoprolol succinate and lisinopril were prescribed. Mitral insufficiency persists 5 months later. Case 2: 15-year-old boy, developed chest pain after the 2nd dose on PVD2 and lasted for 7 days, he was admitted on PVD11 with a peak troponin level of 19pg/ml. MRI (PVD15) showed widespread myocardial oedema. Metoprolol succinate was prescribed. Case 3: 15-year-old boy, developed chest pains on the day of the 1st dose and persisted for 35 days, he was admitted on PVD24 with peak troponin level 15ng/ml. MRI (PVD29) showed mild myocardial oedema, myocardial and pericardial fibrosis. Case 4: 13-year-old boy, developed chest pain on PVD2, which lasted for 65 days, he had an episode of syncope. Holter monitoring showed frequent PVCs, and NSVT, on PVD34 troponin level was 2,5pg/ml. The child received a course of NSAIDs and was referred to us on PVD68. MRI (PVD69) revealed widespread myocardial oedema, fibrosis, and pericarditis. Methylprednisolone was given, and betaxolol was prescribed.
Conclusion: Our case series show that some cases of postvaccination myocarditis develop complications requiring long-term treatment.
Keywords: vaccination, COVID-19, myocarditis, pericarditis, mRNA

P-399

What is the role of sildenafil in patients with single-ventricle anatomy prior to fontan surgery?

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Background and Aim: Sildenafil is frequently used to treat patients with single-ventricle anatomy and increased pulmonary artery pressure. The aim of our study was to analyze the influence of sildenafil on the hemodynamic parameters of patients undergoing cardiac catheterization prior to Fontan surgery.
Methods: 106 patients after the Glenn procedure who had cardiac catheterization prior to the Fontan surgery between January 2013 and July 2019 were enrolled in the study. Of this group, 46 (43.4%) patients were treated with sildenafil. This study compares the characteristics of patients treated with sildenafil versus non-treated.
Results: It was found that patients who were taking sildenafil were more likely to undergo cardiac catheterization prior to Fontan surgery ($p = 0.0003$) and more likely to require interventional procedures ($p = 0.00056$), in particular dilatation of pulmonary arteries stenosis ($p = 0.0046$) and closure of collateral arteries ($p = 0.004$). Patients who were taking sildenafil had higher pulmonary pressures (mPAp; 14.48 mmHg ± 2.681 vs 12.57 mmHg ± 3.26; $p = 0.0017$) and higher pulmonary capillary wedge pressure (mPCWp; 11.76 mmHg ± 2.626 vs 10, 35 mmHg ± 2.94; $p = 0.012$) than those who were not taking this medication. However, in both groups the pulmonary resistance was comparable. Patients who were treated with sildenafil had a significantly higher cardiac index and lower systemic resistance compared to those who did not receive this drug (CI; 5.96

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l/min/m2 ± 2.103 vs 4.56 l/min/m2 ± 1.53; p = 0.000167 and SVRi; 9.55 WU × m2 ± 3.275 vs 12.43 WU × m2 ± 4.74; p = 0.000661).
Conclusion: The use of sildenafil alone may be insufficient to improve the hemodynamic parameters of pulmonary circulation in patients prior to Fontana surgery. We speculate that sildenafil may have a different mechanism of action and a greater effect on the systemic circulation in patients with single-ventricle anatomy than in patients with normal anatomy.

Keywords: sildenafil, single-ventricle anatomy, Fontan procedure, Glenn procedure

P-400
Sudden cardiac death in children cardiomyopathy – a french study

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Background and Aim: Cardiomyopathy in children is a rare and heterogeneous disease in which sudden cardiac death (SCD) is difficult to predict. Recent pediatric studies have proposed models to predict SCD in hypertrophic cardiomyopathy (HCM).
Methods: We conducted a retrospective and multicenter study in 11 medical centers in the North of France from 2009 to 2019. Our objectives were to describe patients presenting severe arrhythmias in dilated cardiomyopathy (DCM) and HCM, to highlight risk factors and to calculate the Norrish et al. HCM Risk-Kids score in HCM patients. We included 224 children with cardiomyopathy (diagnosis ≤18 years): 72 HCM, 69 DMC, and 84 other types.
Results: Among DMC patients, event-free survival (recovered SCD or heart transplant) was 70% at 5 years. We report only 3 recovered SCD: one in a patient with laminopathy, one in a patient with Barth syndrome and one in a context of acute heart failure with important mitral regurgitation. Among HCM patients, event-free survival was 82% at 5 years. We report 11 SCD. HCM Risk-Kids score was calculated at 6.2% on average. An HCM Risk-kids score ≥6% was significantly associated with SCD risk (p = 0.015). But for a threshold of 6%, the Positive Predictive Value of the score was only 27% and the Negative Predictive Value of 95%. Among the 21 patients implanted with an implantable cardiac defibrillator (ICD) for primary prevention in whom the HCM Risk-Kids score could be calculated: 4 had a score <4%, 7 a score between 4 and 6% and 9 a score ≥6%. The only patient who used his ICD had a score of 9.4%.
Conclusion: In DCM, apart from very specific etiologies (laminopathy, Barth, ischemic), the indication of ICD in primary prevention must be rigorously discussed on a case-by-case basis because of a very low risk of SCD. Concerning the HCM Risk-Kids score, even if this score has the merit of existing, its low positive predictive value will conduct to a high rate of ICD implantation in pediatric patients putting them at risk of potential complications. This score should probably be coupled with patient's genetic pattern and MRI data to improve risk stratification.

Keywords: Sudden cardiac death, children cardiomyopathy, hypertrophic cardiomyopathy, dilated cardiomyopathy

P-401
Effects of COVID 19 pandemic on children attending to emergency units with chest pain

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Background and Aim: Chest pain is a one of the most common complaints in children admitted to the Hospitals. Although it was among the most common reasons for referral to the pediatric cardiologist before COVID-19 era, this tendency is changed during covid pandemic. The primary objective of this study was to investigate the aetiological causes, clinical characteristics and the follow up symptoms in terms of changing habits of parents and children admitted to the ED for acute chest pain during pandemic.
Methods: We reviewed the medical records of children under the age of 18 who presented with chest pain as the chief complaint from 1 January 2020 to 1 April 2021, at Istanbul University – Cerrahpasa Pediatric Emergency Clinic retrospectively from the hospital database. The study population comprised 128 boys and 119 girls.
Results: All the children underwent ECG examination. While the ECG results of 239 children were normal, 6 sinus tachycardia, 1 supraventricular tachycardia and 1 incomplete left bundle branch block were observed. 33 patients had an echocardiography. Eight patients with an abnormal Echo result already had been examined and diagnosed prior to their emergency admission. Blood samples were taken from 48 children for troponin. 17 samples were above the cut-off value which was set to 0.004 g/dl A total of 32 SARS-Cov2 swab samples were taken from suspicious cases and analyzed with RT-PCR. Consequently, 8 of these children were Covid-19 positive. 7 patients had no history of chronic disease, while 1 patient had ALL. All of these patients had mild symptoms and none of them required hospitalization. The total number of children who were referred to a pediatric cardiologist for a further examination together with the follow-up patients of the pediatric cardiology department is 52
Conclusion: In conclusion, Chest pain is a common referral complaint in children and is rarely due to cardiac diseases. To date cardiac reasons of chest pain was the major concern of patients and families attending the ED. However we found that Patient/family concerns regarding 'vulnerability to the severe covid infection has emerged as an important discourse during the pandemic on attendences to ED because of chest pain.

Keywords: chest pain, children, Covid 19

P-402
Common arterial trunk with patent ductus arteriosus, a rare diagnosis from fetal life

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Background and Aim: In common arterial trunk (CAT), the involution of the ductus arteriosus is the most frequent finding.

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Truncus with a patent ductus arteriosus (PDA) and a normal aortic arch is very rare. Their coexistence is very rare, with only a few cases reported.
Methods: We present a rare case in which a TAC with a PDA was prenatally diagnosed and confirmed postnatally by imaging and by direct inspection during surgery.
Results: We present a fetus who was diagnosed at 35 weeks of gestation with TAC and a PDA. Genetic study was normal. A male newborn was born at 38+6 weeks with 3120 grams. Postnatal echocardiography, and computed tomography angiography confirmed the diagnosis of CAT with the presence of PDA, which did not present spontaneous closure in successive controls. Truncal valve was dysplastic and quadricuspid, with mild regurgitation. Differential diagnosis by echocardiography included pulmonary atresia with ventricular septal defect, and aorto-pulmonary window. The clue to the diagnosis was the presence of a dysplastic truncal valve. Corrective surgery was performed at 7 days of life. Intraoperatively, the presence of PDA was evidenced, which was ligated in the same surgical procedure. The patient was discharged 10 days after surgery. At 4-month follow-up, the patient did not present coarctation of the aorta and also presented adequate neurological development.
Conclusion: The simultaneous presence of PDA and CAT has been reported with extreme rarity, with only a few cases described in the literature. Prenatally diagnosis of this rare anomaly can be done. It is important to the echocardiographer to bear in mind the possibility of the presence of both TAC and PDA, and make the differential diagnosis mainly with aorto-pulmonary window.

Keywords: PDA, CAT, common arterial trunk, patent ductus arteriosus

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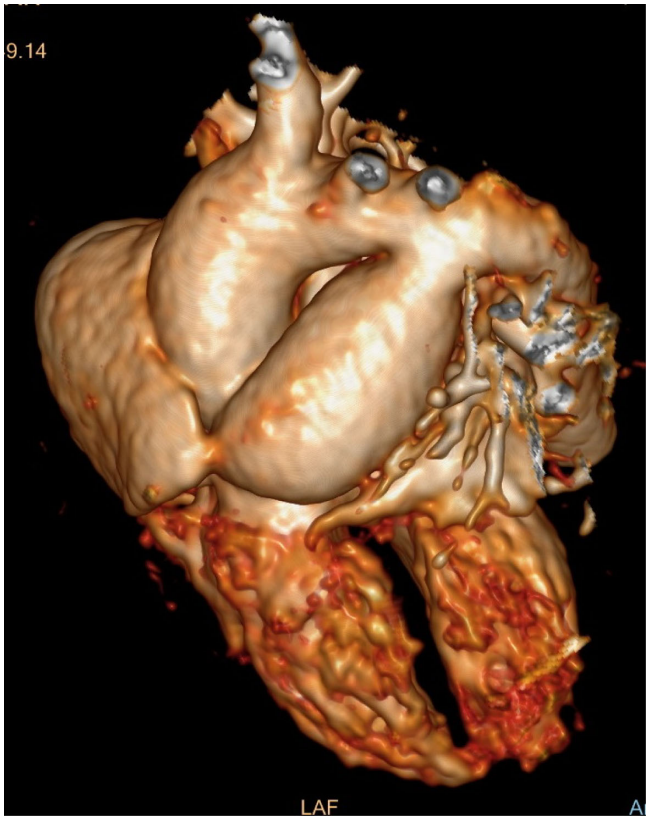

P-403
Correlation between conventional echocardiography and myocardial deformation imaging in the assessment of patients with coronary artery aneurysms

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Background and Aim: The aim of this study was to assess global and regional LV myocardial function and to predict the long-term prognosis of Kawasaki diseases.
Methods: We analysed 62 children (mean age 9,4 ±3,9 years), 8 of them with a residual coronary aneurism and 20 healthy children (mean age 9,6 ±4,8). Apical and basal short axis for 2D images were acquired (65±7 frames/s) in addition to apical four, three and two-chamber views. Global and regional peak systolic strain - longitudinal, radial and circumferential on the LV was derived and the strain curves (eLL, eCC, eRR) were extracted using a commercial software.
Results: In the measurements of LV function, there was a clear tendency toward a decrease in the ejection fraction (65,9 ± 4,1vs. 71,9± 3,2) with a concomitant increase in LV diastolic diameter (LVDD) (z-score 0.7±1,0 vs.-0,3±0,8). Global longitudinal strain measurements of the LV were non-significantly different between the Kawasaki group without an aneurysm and normals (GLS -19,23±1,98% vs -21,73±0.92 %, p=0.03). The regional peak systolic longitudinal strain (LS) were reduced significantly, especially on the basal and middle LV - segments 2,8,14 on anteroseptal wall and 3,9 on inferoseptal wall, as well as the global longitudinal systolic strain. In the group - Kawasaki with an aneurysm was found significantly reduced GLS compared with normal patients -17,94±1,69 % vs -21,73±0.92 %. (p<0,001).
Conclusion: Decreased segmental GLS values are a sign of regional hypokinesia with a possible local segmental ischemia in the group with coronary aneurysms on coronary artery and are a marker for developing severe LV dysfunction in the future. The contribution in this study was the introduction of a new non-invasive assessment of myocardial function after BC, despite the "normal" global systolic heart function.

Keywords: Kawasaki disese, Myocardial deformation.

P-404
Cardiovascular manifestation in multisystem inflammatory syndrome in children temporally associated with COVID-19-single center experience

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Background and Aim: Cardiovascular manifestations are common (35–100%) in multisystem inflammatory syndrome in children (MIS-C), including ventricular dysfunction, shock, coronary artery dilation, pericardial effusion and conduction abnormalities. Our study aimed to analyse cardiovascular involvement in our patients with MIS-C treated in our hospital.
Methods: The retrospective cohort study included all patients with MIS-C treated from April 2020 to December 2021 in the Mother and Child Health Institute of Serbia. In every case, cardiovascular manifestations were analysed: ventricular dysfunction, coronary artery dilatation, pericardial effusion, shock and ECG changes.

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Results: The study included 77 patients, 45 boys and 32 girls, average years of age 9.3 ± 4.8. Elevated cardiac troponin I and pro-BNP were observed in 35.9% and 87.8% of patients, respectively. Myocardial dysfunction was observed in half of our patients (50.6%), with an average ejection fraction of 50.5 ± 8.9%. Children older than 10 years had 4 times higher chances for myocardial dysfunction (OR 4.3, 95%CI 1.6-10.8; p = 0.003). Shock syndrome had 21.1% of children on admission, while 5.3% developed shock during the in-hospital stay. Transient coronary artery (CA) dilatation was observed in 6.5% of patients; left CA in 3 pts (Z score +2,95 ± 0.3), right CA in one patient (Z score +2), and in one LCA and RCA (RCA Z score 2.6). Transient CA dilatations were observed only in patients with KD-like clinical presentation (5/54 pts). Mild pericardial effusion with spontaneous resolution was detected in 28.6% of children, while one female adolescent had severe pericardial effusion with threatening cardiac tamponade. On the standard ECG, 53% of children had negative T wave in inferior or/and precordial leads averagely on day 2 (IQR 1-3 day); transient QTc prolongation was registered in 46% of patients, averagely on day 7 (IQR 5-9). Sinus bradycardia and coronary rhythm were registered in 42.1% of patients, while premature ventricular beats were observed in 2.7% of pts. left ventricle thrombus was detected in one patient with normal echocardiography finding. In this patient, increased activity of Factor VIII and XII was proven.
Conclusion: Cardiac manifestations are common and potentially life-threatening in MIS-C and should be assessed for at presentation and during the clinical course as indicated.

Keywords: multisystem inflammatory syndrome in children, ventricular dysfunction, coronary artery dilatation, pericardial effusion, shock, ECG changes.

P-405/Moderated Poster

Pediatric cardiology fellowship programs in countries with limited resources: achievements and challenges

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Background and Aim: Pediatric cardiology (PC) is a rapidly advancing specialty that needs advanced technical resources. A high level of training is needed for prompt diagnosis and management of pediatric/congenital heart disease. In a low-income setting, establishing and maintaining such programs is challenging. We describe our experience with establishing a training program for PC in Sudan, a country with population of 42 million and poverty rate of 65%.
Methods: Narrative report of Sudan PC Fellowship Program
Results: In 2012 a Pediatric Cardiology Fellowship Program accredited by the Sudan Medical Specialization Board was established. The program is 3- years and has a structured curriculum which consists of didactic lectures, clinical training, echocardiography and cardiac catheterization training as well as pre and post cardiac surgery management. In addition, participation in research projects is required to complete the program. The local trainers are only 3 which posed a huge challenge on the program. To overcome this, collaboration with visiting trainers, online education and complementary training abroad enabled the program to continue despite shortages. Online training as well as examination were utilized without major difficulties. From 2012-2022, 15 candidates graduated and 6 are on training. 2 candidates were sent abroad for interventional catheterization training. The program significantly promoted the PC services in the country (Figure)
Conclusion: PC training programs can be established utilizing scanty resources through online training and evaluation.

Keywords: Pediatric cardiology, fellowship, developing countries

P-407

Features and prognostic factors of scimitar syndrome in children

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Background and Aim: The aim of this study was to assess the features of children with Scimitar syndrom and to determine factors for long term of outcome
Methods: This is a retrospective two-centers study of all patients less than 18 years diagnosed with isolated SS. Demographic data were collected, echocardiographic measurements at diagnosis and last follow up, hemodynamic data when available. Prognostic factors for survival and bad outcome were analyzed.
Results: 52 patients (35 from Lyon center, France and 17 from Padova, Italy). Patients presented (median age 4.9 months) with respiratory symptoms in 48% or heart failure in 6% or were asymptomatic in 46%. Mean Z-scores for RV diameter, LV diameter, right PA and left PA branch were respectively +0.79, -0.6, -1.9 and +2.2. Mean pulmonary systolic pressure was 59mmHg: 52.5% of cases had no or mild PHT and 47.5% moderate or severe PHT. Fifteen patients were operated on, 26 received medical treatment and 9 percutaneous embolization of systemic artery were performed at a median age of 0.6years. Nine deaths occurred (17%) at median age of 0.4years: mortality was 35.7% in neonates, 16.6% in infants and 5.5% in children. Median FU was 13.4years. Survival rates were 87% at 6months, 85% at 1 and 5years and 78% at 12years of FU.
Stenosis of the scimitar vein, neonatal onset, symptoms at onset, systemic artery and moderate/severe PHT were associated with worse survival ($p<0.0001$). RV systolic pressure, PA systolic, dastolic and mean pressure were higher (respectively: 70 vs 39.6, 61.9 vs 33.3, 22.7 vs 13.5 and 38.1 vs 20.5mmHg), and LV was lower (20.5 vs 29.9mm) in deceased cases than in survivors ($p<0.0001$).
Conclusion: Overall outcomes of children with Scimitar syndrome is favourable except in cases with very early onset symptoms and/or moderate/severe PHT and/or stenosis of the scimitar vein.

Keywords: scimitar syndrome, children, prognosis

P-408

Total cavopulmonary anastomosis in patients with univentricular heart disease: results, and prognosis by age at surgery

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Background and Aim: The optimal age at which to perform total cavopulmonary bypass (TCDC) is a subject still debated. The objectives of this study were to evaluate the characteristics of the patients who underwent a DCPT in our center, to study the postoperative results, and to analyze the impact of the age at the time of the surgery on the long-term morbidity and mortality.
Methods: All patients who underwent DCPT at our center between 1981 and 2017 were included, with retrospective collection of demographic, clinical, hemodynamic, and long-term event data. Patients were divided into 4 groups according to their age at the time of DCPT: under 3 years (<3y), 3 to 6 years (3-6y), 6 to 18 years (6-18y) and over 18 years (≥18y).

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Results: 203 patients were analyzed, 12 in the <3y group, 56 in the 3-6y, 108 in the 6-18y and 20 in the ≥18y. Median age at DCPT was 7.6 years (72.4% extracardiac DCPT), and decreased from 9.3 years before 2007 to 6 years after 2007. Overall survival was 93.4% at a median follow-up of 11.4 years (to 36.7 years). Seven patients died and 7 were transplanted. The incidence of wet enteropathy was 5.9%, arrhythmia 19.6%, and prolonged pleuropericardial effusions 11.3%. Ventricular systolic function was impaired in 21.6% of patients. In multivariate analysis, exsudative enteropathy was the only risk factor for death and/or transplantation ($p = 0.04$). Morbidity and mortality were associated with extreme ages. Patients in the ≥18y group had the highest rate of heart transplant (10.7%), exsudative enteropathy (10.7%), prolonged effusions (27.8%), arrhythmias (33, 3%) and systolic ventricular dysfunction (33.3%). Patients in the <3y group had a significantly longer mechanical ventilation and intensive care stay than the other groups, highest mortality rate(8.3%) and highest incidence of exsudative enteropathy (9.1%), prolonged effusions (18.2%), infections (62.5%), and chylous effusions (25%). The best results were observed in the 3 to 6 year old age group.
Conclusion: Post-DCPT survival is favorable, and age at the time of surgery has decreased. Extreme ages are associated with increased long-term morbidity and mortality. The most favorable results were observed in patients aged 3 to 6 years.

Keywords: cavopulmonary anastomosis, univentricular heart, children, age groups

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The large bifurcation angle of left coronary artery influences the development of coronary aneurysm associated with kawasaki disease

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Background and Aim: Coronary arterial lesions (CALs) associated with Kawasaki disease (KD) frequently develops at the bifurcation of coronary arteries; however, there is little information regarding the relationship between underlying coronary geometry and the development of CALs. This study aimed to explore the relationship between branching angles of the coronary arteries and the susceptibility of CALs.
Methods: We reviewed echocardiographic data in patients with KD who were admitted to our hospital between 2017 and 2020, and measured the branching angles between the left main trunk (LMT), the left descending artery (LAD), and the circumflex artery (LCX). The branching angles were compared between patients with and without CAL.
Results: A total of 91 patients were studied. There were 9 patients (9.6%) with CALs, including the left side CALs in 8 patients (8.5%). Thirteen patients (14.3%) were identified as non-responders for intravenous immunoglobin therapy (IVIG). The IVIG non-responder ratio was higher in patients with CALs (50.0% vs 10.8%, $p = 0.010$). The angle between LAD and LCX in the patients with CALs was significantly greater than that in patients without CALs (95.7 ± 16.1 vs 62.8 ± 13.5 degrees; $p<0.001$), whereas the angle between LMT and LCX in patients with CALs was significantly smaller than that in patients without CALs (109.9 ± 20.3 vs 146.4 ± 14.2 degrees; $p<0.001$). The logistic regression analysis showed the development of CALs was significantly associated with the greater branching angle between LAD and LCX (Odds ratio, 1.16; 95% confidence interval, 1.003−1.444; $p = 0.045$).
Conclusion: The branching angle of the left coronary arteries, especially the angle between LAD and LCX, can be a previously unrecognized risk for the development of CALs associated with KD, which suggests that the hemodynamic verification of the coronary artery as well as the control of inflammation is important in the management of patients with CAL.

Keywords: Kawasaki disease, coronary artery lesions

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The hidden risk of kawasaki disease: the case of an 11-year old boy with myocardial infarction without evidence of coronary artery aneurysm

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Background and Aim: Myocardial infarction is the principal cause of mortality in children with Kawasaki disease (KD) and occurs most frequently among patients with giant coronary artery aneurysms. However, endothelial dysfunction, an underrated feature in KD, has been observed in patients without coronary artery aneurysm during the acute stage of KD. So far, there is speculation about the impact of this observation.
Methods: We present the case of an 11- year old boy with acute ST-elevation myocardial infarction (MI) as first cardiac symptom of suspected KD. The general symptoms had started two months prior with arthritis, conjunctivitis and scleritis, exanthema, cervical lymphadenopathy and fever >10 days, but had not been recognized as KD and thus, was not treated with immunoglobuline or ASA. The echocardiogram demonstrated normal coronary arteries. There was no past or family history of cardiovascular disease. After out-of-hospital cardiac arrest with successful cardiopulmonary resuscitation, percutaneous transluminal angioplasty of the left anterior descending coronary artery (LAD) was performed. There was no coronary artery aneurysm (CAA).
Results: Initially severely diminished myocardial function recovered throughout follow up (ejection fraction 50% after two years), but hypokinetic deformation is still detectable in the anterior, infero-septal, and antero-septal regions. In cardiopulmonary exercise testing O2pulse as a surrogate parameter for stroke volume improved. There was no worsening of regional wall motion abnormalities during stress echocardiogram. Preserved coronary blood flow to the LAD was demonstrated with coronary angiogram two months after the initial event. However, optic coherence tomography (OTC) was able to show concentric fibrotic intima thickening leading to a 60% stenosis at bifurcation LAD/D1. This continued into an excentric intima thickening further distally. Again, no CAA was seen. Extensive workup for other causes of vasculitis was done with no pathologic result.
Conclusion: Autopsy studies of deaths occurring one month after onset of KD describe progressive neointimal proliferation and neoangiogenesis. Normally, these changes are associated with the formation of aneurysms. Our patient had unusual findings in the OCT with endothelial dysfunction, but no coronary artery aneurysm. Thus, even without visible CAA there may be a risk for myocardial infarction after KD.

Keywords: Myocardial infarction, Kawasaki disease, endothelial dysfunction, coronary artery intima thickening

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Optical coherence tomography (OCT)

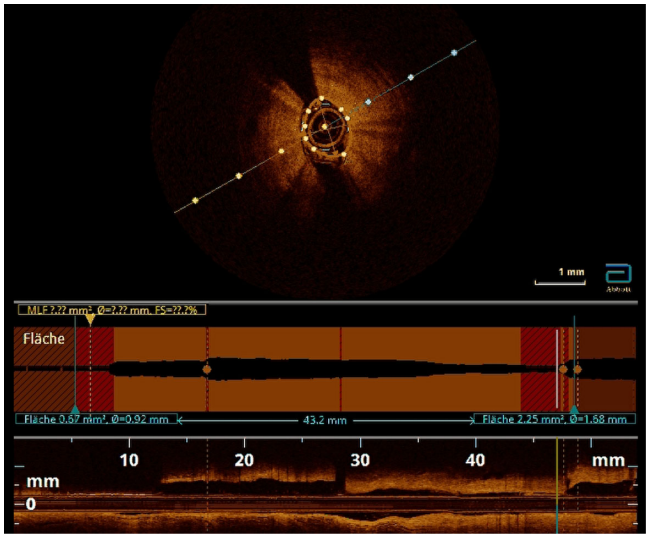

Image demonstrates concentric intima thickening of the left ascending coronary artery at level of stenosis

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Kawasaki disease Arab initiative [kawarabi]: establishment and results of a multicenter survey

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Background and Aim: Studies on Kawasaki disease (KD) in Arab countries are scarce, often providing incomplete data. This long with the benefits of multicenter research collaboratives led to the creation of the KD Arab Initiative [Kawarabi] consortium. An anonymous survey was completed among potential collaborative Arab medical institutions to assess burden of KD in those countries and resources available to physicians.
Methods: An online 32-item survey was distributed to participating institutions after conducting face validity. One survey per institution was collected.
Results: Nineteen physicians from 12 countries completed the survey representing 19 out of 20 institutions (response rate of 95%). Fifteen (79%) institutions referred to the 2017 American Heart Association guidelines when managing a patient with KD. Intravenous immunoglobulin (IVIG) is not readily available at 2 institutions (11%) yet available in the country. In one center (5%) IVIG is imported on-demand. The knowledge and awareness among countries' general population was graded (0 to 10) at median/interquartiles (IQR) 3 (2 – 5) and at median/IQR 7 (6– 8) in the medical community outside their institution.
Conclusion: Practice variations in KD management and treatment across Arab countries require solid proactive collaboration. The low awareness and knowledge estimates about KD among the general population contrasted with a high level among the medical community. The Kawarabi collaborative will offer a platform to assess disease burden of KD, among Arab population, decrease practice variation and foster population-based knowledge.

Keywords: Kawasaki disease, Arab, multicenter collaborative, registry, cardiac complications, survey

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Critical congenital heart defects in the newborn period: prevalence and clinical presentation

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Background and Aim: Critical congenital heart defects (CCHD) comprise 20 to 25% of all CHD. Delayed diagnosis of CCHD is associated with worse preoperative condition and less favorable surgery outcomes. Our study aimed to assess the prevalence and time of presentation of critical CHD.
Methods: We performed a retrospective analysis of the newborn babies who were admitted to our clinic in critical condition during the period January 2015-November 2021.
Results: During this 7-year period, a total of 265 neonates presented with critical hypoxemia, acute heart failure, or cardiogenic shock. The distribution of the diagnoses was as follows: coarctation of the aorta (CoA) - n = 77 (29%), transposition of the great arteries (TGA) – n = 64 (24%), single ventricle (SV) and complex CHD - n = 53 (20%), pulmonary atresia (PAtr) - n = 25 (9%), critical aortic stenosis (AoSt) - n = 18 (7%), critical pulmonary valve stenosis (PSt)- n = 15 (6%), hypoplastic left heart syndrome (HLHS) - n = 13 (5%). The mean age at admission for the newborns with left-sided obstructive lesions was 8.28 days (1-25 days) for CoA, 6 days for HLHS (2-15 days), 4.5 days for AoSt (1-11 days). Cyanotic heart defects presented at a much earlier age – mean

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age 0.46 days (0.04-1 day) day for TGA, 2.6 days for PAtr (0-4 days). The mean age at presentation was significantly higher for left-sided obstructive lesions compared to cyanotic CHD ($p<0.0001$). Of note, 21/77 (27%) of the newborns with CoA were diagnosed prenatally, and the remaining two-thirds had late diagnosis. 19/77 (25%) of the babies with CoA remained unrecognized and were discharged home where they deteriorated and were admitted to the hospital in critical condition. The remaining 37/77 (48%) of the newborns with CoA were diagnosed by a longer stay at the neonatology unit because of comorbidities.
Conclusion: Critical left-sided obstructive lesions in the newborn period present later when most babies are already discharged home, which poses them at much higher risk for an unfavorable outcome, compared to children who develop symptoms while still at the hospital or who have a prenatal diagnosis. Our findings underscore the extreme importance of broad neonatal pulse-oximetry screening for timely detection of duct-dependent CHD.

Keywords: critical congenital heart defect, newborns

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Needs assessment for an executive functions intervention in children with congenital heart disease after cardiopulmonary bypass surgery: e-fit study

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Background and Aim: Many children with severe congenital heart disease (CHD) after cardiopulmonary bypass surgery face academic challenges during adolescence, which are associated with executive dysfunction. Executive functions (EF), higher-order cognitive processes allowing goal-directed behavior (e.g. planning and remembering to bring home school books to study), can be particularly affected in children with CHD. To improve EF in affected children, an intervention targeting EF abilities is currently developed. To date, there is conflicting information on the effectiveness of intervention programs. As motivation and adherence of patients and their families are essential for the success of an intervention, a patient-centered needs assessment was performed.
Methods: A multi-step patient-centered needs assessment was performed. In an online focus group interview with families, five children with CHD ($n = 5$; *mean*age = 13.3 years, *SD*age = 1.4; cardiopulmonary bypass surgery in infancy) and five parents ($n = 7$) were asked about their wishes and needs in respect to the content and scope of the intervention. The interviews were evaluated with a Qualitative Content Analysis. The ideas derived from this analysis were transformed into a Delphi survey: An iterative process until agreement is obtained. The importance of the identified proposals was individually rated by other children with CHD between 10 and 12 years of age ($n = 6$), their parents ($n = 6$) their teachers ($n = 5$), and intervention experts ($n = 5$) on a 9-point Likert-scale.
Results: Resulting from the needs assessment, the most marked wishes of the families and the suggestions of the intervention experts are considered and will be implemented in the intervention design: intervention duration of 8 weeks, multimodal features including cognitive online training (3x/week for 30 minutes), virtual coaching to target EF in everyday life (1x/week) and analogue games requiring EF to play with their families and friends at home.
Conclusion: Following a patient-centered approach by involving patients and their families in the development of an intervention will promote the fit of the intervention with families' lives and reinforce perseverance. By creating an intervention to improve EF in children with CHD, their academic achievement and quality of life may be substantially ameliorated.

Keywords: executive functions, intervention, congenital heart disease, children, adolescents

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Systemic sirolimus for pulmonary vein stenosis following repair of total anomalous pulmonary venous connection in an infant

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Background and Aim: Pulmonary vein stenosis (PVS), either acquired or congenital, is often progressive and associated with poor survival. Due to the fragile organisation of intimal meshwork of pulmonary veins to suture lines and interventions, restenosis after total anomalous pulmonary venous connection (TAPVC) repair is not uncommon. Although transcatheter balloon angioplasty and stent placement procedures are frequently used for the relief of pulmonary venous stenosis, the use of systemic sirolimus is currently becoming popular in pediatric age group. Here we report our systemic sirolimus treatment experience with PVS after TAPVC repair.
Methods: A 3 month-old girl with complaints of respiratory distress and failure to thrive was diagnosed as non-obstructive supracardiac type TAPVC and had surgical repair with sutureless technique. Two months after the surgery, she presented with clinical manifestations that required 2 pulmonary vein stent placements within a year. Institutionally the decision was made to start oral sirolimus treatment in order to prevent in stent stenosis of both stents. The drug was initiated at dosage of 1mg/m2 once daily on the day of last stent placement. Her echocardiographic and clinical follow up is free from reintervention for 2 years. Right atrial and ventricular dimensions were diminished and right ventricular systolic pressure was decreased to 35mmHg from 70mmHg gradually with mild gradient of maximum 5mmHg in pulmonary stents with PW Doppler at last visit. She had a course of 2 months of sirolimus theraphy targeting serum drug level at 6–10 ng/ml. No adverse events were detected related to sirolimus therapy.
Results: Systemic sirolimus has been used to prevent in-stent stenosis for arterial diseases. PVS in pediatric cases is an area pending remedy. These therapies in conjunction with anatomic therapies have already demonstrated encouraging preliminary results.
Conclusion: Our case is one promising report of intervention free survival of a child after sirolimus treatment suffering from PVS.Therefore, sirolimus may be a therapeutic option for these patients with restenosis of pulmonary veins but further studies are required.

Keywords: pulmonary vein, sirolimus, stenosis

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SARS-Cov-2 infection in patients with univentricular heart after fontan operation

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Background and Aim: COVID-19 pandemic caused by SARS-Cov-2 coronavirus affects all groups of patients. Although pediatric population seems to be less affected with milder or asymptomatic course of SARS-CoV-2 infection, there are few groups of patients with potential high risk of severe or fatal course of coronavirus disease. These include children with congenital heart defects.
The aim of this study was to evaluate the course of SARS-Cov-2 infection in patients with univentricular heart after Fontan operation.
Methods: From September 2020 to May 2021 (before vccination started in pediatric population in Poland) we screen all 38 Fontan patients admitted to Cardiology Department, Polish Mother's Memorial Hospital Research Institute for SARS-Cov2 antibodies.
Results: We found positive SARS-Cov-2 antibodies in 21 unvaccinated Fontan patients (55% of all hospitalized Fontan patients), 15 boys (71%) and 6 girls in the age 3-22 years (mean 11 years). 14 patients (67%) had hypoplastic left heart syndrome. Course of SARS-CoV-2 infection: asymptomatic course in 11(52%) patients, fever in 7 (33%) patients, cough 4 (19%) patients, diarrhoea in 2 patients, loss of smell and taste -1 patient. One, 18 years old patient suffered from Covid fog symptoms (impairment of sustained attention and memory problems), he hasn't notice any SARS-Cov-2 symptoms but the level of antiobodies was high. Only 3 patients were hospitalized in acute SARS Cov2 infection: 2 due beacause of need for intravenous rehydratation during severe diarrhoea, 1 because of JET (junctional ectopic tachycardia) during fever. There was no case of PIMS (pediatric inflammatory multisystem syndrome) in study group. Medications used in study group: aspirin in 19 (90 %), warfarin in 2, spironolactone in 18 (86%), sildenafil in 9 (43%), angiotensyn-converting enzyme inhibitors in 17 (81%), beta-blockers in 4 (19%) of patients.
Conclusion: 1. In our study severe congenital heart defect such as univentricular heart was not a risk factor of severe course of SARS-Cov-2 infection. 2. Absence of PIMS in analized group of patients may be connected with changed immunologic response in Fontan patients and chronic use of ASA (acetylsalicylic acid). 3. The impact of SARS CoV 2 infection on patients with congenital heart defects needs further studies.

Keywords: SARS-Cov-2 infection, Fontan operation

P-418
Long-term impacts of kawasaki disease on child development: a montreal pilot study on cognition, behavior, and electroencephalography markers

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Background and Aim: Acute Kawasaki disease (KD) induces excessive irritability and central nervous system inflammation. Little is known about KD impacts on neuropsychological and brain development. The aim of this pilot study on primary-school-aged children is to investigate long-term impacts of KD on neuropsychological development and on electrophysiological markers associated with attention deficit (theta/beta ratio; TBR) and brain maturation (alpha peak; AP).
Methods: Fifteen children aged 8.8 ± 2.5 years participated in this project 4.9 ± 2.7 years after KD onset. Intellectual abilities, long-term verbal memory, and auditory sustained attention were measured during formal neuropsychological evaluation. Parents completed four standardized questionnaires assessing: 1) symptoms of Attention Deficit Hyperactivity Disorder; 2) executive functioning; 3) symptoms of Autism Spectrum Disorder; and 4) internalizing and externalizing difficulties. A 3-minute eyes-open resting electroencephalography (EEG) was recorded and compared to 32 matched controls (8.9 ± 2.1 years). Fast-Fourier transformations were calculated in six scalp regions to obtain total spectral density power, TBR and AP. AP parameters were integrated in a reduction algorithm to detect group tendencies.
Results: Performances showed preserved intellectual abilities and long-term memory. Sustained attention was in the limit or low average range for 4 children (29%), with considerable parental reports of inattention (43%) and hyperactivity symptoms (36%). Parents reported executive difficulties; mainly inhibition (29%), working memory (50%), and material organization (43%). Parents also declared potential internalizing difficulties (29%) and externalizing difficulties (21%). EEG findings yielded no difference in TBR, but showed significantly lower AP amplitude ratio in KD compared to controls in 4/6 regions (frontocentral, central, parietal and occipital; $p<0.05$). A weaker and less salient AP was identified in KD patients, with a clear separation of the KD cohort into two clusters showing that acute irritability is associated with a globally weaker AP ($p = 0.023$).
Conclusion: Despite overall preserved cognitive functions in primary school-level children, there is a possible association between KD and attention deficit concerns. Executive, internalizing, and externalizing difficulties were similarly substantial. This study provides a first indication of EEG abnormality, predominantly in children with acute irritability, suggesting that KD impacts cerebral mechanisms years after disease onset. This first EEG-based study merits large-scale longitudinal studies following KD to better characterize patients' developmental trajectory.

Keywords: Kawasaki disease, child development, neuropsychology, electroencephalography

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Physical activity and aerobic fitness in children with inherited cardiac diseases

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Background and Aim: Because of sports and exercise restrictions, children with inherited cardiac disease are at risk of physical deconditioning. Guidelines on sports participation in cardiovascular disease have been less restrictive over time, but their real-life application and behavioural impact have been scarcely evaluated in children. The objective was to evaluate the adherence to the 2020 European Society of Cardiology (ESC) guidelines on sports and exercise in children with inherited cardiac arrhythmia and inherited cardiomyopathy. We also sought to evaluate the aerobic fitness and the behavioural impact of inherited cardiac diseases on physical activity in children.
Methods: Children aged 6-18 with inherited cardiomyopathy or inherited cardiac arrhythmia were eligible for this prospective cross-sectional study. Clinical, demographic and qualitative data were analysed.
Results: A total of 32 children were included in the study (mean age 12.7 ± 3.5 years old). Most children (81.3%) complied with the 2020 ESC guidelines. They were physically active and had a good overall aerobic fitness with a mean peak VO2 value of 36.5 ± 8.0 mL/kg/min (84.0 ± 17.2%). Due to personal or parental behaviours, some children at risk of sudden cardiac death did not comply with the upper limit of physical activity intensity, while others with low risk did not comply with the lower limit.
Conclusion: Most children with inherited cardiac arrhythmia and inherited cardiomyopathy complied with current 2020 ESC guidelines on sports cardiology and exercise in cardiovascular disease. Holistic education and rehabilitation programs dedicated to children with inherited cardiac disorders and their family may be needed to obtain complete compliance.
ClinicalTrials.gov: registration number NCT04650009.

Keywords: physical activity, inherited cardiac arrhythmia, inherited cardiomyopathy, paediatrics, guidelines, aerobic fitness

Table 3 Physical activity and aerobic fitness.

Variable	Total	2020 ESC guidelines			WHO guidelines		
		Adherence	No adherence	*P*	Adherence	No adherence	*P*
	(*n* = 32)	(*n* = 26)	(*n* = 6)		(*n* = 25)	(*n* = 7)	
Physical activity level	24.8 ± 5.3	24.9 ± 5.4	24.5 ± 5.8	0.88	26.2 ± 4.6	19.7 ± 4.8	< 0.01[a]
Peak VO_2 (mL/kg/min)	36.5 ± 8.0	36.2 ± 7.1	38.0 ± 13.2	0.69	36.3 ± 8.5	37.5 ± 5.9	0.79
% predicted peak VO_2	84.0 ± 17.2	84.1 ± 15.4	83.3 ± 28.3	0.93	84.3 ± 18.1	82.0 ± 14.1	0.81
VAT (mL/kg/min)	26.3 ± 6.8	26.5 ± 6.3	25.5 ± 10.2	0.80	26.1 ± 6.8	27.8 ± 7.7	0.66
% predicted VAT	60.6 ± 13.4	61.1 ± 12.2	57.5 ± 20.8	0.63	60.4 ± 12.8	61.5 ± 18.6	0.88
VE/VCO_2 slope	30.8 ± 5.9	30.1 ± 5.8	35.3 ± 6.1	0.16	30.9 ± 6.2	30.3 ± 5.0	0.86
OUES	1427 ± 853	1415 ± 796	1549 ± 1766	0.84	1398 ± 844	1605 ± 1079	0.71
OUES/kg	32.8 ± 15.8	32.2 ± 14.7	38.3 ± 31.8	0.62	34.0 ± 16.4	25.5 ± 10.6	0.40
Peak RER	1.14 ± 0.10	1.14 ± 0.10	1.11 ± 0.08	0.56	1.14 ± 0.09	1.16 ± 0.15	0.43
Peak HR (beats/min)	147.8 ± 28.0	152.9 ± 24.0	125.6 ± 36.4	0.03[a]	148.2 ± 28.2	146.0 ± 30.5	0.88
% predicted HR	71.2 ± 13.3	73.5 ± 11.6	61.2 ± 16.9	0.03[a]	71.4 ± 13.1	70.4 ± 15.6	0.85
Maximum load (watts)	118.9 ± 57.7	119.6 ± 56.5	116.0 ± 69.7	0.85	112.5 ± 54.1	147.0 ± 70.9	0.16

ESC: European Society of Cardiology; HR: heart rate; OUES: oxygen uptake efficiency slope; peak VO_2: maximum oxygen uptake; RER: respiratory exchange ratio; VAT: ventilatory anaerobic threshold; VE/VCO_2 slope: ventilatory efficiency; WHO: World Health Organization. Data are expressed as mean ± standard deviation.
[a] Significant *P*-value.

Detailed aerobic fitness in studied population

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Community based echocardiographic survey of rheumatic heart disease among young adults aged 18-25 years in a rural north indian region

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Background and Aim: Rheumatic Heart disease (RHD) continues to be a significant health problem in low- and low-middle income countries. Echocardiography helps identification of subclinical RHD cases, thereby making it an ideal screening tool for estimating community prevalence of RHD. In contrast to the data in school-aged children, there is only limited data regarding the prevalence of RHD in young adults.
Methods: A community-based echocardiography survey of RHD was carried out in rural areas of Haryana, a state in the northern part of India. 2842 young adults (aged 18 to 25, 49.3% Females) were selected randomly by cluster sampling method. All participants were subjected to cardiovascular examination followed by echocardiography. RHD was diagnosed based on the 2012 World Heart Federation criteria.
Results: Echocardiography detected 15 cases of definite RHD and 21 cases of borderline RHD giving an overall prevalence of 12.7 cases per 1000 population (95% CI 8.9–17.5).Of these 8 had clinically detectable RHD prevalence of 2.8 cases per 1000 population (95% CI 1.2-5.5). Mitral valve was the most commonly affected valve in 31 (86.1%) patients, mitral regurgitation being the commonest lesion. RHD prevalence was significantly higher in females, subjects living in kuccha-pukka (temporary) houses, houses with overcrowding and those with two or lesser rooms. On multivariate analysis female gender (OR 3.35, 95% CI 1.52 to 7.42 p = 0.003), overcrowding (OR 3.57, 95% CI 1.58 to 8.07; p = 0.002) and living in kuccha-pukka house (OR 2.49, 95%CI 0.98-6.37;p = 0.05) were found to be independent predictors of RHD. When compared to prevalence among children in the same region (from a previous study by the same group), prevalence of definite RHD was higher among adults(5.3, 95% CI 3.0-8.7 vs 1.9, 95% CI 0.1-2.8; p = 0.003). The prevalence of clinically detectable RHD among young adults was twice as higher as that among school children (2.8, 95% CI 1.2-5.5 vs 0.4, 95% CI 0.04-0.8; p = 0.003). This may indirectly suggest that subclinical RHD progresses to clinical RHD.
Conclusion: RHD continues to prevail at high rates among young adults in this rural pocket in India. Policy changes aimed at improving RHD surveillance, thereby enabling early diagnosis and management, should be reinforced.

Keywords: Rheumatic heart disease, subclinical RHD, community screening, Echocardiography, World Heart Federation criteria

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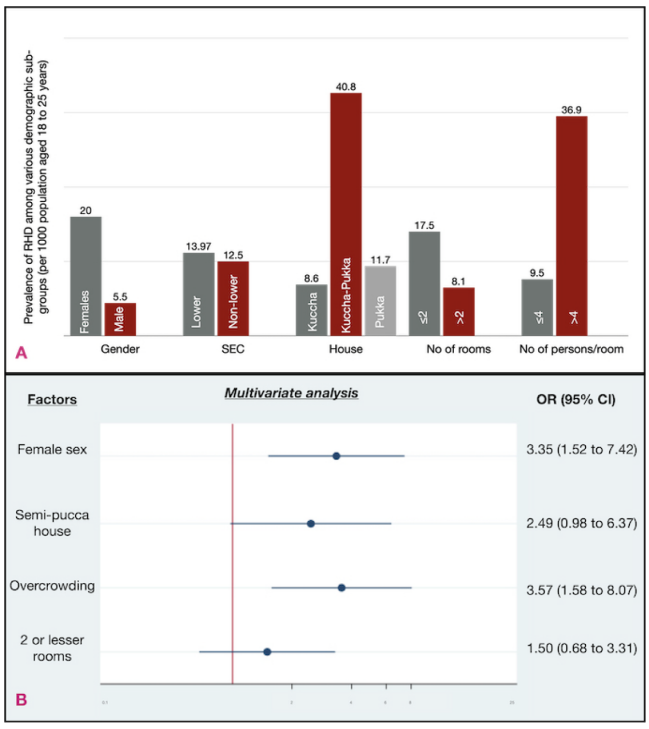
Prevalence of RHD among various demographic groups and the risk factors associated with RHD.



Panel A shows the prevalence of RHD among various demographic sub-groups. Panel B shows the forrest plot of risk factors associated with the prevalence of RHD. OR, odds ratio and 95% confidence intervals are presented

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Necrotising enterocolitis pre cardiac surgery. damned if you, damned if you don't

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Background and Aim: Infants with congenital heart disease (CHD) have the potential to develop Necrotising Enterocolitis (NEC) with devastating consequence. However, simply a suspicion of NEC can delay cardiac surgical intervention causing increase in mortality and morbidity in patients awaiting a "time critical" procedure. This highlights the necessity of accurate diagnosis. We assessed the incidence of NEC in infants with CHD awaiting surgery and the subsequent impact on outcomes at our centre.
Methods: This retrospective cohort study over a 24-month period utilised data obtained from NICOR and local surgical databases. We included infants <90 days of age with a diagnosis of CHD and a diagnosis of NEC prior to cardiac surgery requiring bypass. We collected data on demographics, cardiac lesion, feeding patterns, biochemical markers, diagnostic imaging, clinical assessment and patient outcomes.
Results: 24 patients were diagnosed with NEC prior to a cardiac operation involving bypass in this period, 38% of whom were born prematurely. Within this cohort, 38% of patients had transposition of the great arteries, 21% arch lesions and 17% pulmonary atresia's with lesser incidence of other lesions. Feeding was mixed and 71% were on prostaglandin at time of diagnosis. Clinical signs and biomarkers were varied. 7 patients had an abnormal x-ray, 4 patients ultrasound changes (3 of which had normal abdominal films). 7 cardiac operations were definitively delayed and 4 patients underwent surgical management for NEC. Mean ICU stay post cardiac surgery was 11 days with 0% mortality at 30 days.
Conclusion: The question remains are the typical diagnostic pathways appropriate for our population, Certainly, in our centre the utilisation of ultrasound is under review in light of these findings, as is a review of the feeding recommendations in all our cardiac infants. Is it time to look at new ways of diagnosing these infants, incorporating risk stratification in our usual practice?

Keywords: Congenital. Cardiac. Surgery. Preventative.

P-424

Surgery for congenital heart disease (CHD) during childhood does not imply an increased risk for severe COVID 19 disease

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Background and Aim: The pandemic Covid 19 has so far been reported in more than 245 million individuals and caused five million deaths worldwide. Among risk factors for death and serious illness, heart related conditions were identified early. For children and young adults with CHD, there has been contradicting reports regarding an increased risk for severe disease. The objective of this study was to analyze if patients who had had surgery for CHD in childhood are at risk for developing serious disease or death due to Covid 19.
Methods: To compare our Local surgical registry of all individuals with surgically operated CHD between 1993 and 2018 in childhood, with the occurrence of Covid 19 related disease registered at the National Registry of Diagnoses in Sweden between 2020-03-01 and 2021-03-01. Moderately severe disease was defined as hospitalization during more than two days. Severe disease was defined as treatment in the intensive care unit (ICU).
Results: 3950 individuals were identified through the Local surgical registry. Median age was 16 years (1 – 44 years). 1391/3950 (35 %) were classified as simple defects (ASD, VSD and PDA), 2141/3950 (54 %) had more complex defects, and 418 (11 %) had complicated defects including univentricular defects. 32 patients were found in the National registry of Diagnoses concerning Covid 19 (32/3950 = 0.008%). Fourteen of these were registered as hospitalised with symptoms (0.0035%) at a median age of 24 years (11–37 years). Five patients (median age 24 years, range 13 – 27 years) were hospitalized during more than 2 days (5–32 days). 2/3450 (0.0002 %) patients required treatment at the ICU, of whom one died. The incidence of patients of the corresponding ages needing treatment in the ICU in Sweden due to Covid 19 was 0.0002 %.
Conclusion: There was no increased rate of severe disease of Covid 19 among individuals operated for CHD in childhood.

Keywords: congenital heart disease, covid 19 disease

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P-425

Standardised enteral feeding protocols improve growth and clinical outcomes for infants on CICU

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Background and Aim: To determine the effects of standardising feeding on; enteral nutrition (EN), growth, necrotising enterocolitis (NEC), parenteral nutrition (PN) duration and length of stay (LOS) for infants on CICU.

Methods: A single-centre retrospective chart review was performed in CICU for infants admitted in November/December 2020 (pre-protocol) and May/June 2021 (post-protocol). Data was extracted from the patient record by a Dietitian and Doctor.

A clinical team chaired by a Dietitian used diagnoses and clinical observations to determine whether infants should be kept nil by mouth or follow 'high-risk' or 'standard-risk' protocols. The 'standard-risk' protocol advised four hours of feeding at 30ml/kg/day, followed by increasing feeds to fluid allowance. 'High-risk' protocol infants advanced feeds by 15ml/kg/day (previously 0.5ml/kg/hour four hourly). The option of a 'side-step' from 'high-risk' to 'standard-risk' was introduced following day three for stable infants. Donor expressed breast milk (EBM) was introduced for the 'high-risk' group if maternal EBM was unavailable.

Results: 34 infants in the pre-protocol group and 38 infants in the post-protocol group were recruited. There was a significant decrease between the pre-protocol and post-protocol mean days from CICU admission to EN initiation 8.28 days (±10.80) and 4.22 (±3.92) respectively; p = 0.04 (0.55-8.68). Both groups showed decreased weight-for-age z-scores (WAZ) during admission. There was a significantly smaller drop in mean WAZ score in the post-protocol group -0.21 (±0.42) compared to the pre-protocol group -0.59 (±0.68); p = 0.014 (0.72-0.11). NEC incidence reduced from 14.7% to 7.89%, odds ratio 0.17 in the post-protocol group. Days from CICU admission to EN initiation, days on PN, CICU-LOS and hospital-LOS all decreased; although not statistically significant this may be attributable to a small sample size and could be of clinical significance.

Eight patients in the pre-protocol group did not meet their nutritional requirements prior to discharge; this applied to only one patient in the post-protocol group. No patients developed NEC after moving from 'high-risk' to 'standard-risk' protocol.

Conclusion: The introduction of 'high-risk' and 'standard-risk' enteral feeding protocols improved EN delivery and growth whilst reducing NEC incidence. A 'side-step' from 'high-risk' to 'standard-risk' following day three of feeding appears safe in this small cohort.

Keywords: enteral feeding, protocol, NEC, cardiac, infant

Table 1: growth and clinical outcomes pre- and post-enteral feeding protocol implementation

	Pre-protocol mean (SD)	Post-protocol mean (SD)	Difference	*p*-value (95% CI)
Days from CICU admission to EN initiation	2.77 (2.52)	1.82 (1.7)	-0.95	0.062 (0.04-2.04)
Days from EN initiation to meeting nutritional requirements via EN	8.28 (10.80)	4.22 (3.92)	-4.06	0.04 (0.55-8.68) *
Days on PN	1.93 (5.55)	1.29 (3.31)	-0.64	0.55 (-1.57-2.87)
CICU-length of stay (days)	7.59 (8.69)	7.58 (7.07)	-0.01	0.10 (3.75- 10.57)
Hospital-length of stay (days)	16.30 (19.78)	14.19 (15.58)	-2.11	0.62 (-6.29-11.15)
CICU Admission WAZ	-1.82 (1.76)	-1.44 (1.45)	+0.39	0.34 (-1.2-0.41)
Hospital Discharge WAZ	-2.46 (1.79)	-1.60 (1.58)	+0.86	0.49 (-1.76- - 0.02)
Change in WAZ score from CICU admission to hospital discharge	-0.59 (0.68)	-0.21 (0.42)	+0.38	0.014 (0.72-0.11) *

P-426

Implementation of a consensus-based nutritional pathway for pre-surgical infants with congenital heart disease improves pre-operative growth

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Background and Aim: A consensus-based nutritional pathway for pre-surgical infants with congenital heart disease was published in 2018, following evidence correlating better pre-operative growth to improved post-operative outcomes. The primary aim of this study was to assess growth and clinical outcomes prior to and following implementation of a dietetic outpatient service based upon this pathway.

Methods: A single-centre retrospective chart review was performed in a tertiary cardiac centre in infants between June 2019 (before service implementation) and January 2020-August 2021 (post service implementation). The time-lapse allowed service establishment. Patients were included if they were admitted to cardiac intensive care (CICU) post-operatively with a primary diagnosis of a 'high nutritional risk' lesion as defined by the consensus guidelines. Weight-for-age z-scores (WAZ) taken at three time points (birth, referral to dietetic service, surgery) was the primary outcome; secondary outcomes were CICU length of stay (LOS), hospital-LOS, days ventilated and age at surgery. Data was extracted from the electronic patient record by a dietitian and analysed using paired and independent t-tests through SPSS.

Results: 20 infants in the pre-service group (receiving no or community dietetic input) and 19 infants in the post-service group (specialist tertiary cardiac dietetic input following the consensus-based pathway) were recruited. There was no significant difference between the pre- and post-service mean birth WAZ -1.13 (±1.06 SD) and -0.80 (±1.21) respectively; p = 0.263 (95% CI: -1.14, 0.32). The mean surgery WAZ was significantly lower in the pre-service group -2.13 (±1.72), compared to post service group -0.99 (±1.18); p = 0.018 (-2.14, 0.21). In the post-service group, mean birth WAZ 0.81 (±1.21) was significantly higher than WAZ at referral to dietetics -1.49 (±1.16) p = 0.001 (95% CI: 0.31, -0.06). The mean referral WAZ -1.49 (1.16 SD) was significantly lower than surgery WAZ -0.99 (±1.21) p = 0.008 (95% CI: -0.86, -0.15). Age at surgery, days ventilated post op, CICU-LOS, hospital-LOS and age at surgery decreased in the post-service group; although not statistically significant this may be attributable to a small sample size and could be of clinical significance.

Conclusion: Specialist dietetic input following a consensus-based nutritional guideline for pre-operative cardiac infants diagnosed with a 'high nutritional risk' lesion improves growth at surgery.

Keywords: growth, WAZ, surgery, feeding, congenital heart disease, infant

Table 1: growth and clinical outcomes pre- and post-dietetic service implementation for pre-surgical cardiac infants

	Pre-service (±SD)	Post-service (±SD)	Difference	*p*-value (95% CI)
Birth WAZ	-1.13 (1.06)	-0.80 (1.21)	+0.33	*p*=0.263 (-1.14, 0.32)
Surgery WAZ	-2.13 (1.72)	-0.99 (1.18)	+1.14	*p*=0.018 (-2.14, 0.21) *
Age at surgery (mo)	5.5 (3.2)	3.8 (2.1)	-1.7	*p*=0.74 (0.15,3.37)
Time ventilated post-op (days)	1.9 (1.3)	1.6 (1.2)	-0.3	*p*=0.514 (0.56, 1.10)
CICU-LOS (days)	5.3 (8.7)	3.7 (3.0)	-1.3	p=0.458 (-2.71, 5.84)
Hospital-LOS (days)	10.2 (8.7)	8.7 (8.0)	-1.5	p=0.574 (3.90-6.93)

*=achieved significance within 95% confidence intervals

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P-428
Cardiovascular manifestations in multisystem inflammatory syndrome in children (MIS-C) associated with COVID-19

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Background and Aim: Cardiac involvement in multisystem inflammatory syndrome in children (MIS-C) associated with Coronavirus 2019 disease (COVID-19) is often observed with high risk of hearth failure. Early diagnosis and treatment are mandatory for a good outcome. The aim is to describe cardiovascular involvement, management and early outcome for patients with MIS-C and to analyze the differences in cardiovascular manifestations between two groups: younger and older than 6 years old.
Methods: This retrospective observational study describes cardiovascular clinical manifestations, laboratory findings, cardiac imaging, according to different age groups, and treatment in patients with diagnosis of MIS-C admitted to the Pediatric Istitute Giannina Gaslini between March 2020 and September 2021.
Results: We collected 25 patients. Median age at onset of symptoms was 5 years old (interquartile range IQR, 3-12 y), 12 boys (56%). Immunoglobulin G antibodies were positive in 70% cases, Polymerase chain reaction (PCR) nasal/throat swab test for COVID-19 was positive in 15% cases, at the admission. The remaining cases had close contacts of COVID-19 positive cases. Predominant coronary artery abnormalities were observed in age group up to 6 years old (n.13) with development of small and medium aneurysms in half of cases and low rate of mild ventricular dysfunction. While children between 7-18 years of age present myopericardial involvement with ventricular dysfunction in 67% cases, from mild to moderate. Only two cases of transient coronary dilatation. Frequent electrocardiogram abnormalities: ventricular repolarization anomalies and reversibile QTc prolongation interval. Laboratory findings showed rised inflammatory markers and only mild elevation of cardiac enzymes compared to an early and significant NT-pro-BNP increase. All patients were treated with intravenous immunoglobulin and corticosteroids. Some cases needed anakinra. Aspirin and heparin was administrated. No inotropes requied but only cardioprotective therapy. No need of Intensive Care Unit.
Conclusion: This case-series shows the frequent cardiovascular involvement in MIS-C with a peculiar distribution, according to differents age's group: coronary artery anomalies in young ones, myopericardial disease in old ones. Prompt multi target anti-inflammatory therapy could have an effect to favorable outcome.
Keywords: cardiac involvement; children; MIS-C; SARS-COV-2

P-429/Moderated Poster
Paediatric and adult congenital cardiology training in europe

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Background and Aim: Limited data exist on training of European paediatric and adult congenital cardiologists. This study aimed to report on the current status of paediatric and Adult Congenital Heart Disease training throughout Europe.
Methods: A structured approved questionnaire was circulated to national delegates of Association for European Paediatric and Congenital Cardiology in 33 European countries. Eighteen adult CHD (ACHD) practitioners were surveyed on current status of training of adult CHD and potential benefit of establishing certification in ACHD.
Results: Delegates from 30 countries (91%) responded. Paediatric cardiology was not recognised as a distinct speciality by the respective ministry of Health in 7 countries (23%). Twenty one countries (70%) have formally accredited paediatric cardiology training programs, six (20%) have substantial informal (not accredited or certified) training and three (10%) have very limited or no program. Twenty two countries have a curriculum. Twelve countries have a national training director. There was one paediatric cardiology centre per 2.66 million population (range 0.87-9.64 million), one cardiac surgical centre per 4.73 million population (range 1.63-10.72 million), and one training centre per 4.29 million population (range 1.63-10.72 million population). The median number of paediatric cardiology fellows per training program was 4 (range 1-17) and duration of training was 3 years (range 2-5 years). An exit examination in paediatric cardiology was conducted in 16 countries (53%) and certification provided by 21 countries (70%). Paediatric cardiologist number is affected by gross domestic product ($R2 = 0.41$). This result weakened when adjusted for cardiologists per million population ($R2 = 0.014$). Only two ACHD cardiologists from one country stated that certification was required to practice in their country. The majority of ACHD consultants surveyed (fifteen, 83%) expressed that introduction of ACHD certification would be helpful.
Conclusion: Training varies markedly across European countries. Although formal fellowship programs exist in many countries, several countries have informal training or no training. Only a minority of countries provide both exit examination and certification. Harmonisation of training and standardisation of exit examination and certification can reduce variation in training thereby promoting high-quality care by European congenital cardiologists.

Keywords: Adult CHD, Congenital Cardiology, Education, Paediatric Cardiology, Training, Certification

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P-430

Right aortic arch: long-term follow-up in 502 patients at a tertiary center

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Background and Aim: (1) to report our experience with right aortic arch (RAA) and its variants, association with complete vascular ring (CVR), congenital heart disease (CHD), extracardiac malformations and (2) to describe the long-term follow-up in terms of symptoms and surgery for CVR.
Methods: We collected clinical and instrumental data from 502 patients with RAA referred at our tertiary center.
Results: 157 patients (31.3%) had fetal diagnosis (medium follow-up 7.6 years). 330 (65%) had CHD and 89 (17%) genetic anomalies, with DiGeorge syndrome the most frequent (9.5%). Genetic disorders were significantly more frequent in those with CHD (23% versus 7.5%, $p < 0.001$). 58 (11%) had extracardiac anomalies. Among the RAA not forming CVR, left anterior ductus arteriosus with mirror-image (MI) branching pattern of epiaortic vessels (RAA/LADA/MI) was present in 29, 100% associated with CHD (conotruncal in 72%). Right ductus arteriosus (RAA/RDA/MI) was found in 8, 4 with complex CHD. CVR was present in 246 (49%): the most frequent were RAA, left posterior ductus arteriosus with aberrant left subclavian artery (RAA/LPDA/ALSA – 163 cases, 66%) and double aortic arch (DAA – 66 cases, 27%), Left posterior ductus arteriosus (LPDA) with MI branching (RAA/LPDA/MI – 12 cases, 5%) and aberrant left innominate artery (RAA/LPDA/ALIA – 5 cases, 2%) were rare. CVR were most frequently associated with minor CHD (26% versus 17% major CHD). Seventy-one (29%) pts had respiratory symptoms (29 in the first 2 months of life, 22 within the first year of life, 20 afterwards). 80 pts performed lung-function test, 28 (35%) were positive. At CT mean tracheal compression was 46% (57% for DAA, 43% for RAA/LPDA/MI. 109 (45%) were operated (median age 31.2 months). Three among 16 pts were re-operated for persistent symptomatic compression, the majority being DAA.
Conclusion: RAA is commonly associated with CVR, asymptomatic in 2/3 of cases but requiring surgery in 50%. Symptoms resolves almost completely and reoperations are reserved to the DAA. RAA is present along with major CHD in half cases and these patients are at higher risk of genetic syndromes, however not frequent.

Keywords: right aortic arch, tracheal compression, vascular ring,

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Family-centered early motor intervention in infants with complex congenital heart disease: protocol for a randomized controlled pilot trial

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Background and Aim: Children with congenital heart disease (CHD) are at risk for developmental impairments. Motor developmental delay is the first to become visible during the first year of life. Several studies have investigated the neuromotor development in children with CHD. However, only few interventional studies exist that aim to improve neuromotor development in infants after open heart surgery within the first year of life. Study results are inconclusive due to their low level of evidence.
The aim of our study is to conduct a tailored family centered early motor intervention promoting motor development in infants with CHD.
Methods: This prospective, single centre randomized controlled pilot study will investigate an early motor intervention compared to standard of care for infants with CHD after open-heart surgery. Participants will be 20 infants with CHD aged 3-5 months, who underwent open heart surgery within the first months of life. Infants will be randomly allocated to the intervention or control group. Infants assigned to the intervention group will receive family-centered early motor intervention for a duration of 3 months. The intervention promotes infants' self-initiated motor activities, parents' attendance and active involvement and support transactions between child, parents, and therapist. Infants assigned to the control group will receive standard of care. Specific assessments will measure infants' motor outcome and parents' and infants' quality of life with a set of questionnaires at baseline, post-treatment and at a follow-up at 12 months of age. The primary outcome of this study is the Infants Motor Profile. Secondary outcomes include the Alberta Infant Motor Scale and the Bayley Scales of Infant and Toddler Development 3rd version. Parental questionnaires evaluate socioeconomic status, parental physical and psychological experience, parental empowerment, and parents' and infants' health related quality of life.
Results: This pilot study will be evaluated by linear regression analyses to investigate between group, within subject differences across treatment groups and associations between motor development, and parents' and infants' health related quality of life.
Conclusion: This protocol will provide a foundation for a randomized clinical trial for a family-centered early motor intervention for infants after open heart surgery.

Keywords: congenital heart disease, early intervention, neuromotor development, physiotherapy, family-centered therapy

P-433

Communication between a pulmonary artery branch and the left atrium: a case series

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Background and Aim: The communication between a branch of the pulmonary artery (PA) and the left atrium (LA) is a rare cyanotic congenital cardiac malformation, first described by Freidlich in 1950. Until now, isolated cases have been published. Usually, the fistula connects the right PA (RPA) and the LA. A very low number of cases of congenital communication between the left PA (LPA) and the LA have been reported.
Methods: We present a series of three cases of proven and successfully treated communications between a PA branch and the LA.
Results: Case 1. A 5-year-old boy with cyanosis from birth which progressed in recent years with retardation in physical development. Echocardiography showed normal intracardiac anatomy and nonconclusive data for RPA to LA communication. The invasive study proved the RPA to LA communication, forming a giant saccular aneurysm and a large right-to-left shunt (RLS) with oxygen saturation (SatO2) of 67% in the aorta. Successful surgery was carried on. Case 2. One-day-old newborn with pronounced

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generalized cyanosis and SatO2 of 48%. Echocardiography showed normal intracardiac anatomy, small foramen ovale, LA dilation, and reduced left ventricular (LV) systolic function (ejection fraction [EF] - 55%), dilated RPA with direct communication between the right lower lobe branch and the LA with massive RLS. The infant was catheterized, the diagnosis was confirmed, and the communication was closed using a 3/4 mm Amplatzer Duct Occluder II. After the intervention SatO2 increased to 96%. Case 3. A newborn with pronounced generalized cyanosis from birth. The fetal echocardiogram showed inconclusive data for a giant aneurysmally dilated communication between the LPA and the LA. After birth the diagnosis was confirmed by echocardiography, catheterization and contrast computed tomography. Common drainage of the aneurysmal sac and the left inferior lobe pulmonary vein in the LA was demonstrated. The aneurysmal sac compressed almost the entire left lung. The infant was successfully operated on, which required segmental resection of the left lung.
Conclusion: To our knowledge, this is the first presented series of 3 cases, including 2 infants, with congenital communication between a branch of PA and LA proven and successfully treated by transcatheter or surgical methods.

Keywords: pulmonary artery fistula, cyanosis, treatment

P-434

Single-centre study: MIS-C impact on blood pressure, coronary arteries and left ventricular function

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Background and Aim: MIS-C is a hyperinflammatory syndrome caused by Sars-CoV-2 virus. Cardiovascular system impairment is observed up to 100 % of all MIS-C patients with a wide spectrum and severity of symptoms. It is important to identify the course of the disease and its outcome, which could significantly improve public health.
Methods: A single-centre study, prospective cohort study, conducted in the Children's Clinical University hospital in Latvia from January to December 2021. Patients between the ages of one to seventeen years who met the MIS-C criteria were included in the study. We evaluated blood pressure, left ventricular heart function, size of coronary arteries and hospital course.
Results: Thirty-one patients were included who met the MIS-C criteria. The median age was 8.0 years, 52% were boys. Of all patients 77% initially presented with hypotension of whom 42% required inotropic support. Treatment in PICU was required in 58% of all patients. Reduced left ventricular ejection fraction was observed in 35% of all patients. Mildly decreased ventricular ejection fraction (<55%) was observed in 19% of cases but moderate dysfunction (ejection fraction <45%) was observed in 16% of patients. Twelve percent of patients received milrinone to improve left heart function. Left heart function significantly improved in all patients during the hospitalisation. In 6 % of all patients coronary artery dilations was observed. All patients had dilations resolution at the time of discharge. Median length of hospitalisation was twelve days and median length of PICU stay was three days.
Conclusion: All patients cardiovascular symptoms had resolved at the time of discharge. Whether patients will have chronic cardiac impairment is unknown therefore it is crucial to perform long-term follow-up.

Keywords: MIS-C; Hypotension, coronary artery dilation.

P-435

Clinical characteristics, treatment response and short-term general and cardiac outcomes in multisystem inflammatory syndrome in children (MIS-C)

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Background and Aim: In April 2020, clinicians in the United Kingdom observed a group of children with hyperinflammatory shock with significant cardiovascular effects, with features similar to Kawasaki disease and toxic shock syndrome. This new syndrome that is temporally related to previous exposure to SARS-CoV-2 infection, is now known as multisystem inflammatory syndrome in children (MIS-C). The aim of this study is to describe the incidence, the clinical, laboratory and echocardiografic characteristics of hospitalized children who met criteria for the MIS-C and analyse short time general and cardiac outcomes in our region.
Methods: Data from children admitted who fulfilled the case definition of MIS-C were collected between October 2020 and November 2021.
Results: 10 cases of MIS-C were reported; the incidence of MIS-C during this period was 1 per 10000 positive sars-cov2 cases (diagnosed by polymerase chain reaction test or antigen test). The median age was 10 years (IQR 6-12). 70% were male and 50% corresponded to ethnic minority group in our country (20% Latin American and 30% African). 8 of 10 patients (80%) had evidence of current or prior SARS-CoV-2 infection and 2 of 10 (20%) had an antecedent of contact with a COVID positive patient. Fever (100% patients), hematologic disturbances (90%), cardiac involvement (biochemical or echocardiographic) (80%), gastrointestinal (80%) and mucocutaneus (50%) symptoms were common presenting features. 8 of 10 were admitted in the pediatric intensive care unit. When referring to cardiovascular involvement, 1 of 10 (10%) patients had left ventricular systolic dysfunction, 2 of 10 (20%) had mild pericardial effusion and 4 of 10 (40%) mild coronary artery abnormalities.
Conclusion: Although the incidence is low, in this case series most patients show homogeneous clinical and laboratory findings. Since cardiac involvement is described in a high proportion of patients, long-term follow-up is required due to the unclear prognosis and risk of progression of cardiac manifestation.

Keywords: Multisystem Inflammatory Syndrome in Children (MIS-C)

P-436

Impact of coronavirus disease 2019 on functional capacity in paediatric congenital heart disease assessed by cardiopulmonary exercise test

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Background and Aim: Since December 2019 the novel coronavirus disease 2019 (COVID-19) has been burdening all health systems worldwide. However, cardiopulmonary repercussions in paediatric patients with congenital heart disease (CHD) are unknown. The aim of this study is to compare changes in cardiopulmonary exercise test (CPET) in this patients before and after COVID-19.
Methods: Prospective observational study was lead comparing CPET results after COVID-19 in paediatric patients with stable CHD who had a previous routine CPET. All underwent for standardised CPET, using Godfrey ramp protocol as recommended by the European Society of Cardiology (ESC). Measured variables, expressed by predicted values, were: forced vital capacity (FVC), forced expiratory volume (FEV1), ratio of minute ventilation to carbon dioxide production (VE/VO2 slope), peak oxygen consumption (peak VO2), oxygen uptake efficiency slope (OUES), oxygen pulse (O2 pulse) and peak heart rate (pHR). Wilcoxon test was used to compare continuous variables for related samples.
Results: Ten patients (6 boys, 60%; mean age 11,4 ± 2,4 years) with hemodynamically stable CHD (3 Tetralogy of Fallot, 30%; 2 transposition of the great arteries, 20%; 2 dilated cardiomyopathy, 20%; 2 Kawasaki disease, 20%; 1 cardiac tumor, 10%) were selected to repeat a post-COVID CPET. All of them had mild COVID and could follow ambulatory treatment. Comparing before/post COVID tests, there were no significantly changes in predicted respiratory parameters: FVC (90,6 ± 7,4 vs 98,1 ± 23,9%; p = 0,799), FEV1 (89,5 ± 13,8 vs 94,5 ± 8,8%; p = 0,475), VE/CO2 slope (31,6 ± 3,7 vs 30,6 ± 3,9º, p = 0,203). In the same way, no significantly changes were seen in cardiovascular predicted parameters: oxygen pulse (97,3 ± 19,2 vs 98,5 ± 17,4%, p = 0,798), peak VO2 (82,4 ± 19,4 vs 76,8 ± 13,7; p = 0,123) and OUES (1,79 ± 0,4 vs 2,01 ± 0,6; p = 0,066). Respect peak VO2, there was a non-significant slightly decrease in post-COVID test (82,4 ± 19,4 vs 76,8 ± 13,7; p = 0,123).
Conclusion: In our series, post-COVID CPET results showed that paediatric patients with hemodynamically stable CHD had no impairment in their functional capacity in relation to Sars-CoV-2 disease. Contrary to adults with previous cardiovascular disease, children should have mild infections without sequelae in cardiopulmonary function.

Keywords: Coronavirus disease 2019 (COVID-19); Congenital Heart Disease (CHD); Cardiopulmonary exercise test (CPET)

P-438

A redo mitral and tricuspid surgery for hemolytic anemia: does tricuspid regurgitation contribute for hemolysis?

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Background and Aim: We are reporting a redo mitral and tricuspid valve surgery for a severe hemolytic anemia after patient was transfused 22 times in 4 months period in a 10 years old boy underwent mitral and tricuspid valve repair following severe mitral and tricuspid regurgitation due to rheumatic heart disease (RHD).
Methods: A10 years old boy presented with gross hematuria and subsequently diagnosed to have severe hemolytic anemia after he underwent mitral and tricuspid valve repair for severe mitral and tricuspid regurgitation due to rheumatic heart disease (RHD). Mitral valve repair was done using untreated autologous pericardial patch to augment the posterior mitral valve leaflet and a remodeling posterior annuloplasty using Teflon strip. Tricuspid valve repair was done using 28mm three dimensional McCarthy Edwards ring (MC3) remodeling ring. Both trans esophageal and trans thoracic echocardiography confirmed that there is moderate regurgitation in the mitral valve and regurgitation in the tricuspid valve post repair. Patient become severely anemic after presenting with gross hematuria and subsequently transfused 22 times in three months period post the index first surgery. Peripheral microscopic smear shows normocytic and normochromic RBC with frequent schistocytes.
Results: A redo sternotomy approach for a redo mitral valve replacement and redo tricuspid valve repair after explanting the previous ring from tricuspid valve and re-repair using autologous pericardium was done. Intra-operatively there was jet denuded the 2nd segment of the posterior mitral leaflet (P-2) segment of posterior mitral leaflet (PML) and a raw surface of the tricuspid valve ring at the septo-posterior leaflet junction. Post-operatively hematuria stopped, jaundice resolved, hepatomegaly subsided and the anemia corrected.
Conclusion: The regurgitant jet of a tricuspid valve after ring repair can contribute for severe anemia caused by hemolysis from tricuspid regurgitation following valve repair using ring. We believe both regurgitant jets from mitral and tricuspid valve has contributed to the hemolysis and the fast deteriorating clinical presentation. Addressing both valves solved the problem and helped for the fast recovery of this patient. We could not find so far in the English literature mentioning tricuspid valve as a contributory cause of hemolytic anemia and addressed by re-repair.

Keywords: Tricuspid valve ring, mitral valve repair, hemolytic anemia.

P-439

Cardiac findings and short-term outcomes of multisystem inflammatory syndrome associated with COVID-19 single center experiences from türkiye

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Background and Aim: Multi-system inflammatory syndrome in children (MISC) associated with COVID-19 has been described as a potentially life-threatening disease. In this study, we aimed to evaluate cardiovascular findings in children diagnosed with MISC at initial presentation and follow up.
Methods: Between November 2020 and November 2021, 35 children diagnosed with MISC based on WHO criteria were evaluated in this retrospective study.Cardiac markers, electrocardiography and echocardiography were performed in all cases at presentation. Cardiac evaluation were repeated at the mean of 10th week after discharge(range:5 to 33weeks).

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Results: At this period, 633 children had positive PCR test of Covid -19. The freguency of MISC was 5.5% in our cohort. The median age was 9 years at diagnosis. Comorbid diseases were found in 20% cases, but none had preexisting heart disease. All patients had high grade fever and laboratory evidence of hyperinflammation. Most cases had mild form disease, however 12 patients had been hospitalized in ICU median 6 day. 27 cases (77%) had cardiovascular involvement.Kawasaki-like findings were found in 10 patients and 5 cases were presented with shock(Figure-1) Echocardiography;Left ventricular (LV)systolic dysfunction (EF<57%) was detected in 11 cases (31.4%) and coronary artery (CA) dilatation(z score> 2)was found in five(14.2%) cases. Pericardial effusion was seen in 12 cases. Electrocardiography: Sinus tachycardia was the most common finding. 2 cases had prolonged QTc interval and four cases had T wave alterations. Four cases had experienced complex ventricular arrhythmia. Cardiac markers:24 cases had high Pro-BNP level. 18 cases also had high Troponin T levels. Pro-BNP and Troponin T levels were not found to be correlated with LVEF. Only one adolescent boy who had severe cardiac dysfunction died during the acute period. Followup:There were two cases with persistent cardiac symptom, but no case had LV systolic dysfunction. The mean PR intervale was significantly lower than initial measurements. The mean of QT and QTc at follow up were not different from basal measurements.The mean LVEF was significantly higher than the initial levels. The basal CA z scores normalized at followup.
Conclusion: MISC is characterized predominantly by cardiovascular system involvement, but the children with MISC have good cardiac outcomes at short term follow up.

Keywords: multi-system inflammatory syndrome, COVID-19, cardiac involvement, followup, echocardiography

	Value		Value
Age (years)		Clinical course at admission n (%)	
Median (Minimum-maximum)	8.8 (0.83-18)	• Mild	19 (54.3)
< 1 years n (%)	2	• Moderate	5 (14.3)
1-5 years	9	• Severe	11 (31.4)
5-12 years	11	Hospitalization in PICU n (%)	12 (34.3)
12-18 years	13	• Median length of stay (day)	6
Gender (Male / Female) n (%)	21/14	• Range	3-14
Body mass index (BMI)	18.8 (4.7) (12.9-35.5)	Treatment for PMIS n (%)	
BMI categories,		• IVIG	28
Underweight	2	• Steroids	20
Normal	29	• Anakinra	5
Overweight	3	• Anticoagulation	29
Obese	1	• Positive inotrophic drugs	11
Comorbid disease	7	• Plasmapheresis	7
• Allergic rinit	1	Mechanical Ventilation	5
• Kidney disease	1	ECMO	1
• Neurological disease	1	End point	
• Endocrin metabolic	1	Full recovery	31
• Oncologic disease	1	Partial improvement	3
• Milier Tuberculosis	1	Mortality n (%)	1
Prexisiting heart disease	0		
PMIS subtypes			
• Kawasaki like	10		
• Hypotension-Shock	5		

Figure 1 Demographic features, clinical characteristics of patient diagnosed with MISC shown. The treatment modalities of MISC were shown also.

P-440
The influence of glutaraldehyde on the microscopic structure of human pericardium

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Background and Aim: Glutaraldehyde (GA) is used in cardiac surgery for stabilization of valves prostheses and pericardial patch material. Degeneration processes, mainly calcifications of these tissues are observed clinically and necessitate reoperations. However, such redo procedures should be avoided whenever possible. As the influence of GA on calcification processes is unknown, microscopical alterations of pericardial structure were evaluated - in dependence of fixation time and GA concentration.
Methods: Pericardial samples were taken from 9 patients aged 40+ years who underwent cardiac surgery, coronary artery bypass or valve implantation/reconstruction. Specimens were cut in five equal pieces and treated with GA at fixed concentrations (0.3125%, 0.625% or 1.2%) but different exposer times (5min, 10min, 20min, 30min and 60min). Eosin staining was used for microscopic examination of pericardial collagen structure.
Results: The collagen structure studied microscopically depended on both GA incubation time and GA concentration. At low GA concentrations and short incubation times, individual collagen fibers appeared separately, whereas the fibers merged more and more with increasing incubation time. After one hour incubation period, single collagen fibres could not be distinguished at any GA concentration. For fixed incubation times no differences were seen in the collagen structure when 0,3125% and 0,625% GA was used. However, at a concentration of 1.25% GA and fixed incubation times, differences in collagen structure became apparent. Fusion of individual collagen fibers was already observed at low incubation times, but the development was less pronounced with increasing incubation time.
Conclusion: Pericardial collagen structure changes with increasing incubation time and increasing GA concentration by raising fusion of single fibers. For GA concentrations of ≤0,625%, fiber fusion depends plainly on incubation time. That is relevant as this concentration is used in cardiac surgery. At a concentration of 1.25% GA, single collagen fibers could not be separated, even with short incubation times and the fusion process over time was not as impressive as at lower concentrations. Fusion of individual collagen fibers and changes in appearance (less undulating) were responsible for stiffening of GA-fixed pericardium. In which way these microscopic changes influence the calcification process ought to be subject of further studies.

Keywords: Fixation, Glutaraldehyde, Pericadium, Collagen Structure

P-441
Effect of cibenzoline on pediatric hypertrophic cardiomyopathy

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Background and Aim: Cibenzoline (Cs), an antiarrhythmic drug, was reported to be effective in obstructive hypertrophic cardiomyopathy (HCM) in adults. We aimed to observe the effect of Cs on the hemodynamics of pediatric patients with HCM.
Methods: A total of 19 pediatric patients with HCM were included in the study. Beta-blockers were used concomitantly in all patients. From left heart cardiac catheterization, the outflow tract pressure gradient (LVPG) (30 mmHg or more is considered significant stenosis, and a change of 30% or more is a significant change), dp/dt, end-diastolic pressure (LVEDp), and from echocardiography, the E/A ratio of the inflow waveform was measured. These values were obtained before and after acute (intravenous injection, 1.4 mg/kg) and chronic loading (oral administration) of Cs and were compared. The plasma concentration of Cs was measured immediately after intravenous injection for acute loading and 2 hours after oral administration. The values are described using

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medians (interquartile ranges). est K was calculated by LVEDp-left ventricular minimum pressure/ stroke volume index. The Wilcoxon signed-rank test was used for comparison. Values of $p < 0.05$ were considered to be statistically significant.
Results: The median age of the patients was 12 (6–16) years. The patient characteristics and the results of the comparison before and after Cs loading are shown in Table 1. The plasma concentration of Cs (pg/ml) = 44.6 x dose per body weight (mg/kg) − 24.8 was established in the chronic loading group ($R2 = 0.78$, $p = 0.0003$) (Figure 1). Plasma Cs concentration was higher with acute loading (829 pg/ml (296–1866)) than with chronic loading (241 pg/ml (119–324)). Although the LVPG did not improve in one patient in the chronic loading group, overall, Cs loading significantly improved the LVPG. Examining the relationship between the changes in E/A ratio and LVEDp, LVEDp decreased when the E/A ratio increased, except in two cases (Figure 2).
Conclusion: Cs was considered to be effective in improving LVPG. It was considered that a significant decrease in contractility contributed to MantUS System in usethe improvement of the LVPG. Interestingly, the relationship between the E/A ratio and LVEDp suggests that Cs may also improve diastolic function.

Keywords: cibenzoline, hypertrophic cardiomyopathy, left ventricular outflow tract pressure gradient, diastolic function

figure and table20211201

Table1	Cs loading		
	pre	post	
LVPG (mmHg)	53(30-82)	16(8-38)	**p=0.05**
dp/dt (mmHg/sec)	1300(1200-1351)	1081(922-1130)	**p=0.0005**
LVEDp (mmHg)	15.0(12.0-28.5)	14.0(9.0-24.0)	**p=0.01**
E/A ratio	1.0(0.9-1.4)	1.8(1.4-2.2)	**p=0.002**

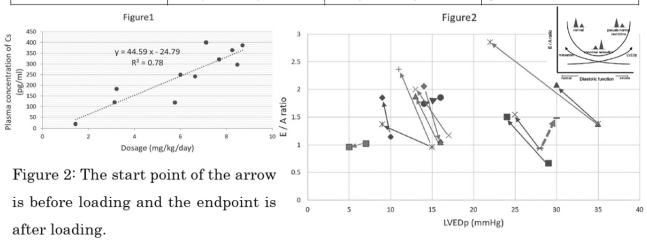

Figure 2: The start point of the arrow is before loading and the endpoint is after loading.

P-443

Idiopathic hypereosinophilic syndrome associated with pulmonary hypertension in an infant

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Background and Aim: Hypereosinophilic syndrom (HES) is defined by eosinophilic infiltration in any tissue or organ and increased eosinophils in peripheral blood. It is quite rare in children and mostly affects skin, lungs, rarely heart and liver. Pulmonary hypertension (PHT) is one of the rare cause of HES in children and diagnostic evaluation and management is difficult like HES. Here, we present an infant with HES-associated PHT who was successfully treated with steroids.
Methods: A 6-month-old girl presented with cough, dyspnea, and tachypnea. However, oxygen saturation was 88% on room air, her physical examination revealed normal findings. white blood cell (WBC) count of 40.6x109/L, and total eosinofil count (TEC) of 18.9x109/L. Peripheral smear showed 42% eosinophils, 34% lymphocytes, 20% granulocytes. She had a high total serum IgE level (100 IU/mL, normal range: 0-8 IU). Her bone marrow smear revealed 28% eosinophilic cells. Flow cytometric analysis was normal. Her electrocardiogram showed sinus tachycardia, right axis and right ventricular hypertrophy. Echocardiography revealed enlargement of the right ventricle, right atrium, main pulmonary artery and an increase in right ventricular systolic pressure (RVSP) (80 mmHg). Right heart catheterization showed pulmonary artery pressure 52/26/37 mmHg, pulmonary vascular resistance 3.8 Wood Units.m2. Genetic analysis for primary pulmonary hypertension and hypereosinophilia were also negative. Searching ethiologic agents for hypereosinophilia showed no parasitic infestation, allergy or autoantibody.
Results: The patient was diagnosed with idiopathic HES associated with PHT. PHT and HES were improved after one month of methylprednisolone treatment. Her TEC was 0.05x109. RVSP regressed from 80 mmHg to 25mmHg.No recurrent episode of PHT and HES has been observed during six months follow up.
Conclusion: HES is a rare cause of PHT in children. Corticosteroids are considered in patients with PHT due to HES. Close clinical follow-up is required after the end of treatment.

Keywords: children, pulmonary hypertension, hypereosinophilic syndrome, corticosteroid

P-444

Features of myocardial infarction in children

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Background and Aim: Mycardial infarction (MI) is a rare event in children. It is characterized by various causes and difficulties in diagnostic criteria and therapeutic strategies. We aimed to detremine diagnostic criteria, causes and managment of MI in infant.
Methods: All cases of MI in infant were retrospectivelly enrolled in this study. Symtoms, electrocardiographic, echocardiographic, and angiographic criteria as well as therapeutic managment were collected.
Results: A total of 6 patients were hospitalized for MI between 2015 and 2020. One child had the history of arterial switch operation, two with familial hypercholesterolemia (FHC). Symptoms on admission were chest pain in two children (8 and 13 years old), vomiting and pallor in a 9-month-baby, dyspnea in two children (15 and 5 years old) and cardiogenic shock in a 3-month-baby. Electrocardiogram showed ST segment elevation in two children, necrosis Q wave in four chirdren. Echocardiography led to the diagnosis of coronary artery (CA) anerysm in a child, a large CA fistula between right CA and right atrium and a case of Anomalous Left Coronary Artery from Pulmonary Artery (ALCAPA). All children had a reduced left ventricle ejection fraction < 50%. Coronary CT scan and /or coronary angiography cofrmed the diagnosis of: one case of CA connexion anomalous after artery switch, one case of CA fistula, one case of large CA anerysm (>6 mm) after kawasaki syndrom, one case of ALCAPA and two cases of atherosclerotic CA disease due to FHC. Three cases were surgically treated (CA fistula, ALCAPA, CA connexion anomalous). CA aneurysm was managed medically

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by oral anticoagulation. One case of FHC (13 years) was treated by percutaneous transluminal angioplasty with a high dose of statin and aspirine. A case of mortality of the other child with FHC because of severe cardiogenic shock.
Conclusion: Various causes could result in MI in children. Specific and differents diagnostic and therapeutic approches are needed to improve prognosis.

Keywords: myocardial infarction, children

P-445
The use of speckle tracking echocardiography for early detection of myocardial dysfunction in patients with duchenne muscular dystrophy

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Background and Aim: With improved respiratory care in Duchenne muscular dystrophy (DMD) patients, cardiac manifestations become the leading cause of morbidity and mortality. Two-dimensional-speckle tracking echocardiography (2D-STE) has recently emerged as a non-invasive functional biomarker for early detection of DMD-related cardiomyopathy. This study aimed to detect subtle cardiac changes using 2D-STE analysis in DMD children.
Methods: The study is a retrospective review of the medical records and transthoracic echocardiograms of DMD male patients followed at our institution. DMD diagnosis was based on clinical features and confirmed by genetic testing in all subjects. The transthoracic echocardiographic studies of DMD patients were obtained according to a standardized protocol in our echocardiography laboratory.
Results: 23 male patients with DMD and 15 normal male controls were included during the study period. Both groups were matched for age (DMD group 10 ± 3.7 years, control group 11.6 ± 3.3 years, p-value 0.17) and were not taking any cardiac medications previously. The DMD group had a significantly lower weight, height, and systolic blood pressure; however, heart rates were higher. Sixteen of the DMD group subjects used a wheelchair, and 12 of them were on steroids for an average of 10 months before the clinic visit. Global longitudinal strain (GLS) was lower in DMD patients compared with controls (-14.2 ± 5 vs. -18.8 ± 2 %, p-value: 0.001). Strain measured on apical 4-chamber and 2-chamber views was significantly lower in DMD patients compared with controls. Segmental longitudinal strains measured in segments 7, 12, 14, 15, and 17 were significantly lower in DMD patients than controls. All other segments showed no significant difference in DMD patients compared with controls.
Conclusion: Early detection of LV dysfunction using 2D-STE analysis is a quick and reproducible method in pediatrics. The existence of an early LV dysfunction visualized using 2D-STE analysis in children with DMD before the onset of cardiomyopathy represents a perspective for future pediatric drug trials in DMD-related cardiomyopathy prevention.

Keywords: Duchenne muscular dystrophy, cardiomyopathy, left ventricular dysfunction

P-446
Early preclinical experience of a mixed reality ultrasound system with active guidance for needle based interventions: the guide study

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Background and Aim: Ultrasound has become routinely used when obtaining vascular access, despite ergonomic and visualization challenges. MantUS system (Sentiar, Inc, St Louis, MO) uses a mixed reality (MxR) interface to display US images and integrate real-time needle tracking. This aim of this study was to prospective assess MantUS versus conventional US in a preclinical vascular access use case.
Methods: Study subjects were recruited from the Department of Pediatrics. Study tasks included obtaining vascular access in 2 trainer models, a femoral access model and head and neck model for 4 vascular access sites (right internal jugular vein [RIJ], right carotid artery [RCA], right femoral vein [RFV] and right femoral artery [RFA]) under 2 conditions: conventional US versus MantUS. Videos were obtained and analyzed. Quality of access was quantified.
Results: Use of MantUS resulted in overall reduction in number of needle repositions ($p = 0.03$), and improvement in quality of access as measured by distance ($p<0.0001$) and angle of elevation ($p = 0.006$). These findings were even more evident in the RFV access site, which was a simulated anatomic variant with a deeper more oblique vascular course. Here, use of MantUS resulted in faster time to access ($p = 0.04$), fewer number of access attempts ($p = 0.02$) and fewer number of needle repositions ($p<0.0001$) when compared to conventional US. Post-participant survey showed high levels of usability (87%) and a belief that MantUS may decrease adverse outcomes (73%) and failed access attempts (83%). Subgroup analysis showed some improvement in access with improvement in quality of access across all career stages. Additionally, users who performed vascular access infrequently or often had more improvement than those who obtain vascular access frequently.
Conclusion: Use of MantUS improved vascular access amongst all-comers, including the quality of access. This improvement was even more notable in the vascular variant (RFV). MantUS was an easy-to-use tool and provided improvement in spatial awareness. Subgrop analysis supported improved performance across all career stages, with less experienced users having more improvement. Further development of MantUS will focus on improving user interface and experience, with larger clinical user and in-human studies.

Keywords: Vascular Access Ultrasound Mixed Reality Vascular Variants

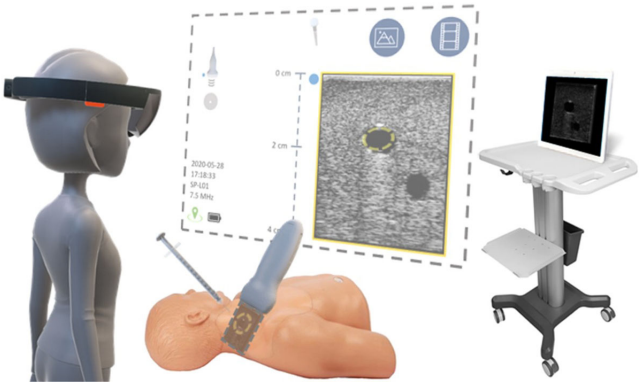

Figure 1. MantUS System in use

P-447/Moderated Poster
Hidden cardiovascular morbidity in children and young adults born with congenital diaphragmatic hernia: a population-based study

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Background and Aim: Congenital diaphragmatic (CDH) hernia is a rare congenital malformation with considerable mortality and morbidity in the neonatal period. The majority of the children today survive but little is known about long term cardiovascular morbidity.
Methods: This was a nationwide population-based prospective case-control study within a cohort of Swedish children with CDH, born 1982-2015. Five controls for each patient were randomly sampled from the population. The outcomes were the corresponding International Statistical Classification of Disease (ICD) codes for cardiovascular diagnoses according to ICD 9 and 10.
Results: There was an overrepresentation of cardiovascular diagnoses in the CDH group after one year of age compared to the control group, 8.0 %v s 0.5% (n = 53 versus n = 16). The risk of having a cardiovascular diagnosis in this CDH group was 15 times higher compared to the control group (HR 15.8, 95% CI: 9-27.6, $p < 0.005$). The diagnoses of cardiac arrhythmias and systemic hypertension were less common in the CDH group before the age of one year compared to the CDH group beyond the age of one year. Arrhythmia 3.7 % vs 15.1%, systemic hypertension 3.7 % vs 7.5%.
Conclusion: CDH survivors have increased cardiovascular morbidity during childhood and young adulthood. This implies that structured follow up programs, covering cardiovascular morbidity, needs to be developed and should be offered in pediatric and adult care. Being born with CDH seems to be a risk factor for future cardiovascular diagnoses.

Keywords: Ccngenital Diaphragmatic Hernia, Cardiovascular morbidity

P-448
Cardiac complications in two patients with distinct mitochondrial disorders

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Background and Aim: Mitochondrial disorders result in decreased energy production thereby affecting multiple organ systems. Cardiac manifestations typically include cardiomyopathy and conduction defects. The purpose of this presentation is to raise awareness of cardiac as well as extracardiac features of mitochondrial disorders using two examples, MELAS and TMEM 70 protein deficiency.
Methods: Two children with pronounced cardiac pathology were selected from our cohort of patients with mitochondrial disorders. Their extracardiac symptoms, echocardiographic, ECG and ambulatory ECG monitoring records are presented here.
Results: Patient one is a nine year old girl who suffers from MELAS syndrome caused by heteroplasmic mutation m. 13513G>A in MT-ND5 gene. Her first symptom was nystagmus noted during infancy. Within the first years of life she developed strabism, ptosis, myopathy and hypertrichosis. Her cardiac pathology includes hypertrophic cardiomyopathy and preexcited ECG with no runs of supraventricular tachycardia documented so far. She has suffered from a stroke-like episode recently causing a significant regression in her psychomotor skills and deterioration of cerebellar symptoms. The second patient is a ten year old boy with TMEM 70 protein deficiency leading to ATP synthase dysfunction. The diagnosis was established at the age of four months during a metabolic crisis, which was triggered by prolonged fasting. It was associated with hyperammonaemia and lactic acidosis. Other features typically related to this condition include history of oligohydramnion, hypospadia and delay of psychomotor development. His cardiac findings include mixed hypertrophic/noncompaction cardiomyopathy and preexcited, extremely high voltage ECG with left axis deviation and intermittent LBBB morphology. He has never had a syncope or documented ventricular or supraventricular tachycardia.
Conclusion: Cardiac manifestations in both patients include hypertrophic cardiomyopathy and pronounced conduction defects. Due to the rarity of both diagnoses, no international guidelines on the management of their cardiac complications exist. Hence, we consider it important to discuss difficult issues, such as risk stratification, preexcitation vs. pseudo-preexcitation and indications for catheter ablation or even ICD implantation in the context of these multisystemic diseases within a broader community of experts.

Keywords: cardiomyopathy, conduction defect, mitochondrial disorders

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P-449

Vascular phenotyping in post-COVID children and adolescents - the lico study

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Background and Aim: An infection with SARS-CoV-2 is associated with systemic inflammation, that also affects the endothelium. This may result in endothelitis, which can influence vascular regulation and morphology. Until now, the specific mechanism of vessel damage after a SARS-CoV-2 infection is still unclear, especially in children and adolescents. The LICO Study (Long term impact of COVID-19) aims to investigate the long-term effects of a SARS-CoV-2 infection on vascular structure and function in children and adolescents.
Methods: Children and adolescents with confirmed evidence of survived SARS-CoV-2 infection are screened 6 ± 3 months post-infection. Vascular function is assessed by flow-mediated vasodilation (FMD) and aortic pulse wave velocity (PWV). Carotid intima-media thickness (cIMT) and retinal diagnostics (arteriovenous ratio - AVR) are used to examine vascular structure. The matched control group without prior SARS-CoV-2 infection undergoes the same examination procedure.
Results: So far, we have been able to evaluate 24 (9 post-covid) subjects (13.5 ± 1.9 years; 9 girls). Compared to the mean reference values of the control group, 5 post-covid subjects have higher cIMT (0.49 ± 0.01 mm vs. reference value 0.46 ± 0.03 mm). Of these, 3 post-covid subjects even deviate from the norm PWV (4.96 ± 0.16 m/sec vs. reference value 4.63 ± 0.29 m/sec). The same 3 post-covid subjects are also below the norm FMD (2.06 ± 1.05 % vs. reference value 4.18 ± 7.04 %). None of the post-covid subjects deviates from the norm AVR values (reference value 0.85 ± 0.07).
Conclusion: It is shown that infection with SARS-CoV-2 has the potential to impair vascular regulation. These initial results provide trends for early vascular changes among children and adolescents after recovered SARS-CoV-2 infection. Due to that this is an ongoing study, the results are constantly being expanded and may still change. To determine lasting changes in morphology, the examination is repeated after 6 months and the further results of this longitudinal study must be awaited.

Keywords: COVID-19, SARS-CoV-2, vascular function, vascular structure, inflammation, children and adolescents

P-450

Acute phase of kawasaki disease: a systematic review of national guideline recommendations

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Background and Aim: Regarding Kawasaki disease (KD), key aspects are not yet supported by a high evidence level, thus making room for individual recommendations. We conducted a structured comparison of existing international KD guidelines to analyse potential differences in the implantation of evidence-based disease specific diagnosis and therapy within existing recommendations.
Methods: To identify country-specific guidelines, we took a multilateral approach including a comprehensive PubMed literature, online research and directly contacting national paediatric associations. We then performed a structured guideline analysis and evaluated the diagnostic and therapeutic differences in the context of evidence-based medicine.
Results: We identified nine country-specific and one European Kawasaki disease guideline. According to them all, diagnosing Kawasaki disease still relies on the clinical presentation and no additional conclusive laboratory test. First-line treatment with intravenous immunoglobulins (IVIG) is consistently recommended in all guidelines. However, recommendations regarding aspirin, corticosteroids, and further therapeutic options vary considerably. In particular, guidance regarding intensified first-line therapy such as additive corticosteroids should rely on different risk variables. None of the current guidelines, however, recommend initial corticosteroids in all Kawasaki disease patients.
Conclusion: According to all guidelines, Kawasaki disease is diagnosed clinically and treated first with IVIG. Recommendations for additional therapeutic strategies are more heterogeneous.

Keywords: Kawasaki disease, National guideline, Coronary artery aneurysm

P-451

A familial history of histiocytoid cardiomyopathy

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Background and Aim: Histiocytoid cardiomyopathy (HC) is a very rare disorder that occurs in the first two years of life. It manifests with complex arrhythmias and sudden deaths. Degeneration of Purkinje cells and abnormal cardiac myocytes are the main histopathologic findings. Few cases are described, questions remain about cardiac and neurologic outcomes in patients with HC.
Methods: Here, we report the case of two siblings with cardiac and unusual neurologic features.
Results: The oldest was hospitalized at 9 months after sudden death. She was diagnosed with refractory ventricular tachycardia and was put on ECMO. Ventricular function was reported initially normal. Decision to withdraw care was made after a diagnosis of cerebral death. Histopathological diagnosis of HC was obtained during post-mortem autopsy. Her youngest sister presented antenatal recurrent arrhythmias, mild heart dilation and pericardial effusion. She was treated with maternal digoxin intake. Pregnancy was otherwise uneventful. At birth, neonatal EKG showed Wolff Parkinson White (WPW). Cardiac ultrasound (CU) demonstrated left ventricular (LV) apical hypertrabeculation despite normal ventricular function. Her sister's CU retrospectively showed the same findings without WPW on EKG. Next generation sequencing on both sisters and mother identified a pathogenic variant in NDUFB11, a X linked complex I deficiency. Holter monitoring revealed auto-resolutive episodes of ventricular tachycardia. CU showed a progressive decrease in cardiac function and worsening mitral regurgitation. ACE inhibitors and beta blockade were started. Trans-esophageal atrial stimulation confirmed a short refractory period malignant pathway. Patient evolved through arythmogenic storm at 7 months with intensive care management controlled with maximal doses of amiodarone, flecainide and beta-blockade. Outcome gradually improved within months. Surprisingly, patient developed refractory eyelid myoclonic epilepsy with absences. She demonstrated mild developmental delay (gross motor and language). Brain MRS was normal without sign of mitochondrial manifestation. She did not demonstrate any dysmorphic features other than short stature and growth delay. EEG

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showed burst of generalized 4hz spikes and waves during seizures without interictal abnormalities.
Conclusion: Patient is now 4 years old with normal ventricular function, same LV apical hypertrabeculation without recurrent arrhythmias. Cardiac treatment has been decreased slowly over a year and is now almost weaned off.

Keywords: histiocytoid cardiomyopathy

P-452
Pediatric cardiac tumor in a tertiary center

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Background and Aim: Cardiac tumors are rare in children. Unlike in adults most are benign and asymptomatic. The majority of primary cardiac tumors can be managed conservatively. This study aims to analyze the characteristics and outcome of pediatric patients with cardiac tumors treated in our center.
Methods: We performed a 15 years retrospective analysis of 27 children (<16 years) diagnosed of cardiac tumor in our institution.
Results: There were 14 males and 13 females, median age at diagnosed of 9 months. Ten patients were younger than 1 year and 10 (37%) were diagnosed prenatally. 25 tumors were primary and benign (14 rhabdomyoma, 7 fibroma, 1 hamartoma, 1 lipoma and 1 indeterminate) two cases were secondary and malignant.
22 (81%) patients were asymptomatic at diagnosis. 2 had chest pain and 3 showed low cardiac output signs. 8 patients (29.6%) developed arrhythmia in the follow up. The diagnosis of cardiac mass was made by Echocardiography in all patients. 19 magnetic resonance image (RMI) studies were performed in 16 patients (59%), 13 to complete the diagnosis and 6 to follow up. 16 patients (59%) did not required treatment; Surgical removal was indicated in four patients due to left ventricle outflow tract obstruction (LVOTO), right ventricle outflow tract obstruction and superior vena cava compression. Although biopsy is the diagnostic gold standard it was only available in 6 patients. Two patients with left ventricle fibroma underwent heart transplant due to severe dysfunction. 10/14 patients with rhabdomyoma were diagnosed of tuberous sclerosis (71%), 3 required additional therapies (2 received m-TOR-inhibitors, one for arrhythmia and LVOTO and other for refractory epilepsy; 1 needed neonatal surgery due to LVOTO. During the follow up 3 of the rhabdomyomas had fatty degeneration in MRI and 1 patient experimented spontaneous regression of the tumor after 6 years. Overall survival was 26 patients (96.3%).
Conclusion: Pediatric cardiac tumors are predominantly primary and benign. Rhabdomyoma is the most common tumor, strongly associated with Tuberous Sclerosis. Most patients are asymptomatic and echocardiography provide consistent assessment of anatomy and function completed by MRI. Surgical resection should be required with good results in symptomatic patients.

Keywords: Cardiac tumors, rhadomyoma, tuberous sclerosis

P-453
Cardiopulmonary exercise test in children with pediatric inflammatory multisistemic syndrome associated with COVID-19 (PIMS)

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Background and Aim: Coronavirus infection (COVID-19) in paediatric population has a generally mild course. In Spain, patients under 15 years old have accounted only for 0,4% of hospital admissions and 0,7% of intensive care admissions. However, in May 2020, cases of children with a systemic inflammatory syndrome related to a recent COVID-19 infection were described. In severe forms, left ventricular systolic dysfunction, mitral regurgitation, pericardial effusion and coronary artery dilatation or aneurysms have been described. The aim of this study is to describe the results obtained in cardiopulmonary exercise test (CPET) in previously healthy patients with PIMS.
Methods: Prospective study of PIMS patients who performed CPET. Godfrey ramp protocol recommended by European Society of Cardiology (ESC) was used in all cases. Measured variables, expressed by predicted values, were: forced vital capacity (FVC), forced expiratory volume (FEV1), ratio of minute ventilation to carbon dioxide production (VE/VO2 slope), maximal oxygen consumption (VO2 max), oxygen uptake efficiency slope (OUES), oxygen pulse (O2 pulse) and maximum heart rate (HR).
Results: Eight patients (75% boys) aged 5-14 years (median 10,5 years) performed CPET reaching a mean peak load of 105,87 W (median 112,5 W and mean load per kg of weight 2,34 W/kg). Only 1 patient (12,5%) presented basal spirometric disturbances in context of asthma without chronic treatment. Obtained mean respiratory parameters were: FVC 97,88%, FEV1 92,7%, Tiffeneau 83% and VECO2p 32,47. Oxygen saturation before and after CPET was greater than 95% in 100% of patients. In 6 patients (75%) the V02max and oxygen pulse was greater than 80% of predicted value (100% of patients reached at least 40% of V02 max at anaerobic threshold). Obtained mean cardiovascular parameters were: VO2 max 1624mL/min (median 1655 ml/min and V02 per kg of weight 36,9 ml/kg), pulse oxygen 9 ml and OUES 1,92.
Conclusion: PIMS may cause severe cardiac disturbances justifying cardiological monitoring of these patients. CPET allows to assess functional capacity of these children after the disease. In our serie, most of patients had a good functional capacity (75%). Studies with more patients are needed to make extended conclusions.

Keywords: Cardiopulmonary exercise test, Pediatric Inflammatory Multisistemic Syndrome.

P-454
High frequency intraoperative coronary imaging, is there anything to gain?

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Background and Aim: Coronary arteries (CA) are one of the smallest vascular structures which can be delineated using two-dimensional (2D) echocardiography. Abnormalities in CAs occur in up to 1% of the population and are associated with significant morbidity and mortality. Intraoperative CA imaging is commonly performed using transesophageal or epicardial imaging via standard probes (7 -12 MHz) however 2D assessment is often limited due to their low spatial resolution. The aim of this study was to determine the feasibility of using high frequency (Hf) ultrasound for intraoperative coronary assessment.
Methods: In this prospective, non-randomized observational study, paediatric patients undergoing surgery involving one of more CAs were enrolled. Patients underwent research only epicardial

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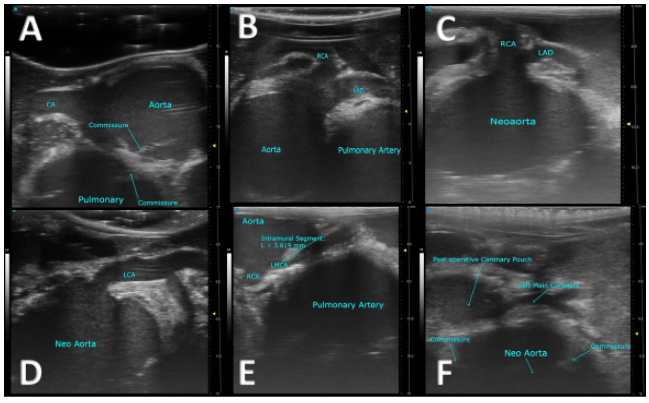
imaging immediately prior and after surgical repair using a 48 MHz probe. Routine transesophageal and/or epicardial imaging was performed post-surgery. All epicardial images were acquired by the responsible surgeon.
Results: A total of 16 patients were included (1 ALCAPA; 15 Transposition of the Great Arteries (TGA)) with a median age of 6 days. Of the TGAs, 8 had usual coronary arrangement (1LCx2R), while 8 had anatomical variations and two of these had intramural segments of the left main coronary. One patient required surgical revision prior to chest closure (intramural) a second patient (ALCAPA) required surgical revision day 4 post op. High frequency imaging was performed on all patients. Pre-operative imaging was able to confirm, delineate, and quantify an intramural course. Post-operative imaging provided improved resolution for clear anatomical assessment coronary ostia and proximal segments (see image 1).
Conclusion: High frequency epicardial imaging for coronary assessment is feasible and may provide valuable information to assist in surgical strategy and post-operative review.

Keywords: Coronary, High Frequency, Epicardial, Ultrasound, Surgery

Image 1



(A) Pre op imaging in a patient with TGA showing an unobstructed coronary orifice well away from the commissure. (B-C) Pre and post-operative imaging in a TGA patient with a single origin of the RCA and LAD (1LR2Cx). (D) Post op imaging of a patient with TGA and usual coronary arrangement (1LCx2R) (E-F) Imaging in a TGA patient with an intramural left main coronary and adjacent RCA. Post–op imaging of this patient (F) demonstrates the newly formed coronary pouch, its relationship to the neo-aortic commissure and an unobstructed left main.

P-455 / Moderated Poster

Oscillometric pulse-wave-velocity measurement in adult and pediatric cohorts shows an excellent accuracy – an evaluation study of the mobil-o-graph

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Background and Aim: Increased arterial stiffness raises the cardiovascular risk. The arterial pulse wave velocity (PWV) is a surrogate marker for arterial vessel rigidity. Non-invasive PWV measurement methods are available and have been validly evaluated in adult but not pediatric cohorts. The purpose of this study was to validate the Mobil-O-Graph, an oscillometric measurement method in children, adolescents and young adults.
Methods: 61 patients (25 men, mean age 16,7 years; range 3-35 years) were included in this study. 52 patients underwent cardiac catheterization for evaluation after heart transplantation and nine for interventional ASD-closure. To assess the invasive aortic PWV, pressure pulse waves were simultaneously documented via a multipurpose catheter in the aortic position and the femoral arterial sheath. A simple mathematical formula (velocity = distance/time) was used to calculate the PWV as follows: PWV = distance between catheter-tip and catheter-exit at the sheaths´ hemostatic valve (Figure 1a)/time delay between both pulse waves (Figure 1b). The invasive PWV results were compared to simultaneously measured brachial cuff readings using Mobil-O-Graph stratified by age (OVERALL | PEDIATRICS <18 | ADULTS ≥18).
Results: Pearson and Spearman correlation analysis showed a positive linear distribution between both PWV measurements (OVERALL: r = 0,628, p<0,001 | PEDIATRICS: r = 0,591, p<0,001 | ADULTS: r = 0,582, p = 0,002). Bland altman analysis revealed a mean difference between both PWV-measurement methods of 0,44 ± 0,55 m/s in the OVERALL-group (95% limits of agreement -0,65 - 1,52 m/s). When stratifying for age, mean difference was 0,41 ± 0,41 m/s in the PEDIATRICS-group (95% limits of agreement -0,39 – 1,21 m/s) and 0,42 ± 0,71 m/s in the ADULTS-group (95% limits of agreement -0,92 – 1,86 m/s). Validation criteria according to artery guidelines are excellent if accuracy is ≤ 0,5m/s ± 0,8 and acceptable if accuracy is <1,0m/s ± 1,5m/s.
Conclusion: Estimated PWV using the Mobil-O-Graph showed an excellent accuracy in adults and pediatric cohorts according to the artery society guidelines and is comparable to an evaluation study in adults. Therefore, the Mobil-O-Graph can be implemented as an ambulatory PWV measuring tool to early predict vascular changes in children and adolescents as well.

Keywords: PWV, Pulse-wave-velocity, pediatrics, Mobil-O-Graph, noninvasive measurement, oscillometric measurement

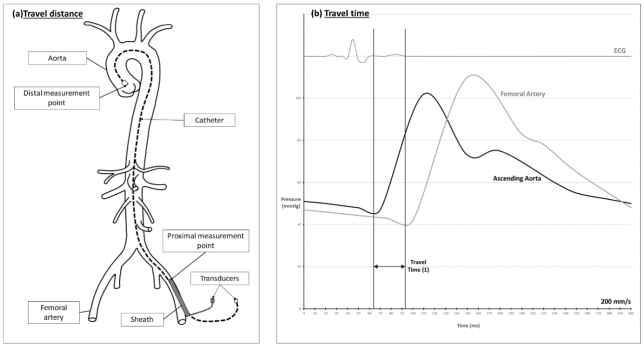

Figure 1. (a) Calculating travel distance between both measurement points. (b) Calculating travel time between both pulse waves.

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P-456

Amino acid metabolism as an indicator of inflammation and subtle cardiomyopathy in patients with marfan syndrome

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Background and Aim: Patients with Marfan syndrome (MFS) have an increased risk of aortic aneurysm formation, dissection, and myocardial impairment. We analyzed amino acid metabolic pathways in adult MFS patients with good systolic biventricular function, without major valve disease, and heterogeneous in terms of presence or absence of prior aortic root surgery and type of surgical approach, seeking biomarker patterns as monitoring tools complementary to imaging studies in early cardiovascular risk assessment with regard to deterioration of myocardial function.
Methods: Comparison of traditional imaging and laboratory values and mass spectrometry-based amino acid metabolomes in 24 adult MFS patients with those in healthy controls. Analytes for which values differed between patients and controls and baseline cardiovascular characteristics as assessed by echocardiography and cardiac magnetic resonance imaging were subjected to uni- and multivariate regression analysis.
Results: A high proportion of patients had signs of impaired diastolic function and elevated serum levels of NT-proBNP. Patients had lower serum levels of taurine and histidine than controls. The variables evidence of diastolic dysfunction, history of aortic root surgery, and aortic root dimensions correlated significantly with the serum concentration of NT-proBNP and taurine.
Conclusion: Incipient cardiomyopathy is present in a high proportion of MFS patients. Alterations in serum levels of isolated amino acid metabolism derived analytes link MFS pathophysiology with altered inflammation, oxidative stress, endothelial dysfunction, and subtle changes in myocardial properties. Particularly the amino acid taurine might play a future role in the detection of possible targets for therapeutic approaches to mitigate inflammation in propagating ventricular deterioration in MFS patients.

Keywords: cardiomyopathy, Marfan syndrome, metabolomics, taurine

P-458

A rare congenital venous anomaly as a potential predisposing factor for atrioventricular nodal reentry tachycardia in an adolescent

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Background and Aim: Persistent left superior vena cava (PLSVC) with absent right SVC, also known as "isolated PLSVC", is found in 0.05-0.07% of the general population. This condition may be associated with cardiac arrhythmias. It is reported that a stretching of the atrioventricular node and the bundle of His due to coronary sinus dilatation as well as a persistence of remnant pacemaker tissue in the LSVC as an ectopic trigger or an arrhythmia substrate might be related to arrhythmogenic effects of the LSVC.
Methods: We report on an adolescent presenting with palpitations who was diagnosed with typical atrioventricular nodal reentry tachycardia in the presence of an isolated PLSVC.
Results: A 15-year-old male with documented narrow-QRS-complex tachycardia was admitted to our unit for catheter ablation. Physical examination was normal. A 12-lead ECG showed sinus rhythm. Transthoracic echocardiography demonstrated normal right and left ventricular function, a dilated coronary sinus as a typical finding of LSVC and the absence of the right SVC. During the cardiac electrophysiologic study with non- fluoroscopic approach guided by three dimensional electroanatomic mapping system, a quadripolar and a steerable decapolar diagnostic catheter were placed in the His bundle position and into the coronary sinus via the left femoral vein, respectively. Mapping confirmed the echocardiographic finding of absence of the right SVC and the enlarged coronary sinus. The steerable decapolar diagnostic catheter was advanced effortless through the coronary sinus into the LSVC to the junction of the left jugular and left subclavian vein. After the patient was diagnosed with slow-fast reentry tachycardia and had successfully and uneventfully undergone radiofrequency catheter ablation of the slow pathway with no fluoroscopy, we performed angiograms to exclude further associated congenital cardiac anomalies. It also confirmed the drainage of the LSVC into the right atrium via the coronary sinus and the absence of the right SVC. Magnetic resonance imaging of the thorax showed the normal cardiac anatomy and excluded further venous anomalies (Figure 1).
Conclusion: Isolated PLSVC is a rare variant anomaly of PLSVC and may be an arrhythmogenic factor that should be considered in all patients with PLSVC. Further cardiacvascular anomalies should be excluded in these patients.

Keywords: persistan LSVC, venous anomaly, coronary sinus

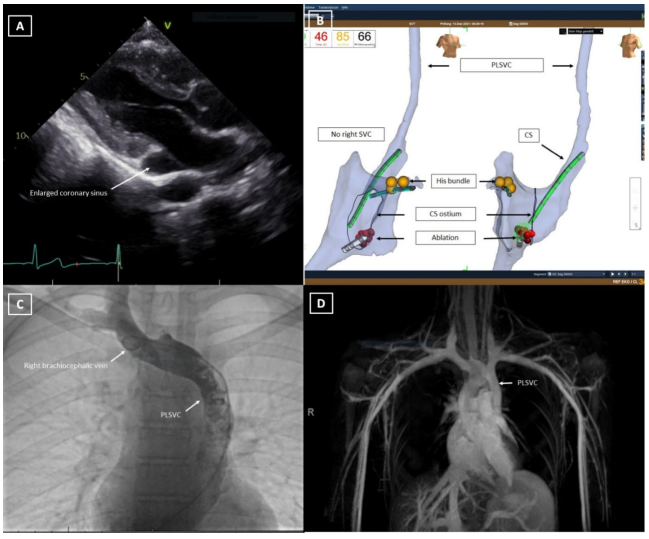

Figure 1. Enlarged coronary sinus (CS) in the parasternal long axis view by transthoracic echocardiography (A); persistent left superior vena cava (PLSVC) and enlarged CS ostium (dark oval circle) by 3D-electroanatomic mapping system (B), selective angiography (C) and magnetic resonance imaging of the thorax (D).

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Total anomalous pulmonary venous connection: 50 years review into clinical and surgical outcomes in south wales

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Background and Aim: We aimed to evaluate presentation, diagnosis, morphology, genetic background, type of surgery, complications, and outcome of patients with total anomalous pulmonary venous drainage in South Wales during a period of 50 years.
Methods: A retrospective review was conducted for all patients diagnosed with total anomalous pulmonary venous drainage in South Wales between 1971 and 2021 (n = 89). Local paediatric cardiology and (1971-2021) and national (CARIS) databases were queried. The cohort was divided into single ventricle (SV) and non-single ventricle (non-SV). Patient characteristics, in-hospital and follow up outcomes were defined.
Results: Of the 89 live-born patients, there was male predominance with 56 male to 33 female patients. Supracardiac type was seen in 36 (40 %), infracardiac 23 (25%), cardiac 16 (18%), and mixed 6 (6%) cases. Common pulmonary vein atresia was identified in one, and two patients had extremely hypoplastic veins but in seven cases no specific location was defined. Eight patients had genetic abnormalities. 72 patients had isolated TAPVD and 17 had TAPVD within complex cardiac anomalies; 11 with right atrial isomerism, and six had other associated complex cardiac conditions. 10 patients had repeat interventions of which six required relief of pulmonary vein obstruction. 18 patients died; five patients with isolated TAPVD and 13 with complex heart conditions. 64 patients remained alive and well but seven had no outcome data.
Conclusion: Although TAPVD is known to carry a low chance of genetic and familial inheritance, in our cohort eight cases exhibited genetic syndromes and two had familial inheritance. Antenatal diagnosis for isolated TAPVD remained challengingly low. Small percentage of patients developed late postoperative complications without increased mortality. Our study showed good survival rate of patients with isolated TAPVD with 64 out of 71 patients being alive and well with high mortality only among patients pertaining to right isomerism and complex heart problems.

Keywords: TAPVD, supracardiac, infracardiac, coronary sinus, surgery.

P-460

Quality of life of children born with a congenital heart defect: a population-based cohort study

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Background and Aim: To describe the health-related quality of life (QoL) at 8 years of age of children born with congenital heart defects (CHD) and to identify subgroups at risk of health-related quality of life (QoL) impairment at 8 years of age according to their medical and surgical management (MSM)
Methods: From a prospective population-based cohort study, CHD associated with chromosomal abnormalities or malformations (non-isolated CHD) were separated from isolated CHD. Then the isolated CHD group was subdivided in 5 subgroups according to their MSM: 1) CHD followed-up in an outpatient clinic, 2) Complete repair before 3 years, 3) Complete repair after 3 years, 4) Palliative repair and 5) CHD with spontaneous resolution (reference subgroup). Self- and parents-reported QoL was measured using the PedsQL4.0 Generic Core Scales (range 0-100) at 8 years of age. Multivariable regression analysis (β) and Cohen's effect size (δ) were used to compare the outcomes across the CHD subgroups.
Results: 751 children were included, 598 (79.6%) had isolated CHD and 153 (20.4%) had non-isolated CHD. For non-isolated CHD subgroup, self and parents reported QoL scores were lower than the isolated CHD subgroup (73.9 ± 0.4 vs 75.8 ± 0.2 $p < 0.001$ and 70.9 ± 1.5 vs 82.7 ± 0.6 $p < 0.001$, respectively). Self and parent-reported QoL scores for the palliative repair subgroup were lower (β = - 2.1 [CI95%: -3.9; -0.2] and β = -16.0 [CI95%: -22.4; -9.5], respectively) with a large effect size δ = -0.9 [CI95%: -1.4: -0.4] and δ = -1.3 [CI95%: -1.8: -0.7], respectively. Parent-reported QoL score for the complete repair after 3 years subgroup was lower (β = -9.2 [CI95%: -15.0; -3.5]) with a large effect size (δ = -0.9 [CI95%: -1.4: -0.5]). Self-reported QoL score for the complete repair before 3 years subgroup was lower (β = -1.3 [CI95%: -1.9; -0.6]) with a small effect size (δ = -0.4 [CI95%: -0.6 -0.2]).
Conclusion: QoL of children with CHD who experienced a hospital intervention is reduced at 8 years of age. The age of the last cardiac intervention might influence the QoL at 8 years of age.

Keywords: Congenital heart defect, quality of life, population-based cohort study

P-461

Pattern of congenital heart disease in sudan (a single center experience)

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Background and Aim: Although Congenital Heart Disease represent a major cause for mortality and morbidity in our population, Congenital heart disease services officially started in Sudan in the year 2000. As a wide spectrum of congenital heart diseases dose present to our clinic, this study was carried out. The aim of this study is to shed light on the pattern, age, and sex distribution of congenital heart disease patients.
Methods: 1,009 patients between the age of 2 days to 70 years old were seen at the pediatric cardiology clinic over a period of 17 months. Currently, more assessment are ongoing and results are being accounted for. Clinical assessments and echocardiography were performed for all patients as the assessment tools.
Results: With respect to age, less than 5% of the studied population were adults. On the other hand, the Male to female ratio was 48% to 52%. The most frequently seen defect in acyanotic heart disease is Ventricular Septal Defect (VSD). This was followed by Common Atrio-ventrcular (AV) canal. The most frequent

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cyanotic congenital heart disease is Tetrolog of Fallot. Double Outlet Right Ventricle (DORV), Transposition of Great Arteries (TGA), Total Anomalous Pulmonary Venous Drainage (TAPVD), and other more rare congenital heart disease such as Single Ventricle physiology are seen not uncommonly. Due to the long queue for surgery, 10% of the studied population die waiting their turn while only 15% of the studied population made it to surgery. Although few patients get sent for surgery abroad, not all can afford the very high cost.
Conclusion: As this study reflects the experience of one center, a multi-center study should take place in the form of an ongoing Congenital Heart Disease Register. This will mostly help plan for better services regarding the management of Congenital Heart Diseases.

Keywords: Congenital, Cyanotic, Acyanotic, Heart Diease

P-462

Recurrent pericarditis associated with postpericardiotomy syndrome in a 6 year old girl

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Background and Aim: In children, recurrent pericarditis occurs in 30% of cases. In 80% of the cases the etiology remains unknown. The prognostic outcome of recurrent pericarditis is good. Nevertheless, it remains a burden for the patients and their parents due to frequent hospitalisations. Furthermore, the management of recurrent pericarditis remains a challenge to doctors.
Methods: The case of a 6-year-old female patient with multiple hospitalizations to pediatric department of the tertiary hospital due to recurrent pericarditis is presented.
Results: The patient was born pre-term, as a second twin, after being diagnosed with Twin Reversed Arterial Perfusion Syndrome. At the age of 5 she underwent an atrial septal defect surgery. The patient first experienced an episode of pericarditis in July 2020, 18 months after the procedure. During this episode the patient experienced fever, nausea, sharp pain in the chest and left shoulder. Blood tests showed leukocytosis, increased reactive protein C levels (156g/l). Creatine kinase-MB levels (0,28ug/l) and troponin (0ng/l) levels were normal. B-type natriuretic peptide (173,7ng/l) was normal. Furthermore, the ST segment was elevated in majority of electrocardiogram leads and pericardial effusion was present during echocardiography. An X-ray showed pleural effusion. The treatment with non steroidal inflammatory drugs (NSAID's) was started. After the first episode of pericarditis, the patient experienced 4 separated episodes within 9 months. Every episode manifested with fever and pain in the chest and left shoulder, increased reactive protein C levels, ST segment elevation and pericardial effusion. antinuclear antibodies, anti-neutrophil cytoplasmic antibody tests, blood test for tuberculosis were negative. Genetic testing for Tumor Necrosis Factor Receptor associated Periodic Syndrome, Familial Mediterranean Fever, mevalonate kinase deficiency were negative. One month later, after the second episode, treatment with steroids was started. Colchicine was added after the third episode, in October, 2020. Despite the triple treatment with NSAID's, colchicine and steroids the patient had 2 more recurrences of pericarditis in December and March, 2021. Due to the recurring episodes whilst treating with colchicine, the patient is now a candidate to IL-1 receptor antagonist therapy. The etiology of pericarditis remains unknown until now.
Conclusion: The inadequacy of diagnostic criteria does not exclude the postpericardiotomy syndrome.

Keywords: idiopathic recurrent pericarditis, children

P-463

Acute myocardial infarction in a 9-year-old boy due to multisystem inflammatory disease

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Background and Aim: Myocardial infarction after coronavirus disease 2019(COVID-19) is a quite uncommon clinical disease in children. We present a case about a 9-year-old boy with total occlusion of the right main coronary artery(RMCA) attending to the hospital with chest pain. It was related pediatric multisystem inflammatory disease (MIS-C).
Methods: It is a case presentation.
Results: Case Presentation:
The patient had no previous cardiac or family history. Electrocardiography(ECG) showed a definite elevation on the extremity derivatives(DI, DII, DIII, and aVF), and marked ST depression on the chest derivatives (V1 to V6) and aVR, aVL, all representing lateral inferior ischemia. Transthoracic echocardiography revealed left ventricular systolic dysfunction, and global wall hypokinesia. Existence of fever and two-body system involvement (cardiac, gastrointestinal), CRP rise, and prior SARS-CoV-2 exposure in a month, MIS-C was a foremost diagnosis. The total antibody for SARS-CoV-2 was positive. Lipid profiles(LDL, HDL, VLDL, triglyceride), lipid electrophoresis, routine coagulation, and thrombophilia tests were evaluated for differential diagnosis and all were normal. Because of the possible MI, it was planned to visualize coronary arteries by angiography. Total occlusion of the right main coronary artery(RMCA) with a large thrombus was detected without any dilatation of the coronary arteries in the coronary angiography. Two coronary stents were implanted into the distal and proximal part of the RMCA. After the procedure, clopidogrel was added to acetylsalicylic acid for platelet inhibition. During the follow-up, LVEF rose to 55% and there was a little hypokinesia on the left inferior wall of ventricles.
Conclusion: . It should be kept in mind that acute coronary thrombosis could be an important complication of COVID-19 exposure or MIS-C. A coronary stent implantation is a good treatment option even in small children.

Keywords: coronavirus disease 2019, myocarditis, myocardial infarction, children

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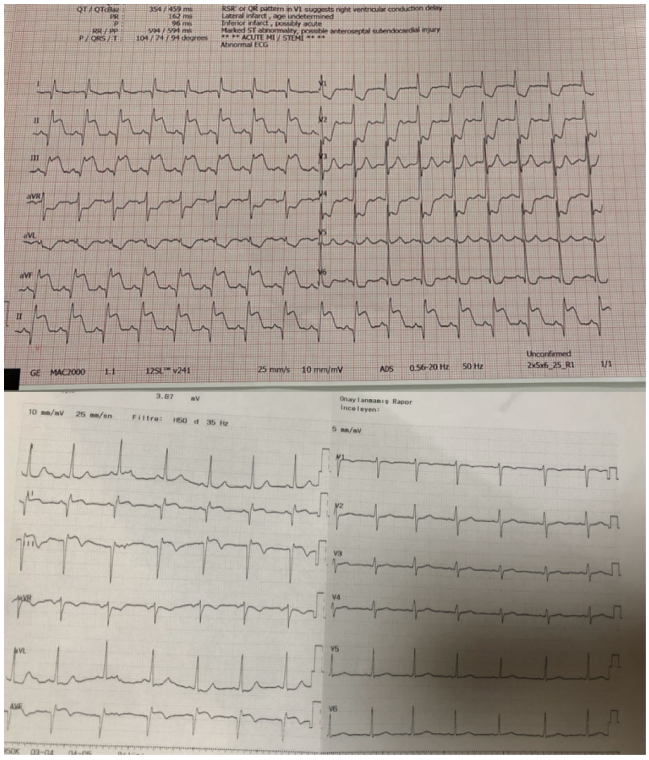

Figure 1. Lateral myocardial infarction findings on ECG on the top and ECG findings at discharge at the bottom.

P-464

Myopericarditis, the disease of predominantly male adolescents: single center experience on clinical, laboratory features, and outcomes

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Background and Aim: Myopericarditis is a primarily pericardial inflammatory disease and is often accompanied by varying degrees of concomitant myocardial involvement.
Methods: In resent study; clinical features, laboratory and imaging findings, and, outcomes over a 10-year-period (2010-2021) of 45 patients younger than 18 years old diagnosed as myopericarditis at the Ankara University School of Medicine, Department of Pediatric Cardiology were retrospectively analyzed.
Results: Patients were predominantly male (42 male / 3 female). Median age at diagnosis was 13.5 years (8-17 years). The most frequent presenting symptoms were chest pain (98%), fatigue (11%), palpitation (9%), and fever (16%). Recent history of infection was recorded in 28 patients (62%). Respiratory viral panel testing was available in 31 patients and identified 15 viruses in 9 patients. Electrocardiography revealed and localized ST segment elevation and T wave inversion in 24 patients (53%). Echocardiographic examination showed normal systolic and diastolic functions in all patients and detected minimal pericardial effusion in only 4 patients. All of the patients had mild to severe increased levels of troponin on admission. Troponin levels returned to normal in 1 to 12 days (median 6 days). White blood cell count, C-reactive protein, and/or erythrocyte sedimentation rates were elevated in 42 patients. Cardiac MRI was performed in 25 of the patients and 12 of them showed abnormal contrast enhancement. 24-hour Holter monitoring carried out in 12 patients, infrequent premature ventricular contractions were found in three of them. The median hospital stay was 4 days (2-10 days). The median length of follow-up was 2 months (1 week-9 years). All patients were well during follow-up.
Conclusion: This is the largest single-center study from Turkey on clinical presentation and follow-up of myopericarditis. Sometimes it can be difficult to differentiate the myopericarditis from the acute coronary syndrome because of the nature of the presenting acute chest pain. However, most cases of acute chest pain accompanied with troponin elevation, especially in adolescent males could be related to myopericarditis. Considering the its benign and self-limiting nature, invasive coronary angiography to exclude acute coronary syndrome is unnecessary in most of the patients. Gadolinium-enhanced cardiac MRI is a powerful diagnostic tool in such cases.

Keywords: Myopericarditis, chest pain, adolescent, troponin

P-465

Successful resolution of left ventricular thrombuses in a patient with MIS-C

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Background and Aim: A 15 year old young man with symptoms and signs consistent with MIS – C was admitted to the Intensive Care Unit for inotropic support as he was exhibiting signs of cardiogenic shock. He was previously fit and healthy and he had been exposed to Covid 19 confirmed cases 6-8 weeks prior to becoming unwell.
Methods: The patient received IVIG and steroids as an immunomodulating regime. On the admission echocardiogram there was a structurally normal heart with large LV thrombuses. The D-Dimers were extremely elevated on admission and the patient received therapeutic heparin infusion. Other prothrombotic causes were excluded.
Results: The surveillance echocardiogram 24h post admission showed resolution of the thrombuses. The patient never exhibited any signs or symptoms of cardiac ischaemia on the electrocardiogram or regional wall motion abnormality on the echocardiogram or neurologic impairment and the brain MRI-MRA one week post admission was normal. The patient was discharged home 5 days post admission and on follow ups up to a year after the acute phase remains very well physically and clinically.
Conclusion: Thromboembolic events are frequently described in COVID-19 patients and in some patients with MIS-C and are the consequence of a hyperinflammatory response and endothelial dysfunction. There might be a potential role of an antiphospholipid syndrome secondary to severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) infection as has been proposed. An increase in D-dimer level has been shown to be associated with thromboembolic events, including arterial thrombosis especially in the older population and should be investigated promptly. With the appropriate immunomodulation and antithrombotic treatment adverse events are prevented. More studies to assess

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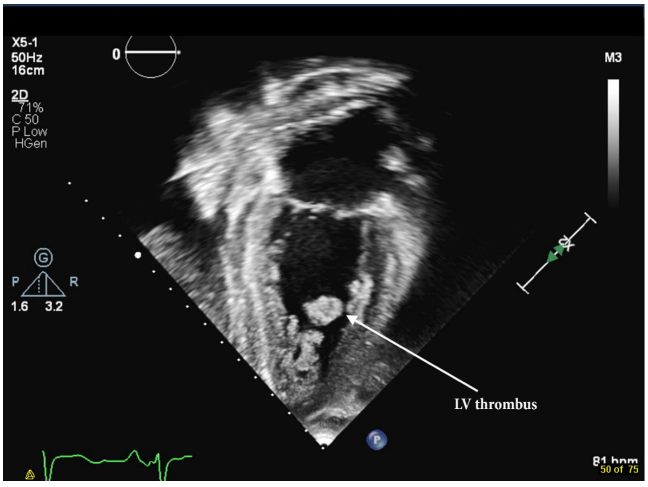
endothelial function and its role in the MIS-C prothrombotic state are necessary.

Keywords: MIS-C: Multisystem inflammatory syndrome in children

Left Ventricular thrombuses



Apical 2 chamber view clearly demonstrating the LV thrombuses

P-466

Parental views on neurodevelopmental follow-up visits and early interventions in children after open-heart surgery during infancy

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Background and Aim: Congenital heart disease (CHD) is the most common birth defect affecting approximately 6-8:1`000 life born children. In Switzerland, developmental follow-up (FU) assessments at 1,2 and 5 years of age are recommended for children with CHD and open-heart surgery within the first year of life. We investigated the compliance, and parental views on FU and early interventions (EI).

Methods: Out of 201 children with CHD (born in 2015/2016) in whom cardiopulmonary bypass surgery was performed atour institution in the first year of life, 26 were excluded due to death (n = 21), and residence abroad (n = 5). 175 were contacted, and 129 answered a semi-structured telephone interview regarding the participation, timing, and place of their child`s developmental FU, and 47 additionally participated in an online-survey on parental views on FU and early interventions.

Results: Of the 129 participating children (51% male) 58% had a cyanotic CHD. In the telephone interview, parents reported, that 89 (69%) children received at least one FU assessment at a mean age of 18 months, with 79% receiving their first assessment during within the first two years of life. Of those children who received a FU, 48% showed a developmental delay in at least one domain (cognitive, language, motor) and for 58% at least one type of early intervention was initiated. In the online survey, 81% of parents rated the FU assessment very positive. Also, early interventions were highly appreciated. From the 25 children without FU, 16 parents were not aware of the necessity of a FU, 11 of them agreed to come to a FU subsequently.

Conclusion: The FU-rate in our study population was lower than we would have expected. Adaptions of our FU program were introduced, accordingly. Furthermore, the nationwide register (implemented in 2019) additionally aims at ensuring good FU rates. The parental satisfaction with the FU's and early interventions was high. However, some parents were not aware of the potential risk of developmental delays and the possibility of specific FU's. Ways to address the parents are to repeatedly include this information in routine consultations and by contacting them by phone.

Keywords: Congenital heart disease, developmental delay, follow-up care

Basic science, Genetics

P-467

Industrial processing causes degeneration of xenografts

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Background and Aim: Besides homografts, different xenografts are frequently used for reconstructive surgery in patients with congenital heart disease. Their degeneration is well known and often induces re-operation. We intended to analyze changes caused by the production process prior to implantation in order to identify whether degeneration begins before implantation.

Methods: Native and processed tissues (Contegra® graft, bovine, equine and porcine pericardium) were analyzed with light and polarization microscopy after standard histological preparation and dying (HE, EvG).

Results: Less birefringence under polarized light indicated a degeneration of the Contegra® graft collagen fiber network as a result of manufacturing. Frequently seen interfibrillary gapping, tissue thickening and collagen network degeneration were among the patterns observed after industrial patch processing.

Conclusion: All analyzed specimen showed severe alterations initiated by the production process. Degeneration begins before implantation. Therefore, the choice of the most suitable material should be carefully evaluated. The ideal material remains to be found.

Keywords: congenital, material science, xenografts, histopathology

P-468

A case of infantile histiocytoid cardiomyopathy with fatal arrhythmia- a diagnostic approach from mitochondrial respiratory chain disorders

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Background and Aim: Histiocytoid Cardiomyopathy (HC) is a rare disease responsible for lethal ventricular arrhythmia in infancy. It maybe caused by genetic mutations involved in mitochondrial

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function, but difficult to diagnose in clinical practice. Here, we experienced an infantile case of HC and diagnosed as mitochondrial respiratory chain disorder by measuring oxidative phosphorylation (OXPHOS) function in skin-fibroblast and genetic study.
Methods: Case Report
She was born by normal vaginal delivery without asphyxia. Immediately after birth, AVRT sustained for several minutes was noted and delta waves were seen on later ECG, which led to the diagnosis of WPW syndrome. The echocardiogram showed left ventricular noncompaction with reduced systolic function. Left sensorineural hearing loss was also observed.
At the age of 15 months, she had a febrile episode, followed by loss of consciousness, which was diagnosed as VT/Vf by the emergency team. Defibrillation was ineffective, and the patient was transported to our hospital while resuscitation was continued. The arrhythmia was a short-coupled variant of torsades de pointes, which siesed with amiodarone and lidocaine after 75 minutes from initial treatment, and finally disappeared after oral administration of Mexiletine. Electrophysiological examination induced Purkinje-related VT in the left posterior bundle branch, and ablation was performed at this site. Subsequently, an ICD was implanted. Fortunately, there were no major neurological sequelae, and the patient is now 7 years old and has never had an episode of VT. She was clinically diagnosed possible histiocytoid cardiomyopathy and also mitochondrial disease was suspected based on the clinical presentation. Thus, genetic study for mitochondrial disease and maximum oxygen consumption rate (OCR) was measured using skin-fibroblast.
Results: The genetic study showed a novel mutation in NDUFB11gene, and maximum OCR in skin-fibroblast was significantly decreased, which confirmed mitochondrial respiratory chain disorder.
Conclusion: HC is probably one of the most lethal mitochondrial cardiomyopathies that can be prevented by early diagnosis. Genetic testing and skin biopsy for detecting OXPHOS dysfunction can be powerful diagnostic tools in infantile period.

Keywords: Histiocytoid cardiomyopathy, mitochondria, Purkinje-related ventricular arrhythmia

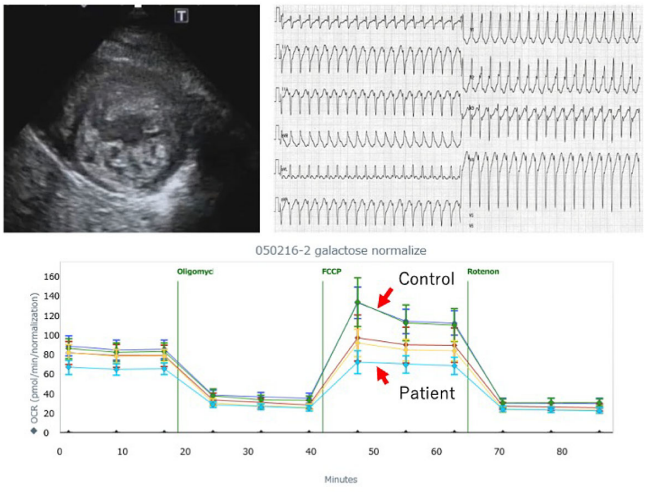

Figure 1. LV noncompation on echocardiogram (top left), sustained VT on electrocardiogram (top right), and oxygen consumption rate in skin-fibroblast (bottom)

P-469

Markers of collagen turnover, endothelial injury and activation in acute rheumatic fever and rheumatic heart disease: a case control study

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Background and Aim: Rheumatic heart disease (RHD) is a major contributor of cardiovascular morbidity and mortality. Diagnosis of acute rheumatic fever (ARF) is based on Jones criteria, which has a low sensitivity and specificity. Since, injury to valvular endothelium and underlying collagenous extracellular matrix is the predominant mechanism of ARF, we measured peripheral blood markers of endothelial activity, endothelial injury and collagen turnover in patients with ARF and compared it with stable RHD patients (without ARF) and healthy controls
AIM: To identify blood biomarkers to diagnose ARF.
Methods: We enrolled 41 patients with ARF, 20 patients with chronic RHD and 10 age matched healthy controls. From the peripheral blood, we measured levels of Hemangioblastic and Monocytic endothelial progenitor cells (EPC) using flowcytometry and fluorescence activated cell sorting. Microparticles released during apoptosis of these cells were characterized and enumerated using dynamic light spectroscopy and transmission electron microscopy. Additional markers such as VEGF, MMP, TIMP, by-products of collagen synthesis and degradation, adhesion molecules were estimated using commercially available ELISA kits.
Results: Patients with ARF had significantly elevated levels of all classes of metalloproteinases and VEGF. Collagen degradation product (CTXI) was higher in those with ARF whereas byproduct of collagen synthesis (PICP) was higher in those with RHD. Levels of VEGF was significantly higher in patients with ARF. Both early and late phases of hemangioblastic and monocytic EPCs were higher in the ARF group compared to those with RHD or controls. The number of microparticles and their proportion to the respective EPCs were higher in those with RHD. Circulatory vascular adhesion molecules were lower in the ARF or RHD group compared to controls.
Conclusion: The milieu is geared towards collagen degradation in ARF and towards collagen synthesis and deposition in RHD. The markers of endothelial injury and activation are significantly elevated in patients with ARF. Patients with RHD exhibit "EPC incompetence" wherein majority of EPCs undergo apoptosis (leading to increase in the number of microparticles corresponding to the specific EPCs) rather than contributing to endothelial repair. Future large studies may help establish any of the above as a potential biomarker to accurately identify underlying ARF in patients with RHD.

Keywords: Acute rheumatic fever, Rheumatic heart disease, endothelial activation, Endothelial progenitor cells, micro vesicles,translational research

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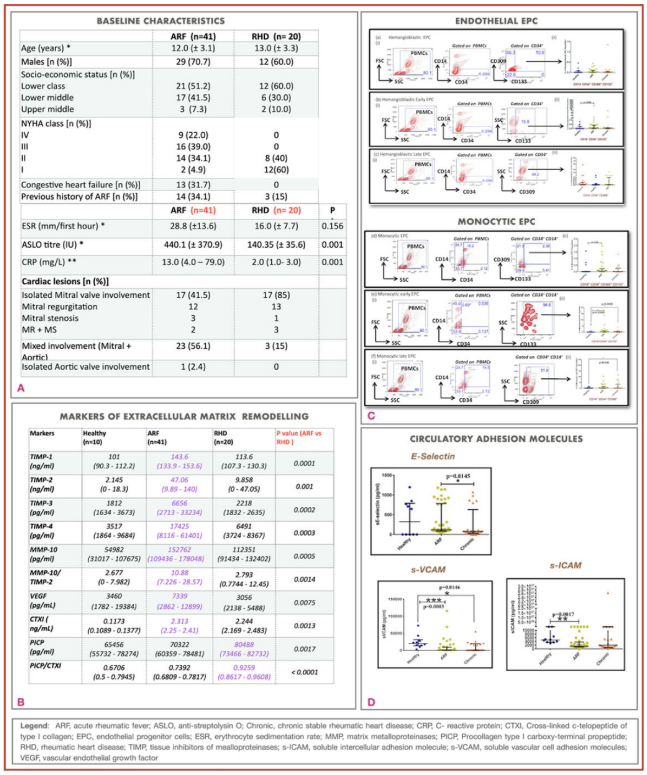
Figure showing the baseline characteristics of study participants and the key findings of the study



	ARF (n=41)	RHD (n= 20)	
Age (years) *	12.0 (± 3.1)	13.0 (± 3.3)	
Males [n (%)]	29 (70.7)	12 (60.0)	
Socio-economic status [n (%)]			
Lower class	21 (51.2)	12 (60.0)	
Lower middle	17 (41.5)	6 (30.0)	
Upper middle	3 (7.3)	2 (10.0)	
NYHA class [n (%)]			
IV	9 (22.0)	0	
III	16 (39.0)	0	
II	14 (34.1)	8 (40)	
I	2 (4.9)	12(60)	
Congestive heart failure [n (%)]	13 (31.7)	0	
Previous history of ARF [n (%)]	14 (34.1)	3 (15)	
	ARF (n=41)	RHD (n= 20)	P
ESR (mm/first hour) *	28.8 (±13.6)	16.0 (± 7.7)	0.156
ASLO titre (IU) *	440.1 (± 370.9)	140.35 (± 35.6)	0.001
CRP (mg/L) **	13.0 (4.0 – 79.0)	2.0 (1.0- 3.0)	0.001
Cardiac lesions [n (%)]			
Isolated Mitral valve involvement	17 (41.5)	17 (85)	
Mitral regurgitation	12	13	
Mitral stenosis	3	1	
MR + MS	2	3	
Mixed involvement (Mitral + Aortic)	23 (56.1)	3 (15)	
Isolated Aortic valve involvement	1 (2.4)	0	

A

Markers	Healthy (n=10)	ARF (n=41)	RHD (n=20)	P value (ARF vs RHD)
TIMP-1 (ng/ml)	101 (90.3 - 112.2)	143.6 (133.9 - 153.6)	113.6 (107.3 - 130.3)	0.0001
TIMP-2 (ng/ml)	2.145 (0 - 18.3)	47.06 (9.89 - 140)	9.858 (0 - 47.05)	0.001
TIMP-3 (pg/ml)	1812 (1634 - 3673)	6656 (2713 - 33234)	2218 (1832 - 2635)	0.0002
TIMP-4 (pg/ml)	3517 (1864 - 9684)	17425 (8116 - 61401)	6491 (3724 - 8367)	0.0003
MMP-10 (pg/ml)	54982 (31017 - 107675)	152762 (109436 - 178048)	112351 (91434 - 132402)	0.0005
MMP-10/ TIMP-2	2.677 (0 - 7.982)	10.88 (7.226 - 28.57)	2.793 (0.7744 - 12.45)	0.0014
VEGF (pg/mL)	3460 (1782 - 19384)	7339 (2862 - 12899)	3056 (2138 - 5488)	0.0075
CTXI (ng/mL)	0.1173 (0.1089 - 0.1377)	2.313 (2.25 - 2.41)	2.244 (2.169 - 2.483)	0.0013
PICP (pg/ml)	65456 (55732 - 78274)	70322 (60359 - 78481)	80488 (73466 - 82732)	0.0017
PICP/CTXI	0.6706 (0.5 - 0.7945)	0.7392 (0.6809 - 0.7817)	0.9259 (0.8617 - 0.9608)	< 0.0001

B

C

D

Legend: ARF, acute rheumatic fever; ASLO, anti-streptolysin O; Chronic, chronic stable rheumatic heart disease; CRP, C- reactive protein; CTXI, Cross-linked c-telopeptide of type I collagen; EPC, endothelial progenitor cells; ESR, erythrocyte sedimentation rate; MMP, matrix metalloproteinases; PICP, Procollagen type I carboxy-terminal propeptide; RHD, rheumatic heart disease; TIMP, tissue inhibitors of mealloproteinases; s-ICAM, soluble intercellular adhesion molecule; s-VCAM, soluble vascular cell adhesion molecules; VEGF, vascular endothelial growth factor

Panel A: Table showing the baseline characteristics of study population. Panel B: Table showing the levels of various markers of extracellular matrix remodelling including the vascular endothelial growth factor. Panel C: Flowcytometry and fluorescence activated cell sorting results of enumeration of hemangioblastic and monocytes endothelial progenitor cells (including the early and late phases) in each group. The box and whicker plot is given as the last column. Panel D: Box and whisker plots showing the levels of circulatory adhesion molecules in patients with ARF, RHD and healthy controls.

P-470

Genome-wide association study in finngen identifies novel congenital heart disease risk LOCI

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Background and Aim: FinnGen is a large-scale biobank study that aims to genotype 500,000 Finnish participants recruited from hospital samples and prospective and retrospective epidemiological and disease-based cohorts. These data are combined with longitudinal registries that record phenotypes and health events over the entire lifespan. Our aim was to identify genetic risk factors associated with congenital heart defects (CHD) in FinnGen.
Methods: We performed a genome-wide association study (GWAS) of general CHD (N = 2436) and five CHD subgroups in FinnGen Data Freeze 7 (N = 309,154) followed by eQTL analysis in the GTEx database and gene expression analysis in human induced pluripotent stem cell derived cardiomyocytes (hiPS-CM) from CHD patients (N = 4) and healthy controls (N = 4).
Results: From the general CHD category GWAS we detected 1 significant lead SNP in chromosome 17 (rs11570508, beta = 0.24, P = 1.2x10-11, intronic region of CDC27). An additional four SNPs were significantly associated with CHD subgroups. Two of these, rs1342740627 associated with left ventricular outflow tract obstruction (LVOTO) defects and rs1293973611 associated with septal defects, were Finnish enriched. Three risk loci for three CHD subtypes had a high beta value. The eQTL calculation resulted in the potential effect by rs11570508 on two nearby genes: MYL4 (eQTL P = 0.0017 in heart - atrial appendage) and KPNB1 (eQTL P = 0.04 in heart - left ventricle). Lower expression levels of these two genes were observed in human induced pluripotent stem cell derived cardiomyocytes (hiPSC-CM) from LVOTO patients compared with healthy controls.
Conclusion: Our study identified a robust risk locus for CHD near candidate genes with a known role in heart development, and MYL4 and KPNB1 were demonstrated as a potentially causal genes for CHD. In addition, three risk loci for three CHD subtypes had a high beta value, suggesting that very rare variants can play a substantial role in CHD. Finally, two lead SNPs were Finnish enriched, highlighting the value of FinnGen study in identifying novel rare or low-frequency disease variants in an isolated population with enrichment of variants due to recent population bottlenecks followed by rapid population growth.

Keywords: Congenital heart defects, Genome-wide association study, Human induced pluripotent stem cells, Genetics

P-471

Indicators of the serotonin system as early biomarkers of PAH development in children with CHD

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Background and Aim: Pulmonary arterial hypertension (PAH) is a serious complication of congenital heart defects (CHD). Pathological changes in PAH are characterized by irreversible changes in all layers of the vascular wall of the distal pulmonary arteries.
AIM: pathogenetic substantiation of the involvement of serotonin in the development of PAH in children with CHD.
Methods: The study included 55 children aged 1 month to 2 years with congenital heart defects with left-right shunting (VSD, PDA, ASD). The examined children were divided into two groups: I - CHD+PAH, II - CHD. In addition to the standard clinical study, serotonin levels in blood serum, platelets and serotonin membrane transporter (SERT) were determined in all children using

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Serotonin ELISA kits, IBL Hamburg. Activity of the phosphorylated SERT forms was determined by Western blotting.
Results: In group I, a high degree of PAH was found in 46% of children, a moderate degree in 12.5%, and a mild degree in other cases. Pulmonary arterial pressure ranged from 30 to 108 mm Hg. In children, tricuspid regurgitation of varying severity was revealed, more often moderate, right ventricular dilatation in 58.3% and right atrial dilatation in 41.7%.
Total amount of membrane serotonin transporter in platelets in I group was significantly higher and amounted to 2122 pg/10^9 compared with group II 409 pg/10^9 ($p<0.001$). Serotonin level in blood serum was 30% higher in group I compared with group II against the background of lower values of serotonin in platelets. At the same time, a decrease in the phosphorylated form of SERT was noted, that is, more than half of it did not have functional activity. Probably, this may indicate the manifestation of a compensatory SERT response to an increase of serotonin in blood serum.
Conclusion: The serotonin system is involved in the development of PAH in children with CHD. Determination of SERT, serotonin concentration in blood serum and platelets can serve as early markers of PAH development in young children with CHD.
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Keywords: children, PAH, CHD, serotonin, SERT

P-472 / Moderated Poster
Homozygous genetic variant in desmin causes severe inflammatory cardiomyopathy
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Background and Aim: Myocarditis is an inflammatory entity of the myocardium that can lead to severe heart failure. Recently, we showed that pediatric myocarditis with a dilated cardiomyopathy (DCM) phenotype is caused by pathogenic genetic variants in cardiomyopathy genes. The role of inflammation in these patients is not well understood.
Methods: We analyzed a 7-years-old girl with chronic myocarditis and phenotype of DCM, her parents, and two younger siblings by whole-exome sequencing (WES). The WES data were filtered for rare (minor allele frequency <10-4) pathogenic genetic defects in cardiomyopathy (n = 89) and immune disorder genes (n = 631). Immune disease genes covered entities such as autoinflammatory diseases, complement disorders, and severe immunodeficiencies. Endomyocardial biopsy (EMB) samples were analyzed histologically, by immunostaining, and viral DNA/RNA RT-PCR.
Results: The girl was first seen with symptoms of severe heart failure with a left ventricular end-diastolic diameter of 51 mm and a left ventricular ejection fraction (LVEF) of 33%. NT-proBNP and Troponin Ihs levels were elevated with 4205 ng/l and 367 pg/nl, respectively. CMR detected no edema but late gadolinium enhancement. The first EMB showed chronic myocarditis without myocardial viral detection. Optimal heart failure therapy did not improve LVEF. Four month later, the second EMB showed progressive inflammation, leading to immunosuppressive therapy. However, the patient needed a left ventricular assist device (LVAD) and heart transplantation. The third EMB during (LVAD) implantation showed again progressive inflammation without viral detection. Genetic analysis with WES identified a homozygous missense variant in the known cardiomyopathy gene Desmin (*DES* c.1244G>A, p.R415Q) implicating a primary cardiomyopathy. Of note, the homozygous *DES* c.1244G>A missense variant affects the donor splice site of intron_6-7. Immunodetection of the Desmin mutant protein showed reduced staining intensity in all EMB. Further, screening of immune disorder genes did not identify pathogenic rare, homozygous, truncating, or *de novo* variants in this patient.
Conclusion: This case report supports the theory of underlying defects in cardiomyopathy genes in myocarditis with DCM phenotype. A pathogenic variant in immune genes was not detected pointing towards different mechanisms leading to inflammation and inflammatory cardiomyopathy in these patients.
Keywords: myocarditis, inflammatory cardiomyopathy, genetic variants, dilated cardiomyopathy, pediatric

P-473
Homozygous 3 BP-deletion in bola3 causes a severe cardiac phenotype in early childhood with lethal outcome
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Background and Aim: Due to deficient lipoate synthesis, variants within the BOLA3 gene result in a deficiency of 2-oxoacid dehydrogenases accompanied by defects of the mitochondrial respiratory chain. Patients typically show a severe and progressive multisystem phenotype with hypertrophic cardiomyopathy, white matter lesions, epilepsy, and involvement of visceral organs. Around 25 patients can be found in scientific literature. We describe the first patient with a severe phenotype and homozygosity for a 3 bp-deletion in BOLA3.
Methods: We report on 2 siblings who showed a severe cardiac phenotype at the age of 5 and 8.5 months, respectively, both with a lethal outcome. We describe the clinical findings of both patients over time together with laboratory and genetic results.
Results: Patient 1 presented at the age of 5 months after a 7-dose vaccination in reduced general condition, circulatory failure necessitating resuscitation, severe lactic acidosis of 22 mmol/l, and hyperammonemia. He died on the day of admission. Endomyomyocardial biopsy (EMB) revealed acute lymphocytic parvovirus B19 associated myocarditis with 80,000 copies PVB19 DNA/µg cardiac DNA. Patient 2 presented at the age of 8.5 months with hypertrophic cardiomyopathy, severely impaired pump function, rapidly developing myocardial failure, lactic acidosis of 10 mmol/l, and hyperglycinemia of 574.7 µmol/l. Sudden bradycardia requiring resuscitation was followed

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by implantation of veno-arterial extracorporeal membrane oxygenation. As EMB was suspicious for non-compaction cardiomyopathy with unlikely recovery, a biventricular assist device was implanted to avoid failure of the right ventricle and to prepare for a heart transplant (Htx). In addition, a focal lymphocytic myocarditis was observed with CD3+ T cells (30/mm2). At that point in time, genetic testing identified homozygosity for a 3-bp deletion (c.220_222del; p.(Glu74del)) within BOLA3. This diagnosis and associated prognosis militated against further pursuit of the plan for Htx, and the patient died after termination of life support.
Conclusion: We report a patient with homozygosity for a 3-bp deletion and sudden early lethal outcome. His genetic variant has once been described in heterozygous expression, then associated with a milder phenotype. Both siblings show the typical manifestation of an iron-sulfur (Fe-S) cluster disorder, affecting many mitochondrial functions.

Keywords: BOLA3, homozygosity, lethal outcome, cardiomyopathy

P-474 / Moderated Poster

Concerns about safety of growth hormone therapy in noonan syndrome and other rasopathies: risk of triggering or worsening hypertrophic cardiomyopathy

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Background and Aim: A new indication for growth hormone (GH) therapy has recently been accepted by the European Medicines Agency, that of GH-therapy for short stature in Noonan syndrome (NS) even in the absence of reduced GH levels, and with hypertrophic cardiomyopathy (HCM) not considered a contra-indication. This is concerning as virtually no safety data have been published, because HCM was considered a contra-indication for inclusion for all early trials for GH-therapy in NS-patients. It is known that excess GH from pituitary adenomas can cause HCM, and that GH is a trophic factor for myocytes in cell culture. We aimed to assess the effect of GH-therapy on the myocardium of patients with NS, and define useful parameters for clinical follow-up
Methods: The NS-patients treated with GH (GH-group, n = 4; 1 with known HCM at start of therapy, 3 without established HCM-diagnosis at start of therapy), were compared with three NS case-controls each with already diagnosed HCM (NSHCM-group), matched for sex and clinical characteristics such as valve stenosis (n = 12). The cardiac ultrasound- and ECG-measurements at start and cessation of GH-therapy in the GH-group, were compared with measurements at identical age in the NSHCM-group.
Results: Age at start of GH-therapy was median 6.0y (range 4.4-9.8), and duration of GH-therapy median 4.5 (2.1-7.0)y. Maximal wall thickness Detroit Z-score increased by 238%, from 1.97 [IQR 1.80-3.12] to 4.69 [3.88-5.83; p = 0.02]. Over the corresponding time-period the NSHCM-group did not change significantly, from 3.10 [2.13-4.27] to 3.17 [2.17-3.64]. Septum-to cavity ratio, left ventricular wall-to-cavity ratio and ECG limb-lead voltage x QRS-duration product were all useful for monitoring and showed significantly greater progression of hypertrophy over time in GH-group with annual changes respectively 0.055 [0.027-0.082] versus -0.003 [-0.009 to 0.002; p = 0.004], 0.050 [0.027-0.081] versus -0.004 [-0.005 to 0.002; p = 0.006] and 0.088 [0.050-0.141] versus -0.001 [-0.010 to 0.011; p = 0.004]. All four in GH-group had GH-therapy terminated because of cardiac deterioration, and one died suddenly 3.5y after discontinuation of GH-therapy.
Conclusion: There is a high risk of worsening pre-existing HCM or triggering HCM with GH-therapy in NS-patients, as we find significantly faster progression of cardiac hypertrophy compared with NS-associated HCM-patients not given GH-therapy.

Keywords: Growth hormone therapy, Noonan syndrome, hypertrophic cardiomyopathy, RASopathy, somatostatin

P-475

Precise in vivo functional analysis of dna variants associated with congenital heart disease

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Background and Aim: Single nucleotide variants (SNVs) are strongly implicated in congenital heart disease (CHD). The recent development and optimizations of base editors now promise to enable direct functional assessment of SNVs in model organisms. We aimed to functionally assess SNVs previously detected in CHD patients using in vivo base editing in fish at sites orthologous to SNV positions in human genes of interest.
Methods: We used state-of-the-art adenine (ABE) and cytosine base editors (CBE) in the medaka (*Oryzias latipes*) fish model to modify specific target DNA bases in medaka embryos and assessed the phenotypic consequences by light microscopy. Design of base editing sgRNAs and off-target evaluation was performed with a newly developed online base editing tool ACEofBASEs.
Results: We demonstrate high efficiency and precision of in vivo base editing by editing eye pigmentation and transgenic GFP genes. We benchmark base editing by altering the coding sequence of genes encoding the cardiac troponin T and the potassium channel ERG, which allowed us to recreate known heart phenotypes from stable mutant lines or CRISPR-Cas9 gene knockouts used as reference. We then validate missense mutations in novel CHD genes *dapk3*, *ube2b*, *usp44*, *ptpn11*, and *myrf* by editing medaka gene loci comparable to the mutations detected in humans and present the gene-specific heart phenotypes.

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Conclusion: This DNA base editing framework in fish provides a straightforward validation of SNVs associated with human CHD. The rapid in vivo assessment facilitates prioritizing genetic variants for detailed mechanistic downstream studies.

Keywords: SNVs, CHD, base editing, CRISPR-Cas9, fish

Other

P-476 / Moderated Poster
Clinical symptoms in children with genetic aortopathy and their response to medical therapy – a monocentric analysis over more than one decade

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Background and Aim: Genetic aortopathy (GA) is a rare form of cardiovascular disease in childhood. Over the last years continuously more genetic mutations presenting as GA have been found. The rarity of each disease makes it difficult to systematically describe the characteristics of these diseases. Here, we describe clinical data from children with all forms of GA collected over the past 13 years in our database and their response to medical therapy.
Methods: Since 2008 we investigated 681 patients in 1755 visits with suspected GA of which 243 patients were diagnosed clinically or genetically. We retrospectively analyzed the type of mutation, prevalence and age of onset of symptoms from birth to adulthood. The progression of Dilatation of the Sinus of Valsalva (SV) (Delta z-Score) was compared between patients on Betablocker vs. Sartans.
Results: 62% of children with diagnosed GA had an FBN1 Mutation. Each of the other mutations (FBN2, TGFB1/2, TGFBR1/2, BGN) occurred in between 1 and 5 %. In 18 % no genetic variant was found. All clinical symptoms present age-dependently. For example, SV dilatation and dural ectasia are present in 50% of cases at the age of 15,9 and 16,8 years, whereas pneumothorax does not reach this mark. At the time of transition to adulthood 15,6, 11,1 and 2,5 % of patients show these symptoms, respectively. Progression of dilatation of the SV was significantly lower in children treated with sartans compared to beta-blockers (Delta z-score $p<0{,}001$).
Conclusion: In our large, monocentric pediatric patient group with GA, Marfan syndrome (FBN1) remains the most frequent form of GA. Each of the other types of GA present rarely. Each of the typical symptoms of GA present in an age-dependent manner, that needs to be known to correctly manage patients. Some typical symptoms of adult GA patients hardly exist in childhood. In our cohort sartans are more effective in reducing the progression of SV Dilatation, making it our therapy of choice in children with GA.

Keywords: Marfan Syndrom, Genetic aortopathy, children, Sartan, Beta-Blocker,

Basic science, Genetics

P-477
Genetic predictors of the clinical presentation of non-syndromic aortic coarctation

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Background and Aim: Coarctation of the aorta (CoA) is a congenital narrowing of the aorta accounting for 4-8% of all congenital heart defects. Aortic coarctation displays a distinct phenotypic variability, and it may present either as an isolated lesion or in combination with other specific cardiac malformations. This study aims to identify genetic factors behind the susceptibility to non-syndromic CoA in the Swedish population, and how these factors might influence the specific clinical presentation of CoA in affected individuals.
Methods: DNA samples from Swedish patients with surgically treated CoA were obtained through the Swedish National Biobank for Congenital Heart Disease. The samples ($n = 196$) were genotyped using the Infinium OmniExpressExome chip from Illumina and compared to Swedish population controls ($n = 5670$) and a separate cohort of patients with bicuspid aortic valve (BAV) without CoA, under-going elective open-heart surgery for aortic valve or ascending aortic disease at the Karolinska University Hospital, Sweden ($n = 633$). In addition to the full cohort, we tested four partially overlapping sub-phenotypes in a case-control setting: CoA-BAV ($n = 84$), CoA with ventricular septal defect ($n = 47$), CoA in combination with other left-sided obstructive lesions (LsL, Left ventricular outflow tract obstruction, aortic/mitral stenosis or hypoplastic aortic arch, $n = 29$) and early-onset CoA (requiring surgical repair before three weeks of age, $n = 107$).
Results: We found a genome-wide significant association between a signal on chromosome 2 (rs167378) and the CoA-LsL phenotype. Additionally, variant rs7541800 located on chromosome 1 was significantly associated with the CoA-BAV phenotype. This variant showed no association with BAV without CoA. Finally,

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variant rs1325713 located on chromosome 6 was strongly associated with early-onset CoA. Nearly two-thirds of female patients with CoA (50/67) were operated within the first three weeks of age, compared to 44% of male patients (p = .000049). Variant rs6738178 on chromosome 2 showed genome-wide significant interaction between genotype and sex in the early-onset CoA subgroup.

Conclusion: This study reveals genetic susceptibility loci that may be involved in developing the distinct morphological and clinical subphenotypes of CoA in Sweden. These results elucidate the role of genetic variants in aortic coarctation and provide a novel approach for detecting previously unknown genetic associations.

Keywords: Aortic Coarctation, Genetics, GWAS, BAV, Left-sided lesions

P-478

Cholestasis is associated with impaired hepatic synthetic function in children, adolescents and adults with fontan circulation

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Background and Aim: Fontan associated liver disease (FALD) is a hepatic disorder arising from hemodynamic changes due to the Fontan physiology. FALD-induced hepatic fibrosis is rarely marked by biochemical alterations, however, a chronic increase in serum bile acid (BA) levels has been observed in some patients indicating cholestasis. We compared clinical-, hemodynamic- and biochemical variables in Fontan patients with and without cholestasis.

Methods: We investigated common biochemical markers and serum human BA, measured by mass spectrometry, in 45 Fontan patients (<18 years n = 18). Using a cutoff value of ≥10 µg/ml BA in serum, two groups of Fontan patients could be distinguished: Cholestatic n = 6, non-cholestatic n = 39. Anticoagulation was taken by 38/45 patients: thrombo ASS n = 18 patients [cholestatic n = 0, non-cholestatic n = 18] and Phenprocoumon [PhC] n = 20 patients [cholestatic n = 4, non-cholestatic n = 16]) with comparable dosages within the respective study groups.

Results: Cholestatic Fontan patients showed significantly increased total bilirubin- (median: 1,4mg/dl; interquartile range [IQR]: 0,6-3,6mg/d) and NTproBNP levels (median: 111 pg/ml, IQR: 170,1-523,5 pg/ml) compared to non-cholestatic patients (bilirubin median: 0,8mg/dl, IQR]: 0,6-1,0mg/dl, p = 0,007; NTproBNP median: 119 pg/ml, IQR: 79,6-230,9 pg/ml p = 0,032). Fontan patients with cholestasis had a significantly decreased platelet count (median: 133.000/µl, IQR: 159.450-280.330/µl) and antithrombin III (ATIII) levels (median: 82%, IQR: 66,3-102,9%) compared to non-cholestatic patients (platelet median: 196.000/µl, IQR: 46.000-.324.900/µl, p = 0,023; ATIII median: 101% IQR: 95,9-104,6%, p = 0,002). In all Fontan patients on PhC therapy, INR (international normalized ratio) and aPTT (partial thromboplastin time) values were significantly increased in cholestatic Fontan patients (INR: median 2,9, IQR: 2,3-3,3; aPTT median: 54 sec, IQR: 38,8-70,1 sec) compared to non-cholestatic patients on PhC (INR: median 2,1, IQR: 1,6-2,3, p = 0,011; aPTT median: 46 sec, IQR: 36,9-63,1 sec, p = 0,036).

Conclusion: Cholestasis in Fontan patients is associated with lower platelet count and ATIII levels as well as with enhanced INR and aPTT values. Our findings argue for an impaired hepatic synthetic function in cholestatic patients with Fontan circulation. However, those findings need further investigation by a larger study cohort.

Keywords: Fontan circulation, cholestasis, bile acids, congenital heart disease

P-479 / Moderated Poster

Arrhythmia and impaired myocardial function in heritable thoracic aortic disease: an international retrospective cohort study

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Background and Aim: Heritable thoracic aortic diseases (HTAD), typically entailing aortic complications, can be caused by pathogenic or likely pathogenic variants (PV/LPV) in several genes encoding proteins of the extracellular matrix, smooth muscle cells or components of the transforming growth factor (TGF)-β signaling pathway. Myocardial features such as impaired myocardial function and/or arrhythmia have increasingly been reported, mainly in Marfan syndrome and are acknowledged as additional causes of morbidity and mortality. The prevalence of these manifestations and clinical characteristics of HTAD patients presenting these features is largely unknown.

Methods: This international multicentre retrospective study collected data on patients with HTAD presenting impaired myocardial function and/or arrhythmia. Patients 12 years or older carrying a PV/LPV variant in FBN1, TGFBR1, TGFBR2, TGFB2, TGFB3, SMAD3 and ACTA2 were screened. Impaired myocardial function included (a)symptomatic reduced ejection fraction (EF<50% - HFrEF) or symptomatic heart failure with preserved ejection fraction (HFpEF). Arrhythmias included atrial fibrillation (AF), atrial flutter (AFL), ventricular tachycardia (VT), ventricular fibrillation (VF) and (aborted) sudden cardiac death (presumed arrhythmogenic) (SCD).

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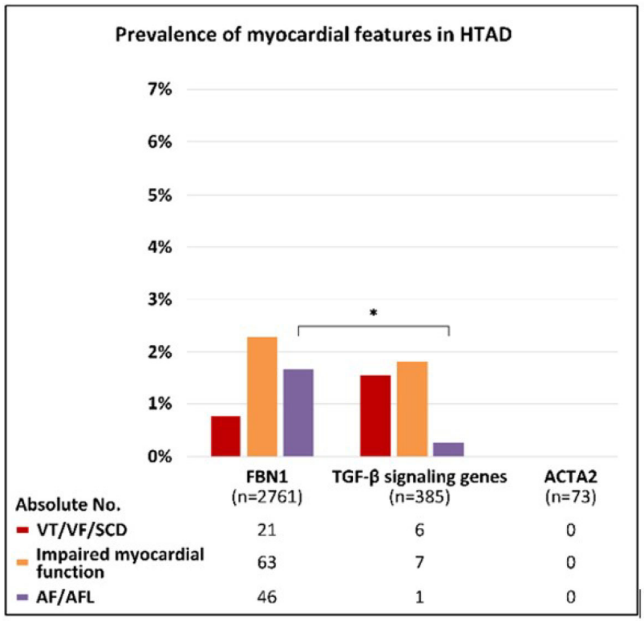
Results: 3219 files of patients with HTAD were screened (2761, 385 and 73 in FBN1, in a TGF-β signaling gene and in ACTA2 respectively). Myocardial features were reported in 102 patients (3.2%) (age 35 [age range 12-77] years, 39% female): 89 carrying a FBN1 variant and 13 carrying a variant in one of the TGF-β signaling genes. None of the features were reported in screened patients with variants in ACTA2. Among the 144 reported myocardial features, 27 (27%) were VT/VF/SCD, 70 (69%) were impaired myocardial function and 47 (46%) were AF/AFL. Among the patients with myocardial features, severe valvular disease was noted in 58% and prior cardiac surgery in 79%, while 19% of the patients developed myocardial features in the absence of any of the latter.
Conclusion: In patients with HTAD, arrhythmia and impaired myocardial function was reported in patients with variants in FBN1 and in the TGF-β signaling genes and not in patients harboring ACTA2 variants. Though infrequent, myocardial features should be acknowledged as potentially severe, also occurring in young patients with no underlying significant valvular or aortic disease.

Keywords: HTAD, myocardial dysfunction, arrhythmia, Marfan, TFG-b, ACTA2

Figure 1: Prevalence of myocardial features in HTAD



P-480

Mitral annular disjunction and cardiovascular outcome in marfan syndrome

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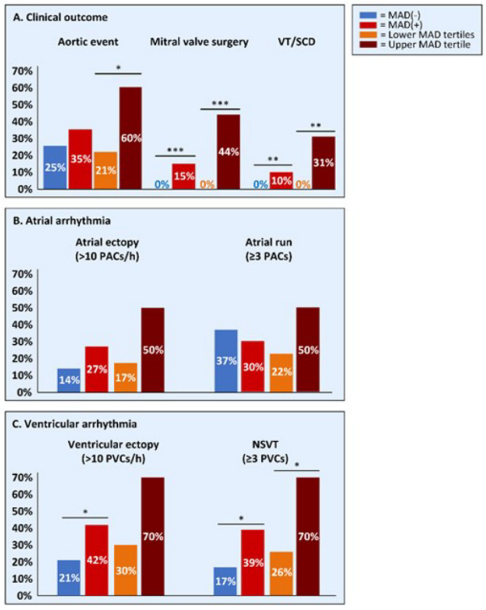
Background and Aim: Mitral annular disjunction (MAD) has received growing interest in patients with mitral valve prolapse (MVP), ventricular tachycardia (VT) and sudden cardiac death (SCD). The clinical significance of MAD in Marfan syndrome (MFS) remains largely unexplored. The aim of this study is to define the prevalence of MAD and examine the relation with cardiovascular outcomes and arrhythmia in patients with MFS.
Methods: This is a retrospective single-center study including patients with MFS who underwent regular follow-up between January 2004 and December 2019. Adults and children with a diagnosis of MFS based on the revised Ghent criteria and a confirmed (likely) pathogenic variant in the FBN1 gene were included.
Results: The presence of MAD was assessed by echocardiography and the extent of MAD was categorized in tertiles. Patients also underwent resting ECG and 24-hour Holter monitoring. Outcomes included aortic events (aortic dissection or prophylactic aortic surgery), arrhythmic events (defined as sustained VT/SCD) and mitral valve surgery. A total of 142 patients (median age 25 years [range 2-64], 51% women) were evaluated. Forty-eight (34%) patients had MAD. Patients with MAD had larger aortic root Z-scores (4.1 [2.8-5.7] vs. 3.0 [1.8-4.0], $p<0.001$) and more often showed MVP (71% vs. 15%, $p<0.001$), ventricular ectopy (42% vs. 21%, $p = 0.027$) and non-sustained ventricular tachycardia (39% vs. 17%, $p = 0.014$) than patients without MAD. During follow-up, aortic events occurred at similar rates in patients with vs. without MAD (35% vs. 25%, $p = 0.242$), but patients in the upper MAD tertile (>10mm) showed a higher occurrence of aortic events (60% vs. 21%, $p = 0.011$) compared to patients with MAD(+)≤10mm. Patients with arrhythmic events ($n = 5$) and patients requiring mitral valve surgery ($n = 7$) were observed exclusively in the group displaying MAD.
Conclusion: MAD in patients with MFS is associated with the occurrence of arrhythmic events, a higher need for mitral valve intervention, and, in patients with extensive MAD, more aortic events. Cardiac imaging in MFS should consider the assessment of MAD as a potential marker for adverse outcome.

Keywords: Marfan syndrome, mitral annular disjunction, arrythmia, myocardial disease

Cardiovascular outcome in Marfan syndrome in relation to mitral annular disjunction



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P-482

Decreased cardiac SDF-1/CXCR7 expression contributes to neonatal hyperoxia induced exercise intolerance in adult rats

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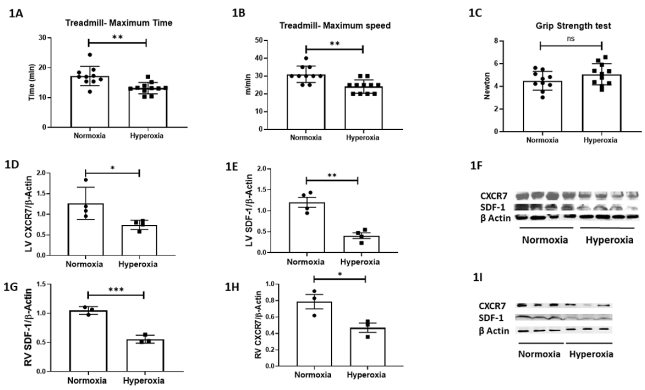
Background and Aim: Neonatal oxygen (O2) exposure leads to reduced exercise capacity and increased systemic vascular stiffness in preterm born adults. Stromal derived factor (SDF-1) and its receptor Chemokine Receptor-7 (CXCR7) play a crucial role in cardiovascular homeostasis and repair. Here we test the hypothesis that neonatal oxygen exposure reduces exercise capacity in adulthood and this is associated with decreased SDF-1 and CXCR7 levels.
AIM: To determine whether neonatal O2 alters aerobic exercise capacity, vascular stiffness and cardiac SDF-1 levels and systemic vascular function in rats exposed to neonatal hyperoxia.
Methods: Newborn rats (n = 24) were randomly assigned to normobaric normoxia (RA) or hyperoxia (85% O2) from postnatal day (P) 1 to P14. Rats were maintained in normoxic conditions until 12 weeks at which time treadmill running exercise, grip muscle strength test, carotid pulse wave velocity (PWV) doppler, left and right ventricular SDF-1 and CXCR7 protein expression were determined. Data were analyzed by student T test.
Results: Exercise stress test revealed that young adults rats exposed to neonatal O2 had significantly reduced exercise capacity and similar limb strength as compared to controls (Fig 1A-C). This was associated with increased systemic vascular stiffness as evidenced by increased PWV in carotid arteries (Fig 2A). In addition, Western blot analyses demonstrated a significant decrease in SDF-1 and CXCR7 expression in both left (Fig 3A-C) and right (Fig 4A-C) ventricles.
Conclusion: Neonatal O2 exposure decreases exercise capacity and increases systemic vascular stiffness in adult rats. This was accompanied by decreased cardiac SDF-1 and CXCR7 expression. Given the reduction in SDF-1/CXCR7 expression, further mechanistic studies elucidating the mechanistic role of SDF-1/CXCR7 signaling in cardiac dysfunction in preterm born adults will be necessary.

Keywords: Cardiac SDF-1/CXCR7, young adults born preterm

Figures for abstract



P-483

Flow stagnation induces endothelial-to-mesenchymal transition in the pediatric endocardium which can be inhibited by statins

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Background and Aim: Congenital heart defects are often associated with fibrotic processes of the endocardium, which have been clinically linked to flow stagnation (e.g., aortic and mitral stenosis). Our group has previously reported that the underlying mechanism of inducing this endocardial fibroelastic process is TGF-β mediated endothelial-to-mesenchymal transition (EndMT). In other disease processes, e.g., atherosclerosis, EndMT can be successfully inhibited with statins through a shift in TGF-β signaling via the transcription factor KLF2, but little is known about similar effects on the endocardium. In the present study, we examine the effect of KLF2 induction through atorvastatin on regulation of EndMT in pediatric endocardial cells.
Methods: To assess the response of endocardial cells to flow stagnation, we performed our experiments on healthy lamb endocardial endothelial cells isolated from left ventricular tissue. These cells were exposed to either flow stagnation (culture in static medium) or flow stagnation + TGF-β-stimulation (10ng/m), with and without KLF2-stimulation through atorvastatin (0,5μmol/l), respectively, for 24h. We analyzed the levels of EndMT (double staining with CD31 and α-SMA), endocardial cell viability (acetylated LDL-uptake), mesenchymal cell function through collagen expression (COL1A1), and KLF2 expression with immunofluorescence staining and confocal microscopy.
Results: Flow stagnation with and without TGF-β-stimulation resulted in increased EndMT (p<0.001, figure 1A,B,D). In both cases endocardial endothelial cells undergoing EndMT maintained their endocardial phenotype (positive acetylated LDL-uptake) while already producing significant amounts of collagen. Under KLF2-stimulation with atorvastatin, we observed a significant reduction in EndMT (under TGF-β-stimulation reduction by 73.2%, 95% CI: 71.0-75.1, p<0,001, n = 3, under no TGF-β-stimulation reduction by 27.8%, 95% CI: 10.7-44.8, p = 0,001, n = 3, figure 1A,B,D) and collagen expression (under TGF-β-stimulation reduction by 23.6%, 95% CI: 10.1-37.2, p<0,001, n = 3, under no TGF-β-stimulation reduction by 23.4%, 95% CI: 12.9-33.3, p<0,001, n = 3, figure 1A+C) and invertedly related to it a measured increase in KLF2 (1.4-fold increase, 95% CI: 1.2-1.6, p<0.001, n = 3, figure 1A+E).
Conclusion: In healthy pediatric endocardial endothelial cells, flow stagnation as well as TGF-β-stimulation result in EndMT. The stimulation of KLF2 through atorvastatin significantly inhibits EndMT. These findings might translate into atorvastatin as possible inhibitor of EndMT in pediatric patients with flow stagnation and fibrotic processes on the endocardial level.

Keywords: Endocardial Fibroelastosis, Endothelial-to-Mesenchymal Transition, Flow Stagnation, KLF2, Atorvastatin

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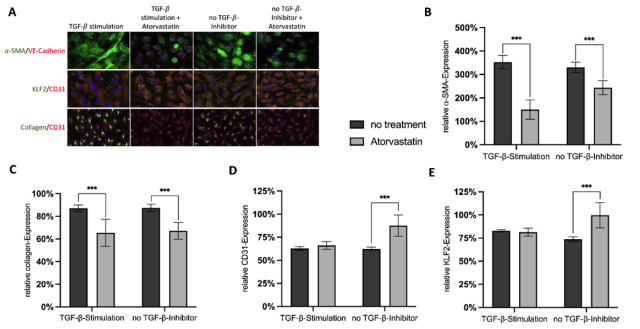

Fig. 1 Atorvastatin inhibits EndMT in pediatric endocardial cells. **A** Healthy lamb endocardial endothelial cells exposed to 24h flow stagnation were stained for endothelial and mesenchymal markers, COL1A1, and KLF2 and analyzed using confocal microscopy. **B-E** Relative expression of α-SMA (**B**), collagen (COL1A1, **C**), CD31 (**D**), and KLF2 (**E**) after 24h flow stagnation ± TGF-β-stimulation ± atorvastatin in relation to expression in standard culture under TGF-β-inhibition (defined as 100%). Error bars represent 95% confidence interval . *** p≤ 0.001.

P-485

Transforming growth factor β level in healthy children and pediatric marfan patients: the tiger for kids study

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Background and Aim: TGFβ plays a crucial role in the pathogenesis of Marfan syndrome. Elevated TGFβ levels correlate with increasing aortic root dilatation and can be reduced by TGFβ receptor blockade. Neither normal values nor TGFβ levels in pediatric Marfan patients are known. We performed our TiGer For Kids study to investigate the role of TGFβ and hereby present our results concerning TGFβ levels in healthy children and pediatric Marfan patients without and with sartan treatment.
Methods: Since 2008, 672 children presented to our pediatric Marfan clinic, of which 152 showed a heterozygous pathogenic *FBN1* variant. From 2017 to 2020 we measured TGFβ in 31 MFS patients (6.1 ± 5.6 y, m:52 %) with a proven *FBN1* variant in our TGFβ study. In 11 patients (5.5 ± 3.8 y, m:73 %), medical prophylaxis with Valsartan was indicated. After starting sartan therapy, we repeated the TGFβ measurement after 1, 2, 6, 12 hours, 6 and 12 months. In addition we measured TGFβ levels in 125 healthy children (9.4 ± 5.7y; m: 58 %).
Results: In healthy children TGFβ declines the older the children get. TGFβ is significantly lower in adolescents in comparison to younger children ($p = 0.001$). In comparison, baseline TGFβ serum levels were 6137 (95%CI: 5360;6914) pg/ml in MFS patients without medication and 6526 (95%CI: 6027;7026) pg/ml in healthy children ($p>0.05$). TGFβ serum levels reached a significant nadir 6 hours after sartan-administration with an average reduction of 1288 pg/ml (95%CI: 85;2491, $p<0.05$), followed by a return towards baseline at the 11.5-hour mark (delta from baseline -590 pg/ml (95%CI: - 2932;1751, $p>0.05$)). Follow-up measurements of TGFβ at random times of the day after 12 months did not show a significant decrease of TGFβ levels in comparison to baseline.
Conclusion: In our sartan (Valsartan) treated patient collective TGFβ was effectively suppressed by the treatment during the day with a nadir 6 hours after drug administration. The pathogenesis of MFS determined by TGFβ can thus be influenced in a relevant way. We conclude that the application of sartans twice daily seems reasonable and essential for a constant suppression. Whether TGFβ is suitable as a screening parameter or for therapy monitoring is still unclear.

Keywords: Marfan syndrome, TGFβ, sartan

P-486

An oligogenic model for cctga: whole-genome sequencing shows a mutation load effect at 156 loci involved in cardiac patterning

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Background and Aim: Congenital heart disease (CHD) is a major public health issue. About 20% of CHD correspond to a misalignment between cardiac segments. We focused on a rare CHD, the congenitally corrected transposition of the great arteries (CCTGA). The aim of the study is to understand the mechanisms and genes that control the alignment of cardiac chambers, and thus lead to the CCTGA.
Methods: We analysed a cohort of 45 trios (43 sporadic cases and 2 familial cases) of non syndromic CCTGA by next generation sequencing analysis: 15 trios with whole-genome sequencing and 30 trios with whole-exome sequencing Selection of variants was done according to four criteria: extremely rare frequency in public databases (DB frequency <1%, prediction of a deleterious effect by prediction softwares (SIFT and PolyPhen-2), strong conservation between species and localization in a gene involved in the morphogenesis of the cardiac tube, the outflow tract development and/or in establishment of cardiac left-right asymmetry in zebrafish, mouse or human.
Results: Under the hypothesis of Mendelian models, or de novo genomic alterations, no consistent major gene involved in the disease could be identified. Under the hypothesis of a complex model of inheritance with incomplete penetrance, a total geneset of 156 genes matched each of our selection criteria. The highly heterogeneous combinations of susceptibility rare variants, mostly inherited from the healthy mother and father respectively, functionally converge to give rise to the CCTGA phenotype. To prove that a given allele combination is associated with the CCTGA phenotype, we showed a significant enrichment of rare variants in cases compared to healthy controls ($p = 9E-7$, OR 1.44)
Conclusion: Taken together, the cases could be explained by a mutation load of segregated alleles at loci mostly involved in heart tube looping, outflow tract morphogenesis and establishment of left-right asymmetry. Our data contradict a monogenic mode of inheritance, favouring a multigenic origin of CCTGA, where both parents harbour genetic predisposition variants for the disease (susceptibility alleles) and both transmit this load of variants to the affected offspring, explaining the low recurrence risk.

Keywords: congenital heart disease, CCTGA, genetics

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P-487 / Moderated Poster

Induced pluripotent stem cell-derived chagas disease model highlights the role of cardiomyocytes as sentinels of the host-parasite interaction

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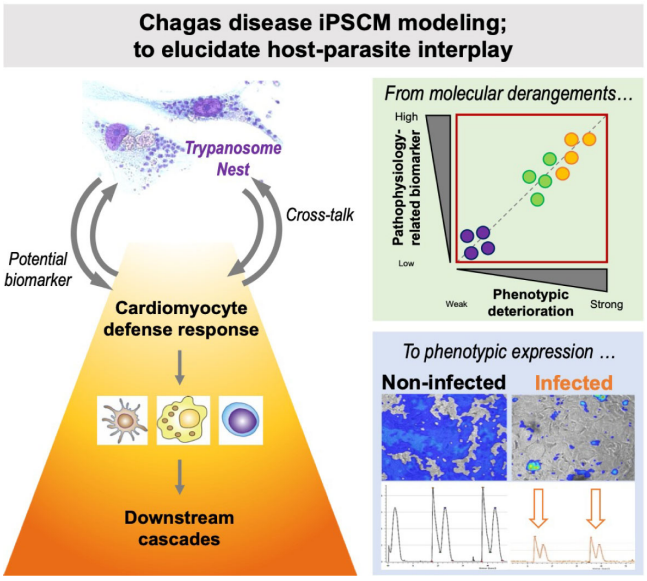
Background and Aim: Chagas heart disease (CHD), a malignant cardiomyopathy caused by *Trypanosoma cruzi* protozoan infection, is the most highly prevalent 'neglected tropical disease' still in urgent need for effective therapeutics. Lack of reliable disease models and relevant biomarkers have hampered development of novel treatment measures. CHD is also one of few model cardiac diseases that provide opportunity for our approaching the yet-to-be elucidated cardiomyocyte response against non-ischemic stress.
Methods: CHD was modeled *in vitro* using human induced pluripotent stem cell-derived cardiomyocytes (iPSCMs) to investigate host-parasite interplay. Motion vector analysis was performed to evaluate the model's phenotypic recapitulation. Gene expression analysis, as well as multiplexed immunoassay for cytokine expression, was performed to investigate the underlying molecular derangements and identify the associated factors of phenotypic severity. Phenotype was assessed before and after benznidazole administration to assess the model's adequacy as a tool to study drug efficacy/toxicity.
Results: Trypanosome-infected iPSCMs, differentially expressing >1400 genes compared with non-infected controls, recapitulated the rhythmic and contraction/relaxation abnormalities in CHD. Trypanosome infection provoked cardiomyocyte defense responses characterized by pro-inflammatory signatures. Pro-inflammatory molecular markers (interleukins and C-C chemokines), correlating to the extent of phenotypic deterioration, were identified, and further validated using multiple protozoan strains. Therapeutic benznidazole doses, above its median effective dose (5–10 μM), only partially rescued the phenotypic abnormalities (100% recovery in beating rate, while 62–65% recovery in contraction/relaxation kinetics). 'Functional cardiotoxicity' was observed even below the recognized toxic dose (>250 μM) for benznidazole.
Conclusion: Cardiomyocytes act as sentinels of pathogen encounter and its emitting defense response lie upstream in the immune hierarchy. Molecular signatures of the cardiomyocyte response provoked by trypanosome invasion constitute candidate biomarkers reflecting magnitude of myocardial stress. Finally, the iPSCM-based *in vitro* CHD model is suitable, not only for studying the host-parasite interplay, but also for revolutionizing trypanocidal drug development.

Keywords: Chagas disease, Cardiomyopathy, Human induced pluripotent stem cell-derived cardiomyocyte, Disease modeling

Graphical abstract of our working hypothesis



Morphology

P-488

Histological structure of pulmonary veins in tapvc patient as independent risk factor of postoperative obstruction

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Background and Aim: Postoperative pulmonary venous obstruction is the main reason of mortality in long-term period. Despite of different type of surgery (conventional or "sutureless") used for TAPVC correction, postoperative pulmonary venous obstruction is still an actual problem. We believe that tissue differences determine the tendency of pulmonary vein stenosis after surgery in the patients with TAPVC. The aim of this study is to reveal histological structure of pulmonary vein in a patient with any type of TAPVC and define the role of this differences in postoperative obstruction.
Methods: The tissue samples of the anomalous pulmonary veins were taken from neonatal patients with different types of TAPVC during surgery, including redo surgery due to postoperative obstruction. The routine H&E staining and some special stains (Verhoeff–Van Gieson, Masson) were used to evaluate tissue structure of anomalous veins. The immunohistochemical assessment (smooth muscle actin, troponin T) of this samples was done

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additionally. The results were assessed by electronic microscopy and compared with normal.
Results: The obvious histologic difference was obtain in TAPVC samples. The wall of the anomalous pulmonary vein was presented by intimal, medial and an adventitial layer. There was no myocardial layer around anomalous pulmonary veins in all types of TAPVC in contrast to normal pulmonary veins. The postoperative pulmonary venous obstruction was caused by neointimal proliferation and medial fibrosis, but the severity of this process can be varying degrees.
Conclusion: The histologic features of anomalous pulmonary veins are typical for all types of TAPVC, which is the non-modified reason of postoperative obstruction. This circumstance can be considered as independent risk factor of postoperative obstruction for all TAPVC patients. Despite of this one, it seems that postoperative obstruction is not mandatory for all patients after TAPVC surgery, which means that postoperative obstruction has multifactorial nature, realizing under condition of addition risk factors, especially hemodynamic ones.

Keywords: total anomalous pulmonary venous connection

Other

P-489 / Moderated Poster
Determining factors for hs-troponin T and NT-probnp in a cohort of healthy children (the life child study)

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Background and Aim: NT-proBNP and hs-Troponin T (hs-TnT) are commonly used biomarkers in the evaluation of cardiac stress in children despite lacking a true understanding of influencing factors. As part of the prospective longitudinal population-based cohort study, "LIFE child" in Leipzig, Germany, we provided new reliable reference values for both cardiac markers from well children and investigated for associations between those markers and age, sex, pubertal status, BMI and serum lipid levels. Subsequently, we explored associations with an individual´s growth hormone status represented by Insuline-like Growth factor-1 (IGF1) and IGF1-Binding Protein-3 (IGF-BP3) levels.
Methods: Serum values, medical history and anthropometric data provided by 2522 children aged 0.25-18 years were collected and analyzed as per study protocol.
Results: NT-proBNP values decreased throughout childhood and dropped rapidly with advancing pubertal stage. The strong negative association between lower NT-proBNP values and higher BMI or elevated serum lipids was also identified between lower NT-proBNP values and higher IGF1 and IGF-BP3 levels. This interaction was modulated by sex and age for IGF-BP3 levels, the latter effect was not found for IGF1 levels.
Approximately half of the hsTnT measurements were below the detection limit. However, young children aged 3 to 6 months frequently had values exceeding the adult cut-off limit. HsTnT levels in boys increased from pubertal stage 2 onwards. Our data showed a positive association between hsTnT and BMI but a negative association with LDL and triglyceride levels in boys. Higher hsTnT levels also correlated with higher IGF1 and IGF-BP3 values with minimal differences between the sexes and an age-dependency for the correlation with IGF1 levels.
Conclusion: We have identified important associations between certain metabolic markers and reference values for NT-proBNP and hsTnT based on data from our large cohort of healthy children.

Keywords: Troponin T, proBNP, reference values, metabolic marker, puberty, growth hormones

Morphology

P-490
Isolated congenital coronary artery anomalies in children: a 42-year single center experience

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Background and Aim: Data on the incidence of isolated coronary anomalies in the pediatric population is limited. We evaluated the incidence of these anomalies during a 42-year period in a tertiary pediatric care center
Methods: Medical records of all patients undergoing selective and non-selective coronary angiography between March 1979 and November 2021 were evaluated retrospectively. All patients with additional congenital heart or acquired coronary disease were excluded from the study.
Results: During a 42 year time period 3398 pediatric patients between 0 to 18 years of age underwent coronary angiography. Seventeen patients (0.5%) (10 females, 7 males; age 21.79 ± 39.18 months) had isolated coronary anomalies: Anomalous Left Coronary Artery from the Pulmonary Artery (ALCAPA) was diagnosed in 9/17 patients (52.9%), coronary fistula was seen in seven (41.2%) and singular coronary ostium in one patient (5.9%).
All patients with ALCAPA presented with either signs of heart failure or myocardial infarction. Eight of them (88.9%) showed ischemia on ECG at the time of admission. Six of the seven (85.7%) patients with coronary fistula and the patient with singular coronary ostium were asymptomatic.
Prior to 1990 diagnosis of ALCAPA or coronary fistula was made solely by angiography, while echocardiography only raised suspicion by showing i.e., poor left ventricular function or left atrial and ventricular enlargement. Seven of the nine ALCAPA patients underwent re-insertion of the anomalous left coronary artery into the aorta while two underwent Takeuchi repair. There were three intraoperative deaths, all of which were prior to 1990. Four of the seven patients with coronary fistula underwent either surgical ligation (two patients) or percutaneous coil embolization (two patients).

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Conclusion: The incidence of isolated coronary anomalies in the pediatric population at our tertiary pediatric care center is 0.5%, which is coincidentally similar to the incidence in the general population. Although patients often present with only unspecific symptoms early diagnosis is important because of the poor prognosis if left untreated, especially ALCAPA.

Keywords: ALCAPA, Coronary fistula, Heart failure, Children, Sudden Cardiac Death

P-491

Cardiac isomerism: a 20 year retrospective cohort at a single institution

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Background and Aim: Cardiac isomerism or 'isomerism of the atrial appendage' is found in about 0,4-2% of congenital heart defects and is associated with a complex spectrum of anomalies. We reviewed isomerism cases from the last 20 years at a single institution.

Methods: Retrospective review of 48 patient records (Jan 2000-Dec 2020). Case selection was based on echocardiographic findings suggesting cardiac isomerism. Definitive diagnosis was made post-mortem or peri-operatively by identifying symmetrical morphological left or right atrial appendages.

Results: Left atrial isomerism (LAI) was diagnosed in 24 (10 confirmed), right atrial isomerism (RAI) in 24 cases (18 confirmed). Prenatal diagnosis was made in 33 cases (17 LAI; 16 RAI). Unconfirmed cases consist of 5 terminations of pregnancy were no autopsy was performed and 15 live-born patients who did not undergo cardiac surgery or autopsy. In LAI, 21 cases (87,5%) have anomalous systemic venous return, being the only cardiovascular abnormality present in 10 patients. Conduction or rhythm abnormalities were present in 11 cases (45,8%). In RAI, only complex univentricular heart defects were found with a large proportion of AVSD, transposition of the great arteries and pulmonary stenosis/atresia. Splenic function is disturbed in 86% of all live-born patients. While other extra-cardiac defects are more incidental, malrotation is more prevalent in this cohort (>10%) than in the general population. After exclusion of termination of pregnancy (n = 11) and patients lost to follow-up (n = 2), overall survival for isomerism patients is 60%, with a median time to follow-up of 9 years. Compared to RAI, survival rates in patients with LAI are much higher: 84% (median follow-up 6yrs) compared to 31% (median follow-up 12yrs). However, for those LAI patients that need univentricular palliation (n = 3), survival rates are low (33%) and similar to RAI. Main causes of death are primary cardiac failure, abstinence of life-prolonging care and severe sepsis.

Conclusion: Survival rates for patients with cardiac isomerism are low for those who need univentricular palliation. These results can be used when counseling parents. Additional pathology like (functional) asplenia and malrotation add to the burden of disease.

Keywords: cardiac isomerism, univentricular heart, prenatal counseling

P-492

Left cervical and circumflex aortic arch with aberrant right subclavian artery arising from a kommerell diverticulum: a rare vascular ring

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Background and Aim: Aortic arch anomalies are a relatively common cause of vascular ring. Cervical aortic arch is a rare anomaly characterized by an elongated arch extending at or above the level of the clavicles. The most common form is characterized by an omolateral descending aorta with normal rise of brachiocephalic vessels.

Methods: We report a rare case of vascular ring due to a left cervical circumflex aortic arch with aberrant right subclavian artery arising from a Kommerell diverticulum

Results: A newborn (age: 20 days, weight: 2,8 kg) come to our attention due to stridor during crying. Echocardiography showed a significant stenosis of the left pulmonary artery, at proximal tract. Aortic arch was not well profiled, but a right subclavian artery was suspected. Catheterization clarified the diagnosis: left cervical circumflex aortic arch with aberrant right subclavian artery arising from a Kommerell diverticulum. Three-dimensional angio-CT scan confirmed the diagnosis and demostrated a significant compression of esophagus and trachea by the descending aorta. The anterior displacement of trachea caused the distortion of the left pulmonary artery at proximal tract with consequent stenosis.

Conclusion: Vacular rings are an important cause of stridor in neonatal age. Several vascular anomalies may cause of vascular rings. In our experience, this is the first case of vascular ring due to a left cervical circumflex aortic arch with an aberrant right subclavian artery arising from a Kommerell diverticulum in a patient with stridor and stenosis of left pulmonary artery.

Keywords: Vascular ring, cervical aortic arch, circumflex aorta, aberrant subclavian artery, Kommerell diverticulum

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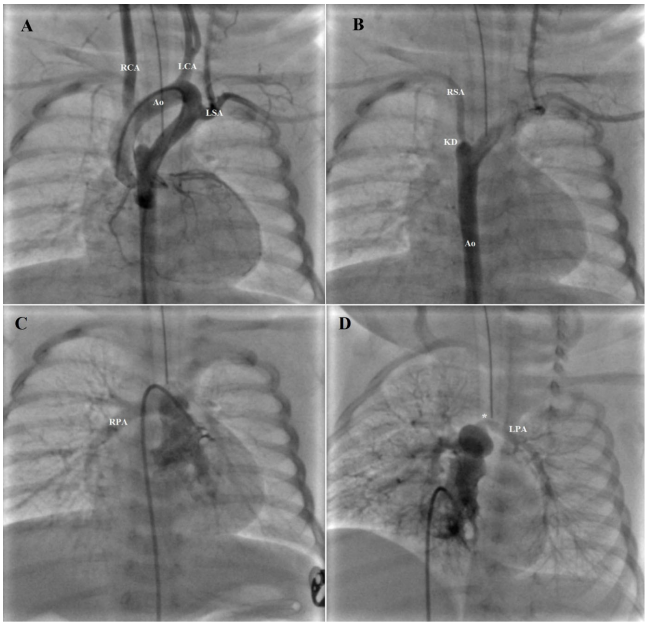
Aortography



Ascending (A) and descending (B) aortography in antero-posterior view showed: a cervical left aortic arch (overriding the left bronchus) with right-sided descending aorta (circumflex aorta), with an aberrant right subclavian artery arising from a Kommerell diverticulum. A right arterial duct was recanalized with a multipurpose catheter from the Kommerell diverticulum (C). Right ventriculography (D) showed an angulated origin of the left pulmonary artery with significant hypoplasia (*). Abbreviations. Ao: Aorta, KD: Kommerell Diverticulum, LCA: Left common Carotid Artery, LPA: Left Pulmonary Artery, LSA: Left Subclavian artery, RPA: Right Pulmonary Artery, RSA: Right Subclavian Artery, RCA: Right common Carotid Artery.

P-493

Aberrant subclavian artery with vertebral artery origin from the carotid artery: incidence in infants requiring cardiac surgery in the first 6 months

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Background and Aim: Aberrant subclavian artery (ASA) has an incidence of 1-2% in patients with congenital heart disease. According to the literature, these patients frequently have an abnormal origin of the vertebral artery from the carotid artery. The purpose of this study was to describe the incidence and clinical significance of this combination among children undergoing cardiac surgery in the first 6 months of life.
Methods: We identified in our hospital database all patients who underwent cardiac surgery in the first 6 months of life from 01/2013 until 06/2021. Patients with double aortic arch were excluded. The findings of echocardiography were reviewed regarding aortic arch laterality and branching pattern. Findings of other imaging modalities were included if available. Patients with ASA were evaluated carefully for possible origin of a vertebral artery from the carotid artery.
Results: A total of 554 infants were included in this study. ASA was found in 41 pts (7.4%). These included 34/512 pts (6.6%) with left aortic arch (LAA) and 7/42 pts (16.7%) with RAA. ASA occurred in 12/178 patients (6.7%) requiring surgery for hypoplastic aortic arch or coarctation. Origin of the right vertebral artery from the right carotid artery was detected in 5 pts with ASA and LAA representing 0.9% of the entire cohort and 14.7% of pts with ASA and LAA. Since this anomaly can mimic normal branching of the innominate artery diagnosis of ASA was missed at initial examination in our first patient while the correct diagnosis was made at initial echocardiography in the other four. Two of these pts had aortic arch hypoplasia or coarctation.
Conclusion: ASA was present in 7.4% of our cohort and in 6.7% of children requiring aortic arch surgery or coarctation repair. In 5/34 patients with ASA and LAA (14.7%) we detected anomalous origin of the right vertebral from the right carotid artery mimicking normal right innominate artery branching. Since preoperative detection of ASA is import in infants requiring aortic arch surgery or coarctation repair preoperative echocardiography of the aortic arch should not rely exclusively on branching of the first vessel but also attempt direct visualization of possible ASA.

Keywords: Aberrant subclavian artery, vertebral artery origin, aortic arch laterality

Other

P-494 / Moderated Poster

Infections predispose kawasaki disease, a nationwide population-based case-control study of incident cases 1987-2018 in sweden

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Background and Aim: The etiopathogenesis of Kawasaki disease (KD) is insufficiently understood. In addition to genetic factors, a multitude of environmental elements have been suggested as causative in the onset of KD. Various infectious agents repeatedly been implicated in the pathogenesis, however with inconclusive findings. The aim of the study was to investigate the load of exposure to infections predisposing KD diagnosis, using data from the Swedish national health-care registers.
Methods: The study was established by identifying all cases diagnosed with KD diagnosed in Sweden during 1987–2018. Matched controls were assessed to the cases using the Swedish Total Population Register. Data on infections diagnosed inpatient and outpatient care was retrieved from the National Patient Register, which contains data on all Swedish residents. The study was conducted as a case-control study, where exposure to infections was assessed in cases and controls. Odds ratios (ORs) for KD were estimated using conditional logistic regression.
Results: The study comprised of n = 1,774 (61% male) cases, matched to n = 17,731 controls. Overall, a history of infection was associated with Kawasaki disease with an OR of 2.3 (95%

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CI 2.0–2.5). Stratified by anatomical site, infections in the skin, respiratory, urogenital, and gastrointestinal tract were all significantly associated with Kawasaki disease. Notably, temporal stratification revealed a prominent clustering of infectious events during the month preceding Kawasaki diagnosis; OR of infections preceding Kawasaki by 8–14 days was 11.6 (95% CI 8.1–16.6), and for 1–7 days 91.5 (95% 66.1–126.8). Similarly, prescription of antibiotics was significantly associated with Kawasaki. Interestingly, bacterial and viral infections associated with the disease with ORs of similar amplitude.
Conclusion: Our findings indicate that infections are closely linked with the development of KD. The study suggests the acute inflammation linked to infections, rather than any pathogen, is causing the syndrome.

Keywords: Kawasaki disease, epidemiology, risk factors, infections, inflammation

P-495 / Moderated Poster
Preliminary results of follow-up of patients with multisystem inflammatory syndrome associated with COVID-19

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Background and Aim: Kawasaki-like (multisystem inflammatory) syndrome associated with SARS-CoV-2 infection is characterized by acute severe systemic vasculitis, often with multi-organ dysfunction and cardiac involvement. Although most patients recover, long-term outcomes are poorly studied [Gema de Lama Caro-Patón et al., 2021; Guimarães D. et et al., 2021; Sharma C. et al., 2021].
Methods: We analyzed the results of laboratory, clinical, radiological, ECG and EchoCG data in the dynamic observation of 15 patients (M 9, 1.5-16 yo, m = 7) in 3 months after the suffered MIS-C.
Results: At the disease onset high refractory fever was observed in all cases, symptoms of Kawasaki disease in 12 (80%) of them, shock with multi-organ dysfunction - in 8 (53.3%), including symptoms of acute heart failure - in 5 (33%), concomitant in two cases with severe left ventricular dilatation with low LV EF. Myocardial damage was seen in 11 patients (73%), pericarditis in 12 (80%), coronary dilatation in two (13%); troponin level increased in 5 (33%), CK-MB - in 5 (33%), BNP - in 3 (25%). After 3 months, there were no signs of myocardial dysfunction and/or cardiomegaly in any patient, troponin and BNP levels normalized in all patients, a moderate increase of CK-MB was seen in 8 (53%), and coronary dilatation persisted in one patient. Arrhythmias were documented at onset in 9 (60%) patients, 3 (20%) after 3 months ($p = 0.028$).
Conclusion: preliminary results of follow-up of children after MIS-C demonstrate favorable course in the majority of patients by clinical, laboratory, ECG and echocardiographic data. Further observations are needed to determine the long-term prognosis.

Keywords: Kawasaki-like syndrome, COVID-19; multisystem inflammatory syndrome in children; MIS-C;

End-stage Heart and Lung Disease

P-496
Long-term mechanical assist device in paediatric patients: our 15-year experience

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Background and Aim: About 10-15% of paediatric patients with severe left ventricular dysfunction will need a mechanical assist device. The limited lifespan of the ECMO (Extracorporeal membrane oxygenation) and the increasing number of patients in the transplant waiting list have forced the development of long-term mechanical circulatory support devices. The Berlin Heart EXCOR© is the most implanted long-term device in children and the only one in newborns and infants. The aim of the study is to analyse clinical and survival results after the implant of a long-term mechanical assist device in paediatric patients in the end stages of heart failure.
Methods: Between January 2006 and July 2021 33 Berlin Heart EXCOR© devices were implanted in paediatric patients. We analysed preoperative and intraoperative characteristics, as well as complications after the implant (haemorrhagic, infectious, thromboembolic and neurological). Outcomes of the patients were also evaluated (transplant vs. death). Statistical analysis with STATA IC 15.
Results: The mean age was 52 ± 52 months (range 1-180), mean weight 18 ± 16 kg (range 3-65). The underlying heart disease was myocardiopathy in 67% and congenital heart disease in 33%. The Berlin Heart EXCOR© device was univentricular in 64% of the patients. Surgical indication was bridge to transplant in 97% of the cases. Mean duration of the mechanical support was 76 ± 73 days (range 1-315). Postoperative complications were common: 61% infectious and 52% neurological (41% ischaemic and 35% haemorrhagic). The mortality of haemorrhagic strokes was 67%, and ischaemic 14%. Since 2019, when our anticoagulation protocol was modified (Heparin changed to Bivalirudin), no mortal strokes were observed. 67% of the patients has been transplanted, 30% has died (60% neurological cause). 1 patient is still on mechanical circulatory support.
Conclusion: As a response to the increasing number of paediatric patients in end stages of congestive heart failure, the use of the Berlin Heart EXCOR© device has spread, specially if the intention is to use it as bridge to transplant. Neurological events, mostly haemorrhagic, are the most feared because of high mortality. Nevertheless, it is an encouraging alternative as a long-term mechanical assist device in children, allowing them to reach the transplant in 67% of the cases.

Keywords: mechanical circulatory support, ventricular assist device, heart failure, heart transplant, children, congenital

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P-497

Application of slide tracheoplasty in pediatric tracheal stenosis and a mid-term follow-up

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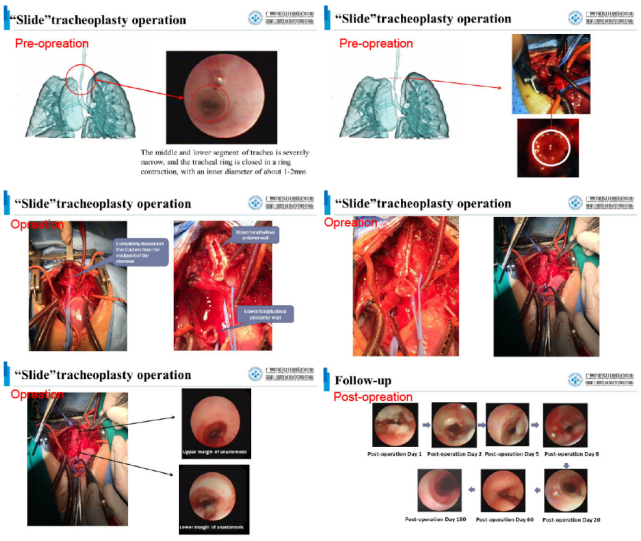
Background and Aim: To summarize the clinical data of slide tracheoplasty in the treatment of congenital tracheal stenosis, and to evaluate its efficacy.
Methods: From January 2015 to May 2019, 27 cases of congenital tracheal stenosis were treated surgically. The routine preoperative fiberoptic bronchoscopy revealed that their stenotic tracheas were all O-shaped, and in some children, a fiberscope with a diameter of 2.8 mm could not pass through their stenotic tracheas. Ultravist was used for tracheography to evaluate the tracheal condition. The minimum diameters of the stenotic tracheas ranged from 1-3 mm. There were 6 cases of short-segment tracheal stenosis and 21 cases of long-segment tracheal stenosis (3 of which were almost complete tracheal stenosis). Before surgery, 14 (51.9%) patients required ventilator-assisted ventilation. All patients underwent corrective surgery for congenital heart disease under mild hypothermic cardiopulmonary bypass, along with slide tracheoplasty. After surgery, Pulmicort Respules was used for aerosol treatment for a week. Patients were followed up via fiberoptic bronchoscopy for 2 months to 4 years.
Results: Among the 27 patients who underwent congenital tracheal stenosis surgery, 2 (7.4%) died at the early stage, both of whom had complete tracheal stenosis. One (3.7%) had anastomotic dehiscence; one (3.7%) had recurrent anastomotic leak, gave up treatment, and died. In one case, anastomotic granuloma formed under fiberoptic bronchoscopy at 2 days after surgery; the granuloma detached at 8 days after surgery, and the trachea was patent. In one case, the stenotic O-shaped ring was not completely relieved, which made it difficult for the patient to wean from the ventilator. Except one case lost to follow-up, no other deaths occurred outside the hospital. According to fiberoptic bronchoscopy during follow-up, the trachea was patent without granuloma formation. Four patients had audible laryngeal stridor after movement, and the others had no discomfort.
Conclusion: Slide tracheoplasty is effective in the treatment of congenital tracheal stenosis. During the surgery, the tracheal blood supply should be preserved as much as possible to prevent poor healing of anastomotic stoma and to thoroughly relieve the tracheal stenosis. The children can obtain satisfactory quality of life after surgery.

Keywords: Tracheal stenosis; Pulmonary artery sling; SLIDE tracheoplasty; Congenital heart disease.

Operation chat



A case of preoperative, intraoperative and postoperative follow-up

P-498

Pulmonary hypertension in children with scurvy

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Background and Aim: Pulmonary hypertension(PH) in children is a serious condition and could be life threatening.
Methods: We reported 2 cases of previously healthy children with reversible PH secondary to scurvy.
Results: The first case is a previously healthy 2-year-old boy who presented with vomiting, diarrhea and fever for 5 days. After rehydration, he had recurrent episodes of hypotension with intermittent abdominal pain. Fluid resuscitation and inotropic medication were given. Then he suddenly collapsed in PICU. After 4-minute cardiopulmonary resuscitation, his hemodynamic was stabilized. The majority of the diagnostic workups was unremarkable except PH from echocardiogram with estimated systolic pulmonary artery pressure(PAP) of 74 mmHg. Pulmonary CT angiography was normal. Transient PH was diagnosed and milrinone was started prior

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to referral. At our institute, physical examination revealed a loud second heart sound without significant murmur. Echocardiogram showed right atrial (RA) and right ventricular (RV) enlargement and mild tricuspid regurgitation (TR) with estimated systolic PAP of 29 mmHg. Right cardiac catheterization revealed PAP 28/13(mean 20) mmHg and PVRi 1.8 WU.m2. According to his nutritional history and sclerotic rim at epiphysis in chest films, vitamin C level was tested with low level result. The second case is a 6-year-old boy with acute dyspnea. He experienced a month of low-grade fever, swollen left knee and progressive dyspnea. Physical examination revealed oxygen saturation at 90%, tachypnea and tachycardia. Subconjunctival pallor and diffuse petechiae were seen. Cardiovascular examination showed loud second heart sound. He had a tender, swollen left knee with a limited range of motion. Echocardiogram indicated moderate TR with estimated systolic PAP of 90 mmHg (systolic blood pressure 90 mmHg). Pulmonary CT angiography showed ground glass opacity at left lower lung field. Milrinone was given. Right cardiac catheterization showed PAP 66/38(mean 50) mmHg and PVRi 5.7 WU.m2. Other medical conditions were excluded. Since he had improper dietary intake and clinical suspicious of scurvy, vitamin C was tested and reported undetectable level. Administration of vitamin C in both cases rapidly reversed the pulmonary hypertension.

Conclusion: Association between vitamin C deficiency with PH has been rarely reported. Since malnourishment-induced PH is a reversible condition, vitamin C deficiency should be recognized and treated.

Keywords: pulmonary hypertension, scurvy

P-499

Eleven years of pediatric ecmo; single center experience in zagreb, croatia

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Background and Aim: Extracorporeal membrane oxygenation (ECMO) is a complex and high-risk treatment used as a last option in neonates and children with severe cardiorespiratory failure. It is a life-saving procedure for highly critical patients which fail to respond to a standard approach with intravenous fluids, vasoactive, and/or inotropic drugs. In the following report, a brief overview of pediatric ECMO in University Hospital Centre (UHC) Zagreb is provided.

Methods: This study is a retrospective review of all pediatric cases who underwent a veno-arterial (VA) or veno-venous (VV) ECMO protocol between August 2009 and August 2020. Patients with both cardiac and respiratory indications for ECMO were included, from neonatal to adolescent age.

Results: There were 52 ECMO runs identified over that period; data were complete for 45 cases with similar equal distribution between sex. Median age was 8 months (range 1 day - 18 years), median body weight 6.7 kg (from 2.3 to 90 kg), and median height 65 cm (from 45 to 190 cm). The overall survival rate was 51%. Circulatory failure was an indication for ECMO in 38 patients. In 17 patients ECMO was started after providing cardiopulmonary resuscitation (E-CPR). Associated malformations were present in 12 cases, four with one of the variants of the heterotaxy syndrome and two with Down syndrome. Normal neurological outcome was found in 76% of children who were discharged from the hospital. Variables during ECMO treatment significantly associated with higher mortality rate were renal failure with renal replacement therapy ($p<0.001$) and presence of neurological deficit ($p< 0.001$). In conclusion, the use of ECMO is expanding in our center; there were 14 ECMO runs between 2009 and 2016, and 38 ECMO runs in the last four years.

Conclusion: This study shows an increasing trend of pediatric ECMO use in Croatia. Overall survival rate in our cohort is comparable to the data published in the literature. The use of hemodialysis was shown to be associated with higher mortality. Normal neurological outcome in 76% of the children who survived ECMO is a good pledge for the future of ECMO treatment in our institution.

Keywords: cardiopulmonary resuscitation, epidemiology, extracorporeal membrane oxygenation, pediatric intensive care unit

P-500

Use of ventricular assist devices in children; single center experience in zagreb, croatia

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Background and Aim: End-stage heart failure in children unresponsive to medical therapy has limited treatment options. Surgical options are heart transplantation or implantation of durable ventricular assist devices (VAD). The main difference between VAD's is based on flow used for providing cardiac output: pulsatile or continuous flow VADs. The VAD support in children is used as a bridge to transplantation, bridge to myocardial recovery or bridge to decision

Methods: Our study is a retrospective review from September 2016 to September 2021 of all children from Croatia who underwent VAD insertion at University Hospital Center Zagreb, Croatia. The clinical outcome was collected prospectively. The decision for biventricular or left ventricular support, although discussed before operation, was made in operating theatre. The anticoagulation management was mainly dependent on Edmonton protocol.

Results: The total of seven children were included in the study. The median age was 53 months (IQR 30 - 133 months), with the mean duration of VAD support of 35 days (1- 124 days). The long term survival was achieved in three patients (43%). The reason for end-stage heart disease and an indication for VAD support in one child was intractable arrhythmia while in five of them cardiomyopathy. In the last one, the VAD was started after unsuccessful repair of supravalvular aortic stenosis. The common causes of death were thromboembolic events or bleeding. In one patient, due to ischemic stroke, successful neurointervention was performed during ongoing VAD support. Four of the patient underwent a hemodialysis in the time of VAD support. Of three survived children, one is in permanent vegetative state.

Conclusion: Our experience with VAD support in children is limited but our learning curve certainly is steep. Problems in the postoperative management have arisen due to coagulation problems, thrombosis and bleeding. When the signs of intracranial events are observed, an emergent neurointervention should be considered regardless of the possible numerous transport problems.

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Keywords: ventricular assist device, children, cardiomyopathy, end-stage heart disease, neurointervention

P-501

Serum proteome analysis in children and young adult with pulmonary hypertension: an observational cohort study

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Background and Aim: Background Pulmonary arterial hypertension (PAH) in children is a complex and heterogenous condition, mostly complicating congenital heart disease (CHD) and carrying an increased risk of mortality. Clinical management is difficult due to the scarcity of data to have an accurate risk assessment. We aim to identify protein proteomic features that correlate with events (life threatening arrhythmias, HF admissions) and might impact on clinical course in children.
Methods: In this observational cohort study, we enrolled children with idiopathic and CHD-PAH in referred to our Institute. Blood samples were collected during routine clinical visits, together with clinical status, biochemical and no-invasive tests. Outcomes were considered deaths or transplant, rehospitalization for heart failure and arrhythmias during one year of follow up. We performed a proteomic analysis (SWATH) on serum aiming at the identification of a panel of prognostic proteins, evaluating their differential regulation in patients grouped by diagnosis. A number of potentially prognostic proteins were then confirmed by statistical analysis: ANOVA, ROC and Kaplan-Meier curves were used.
Results: We enrolled 34 children (mean age: years 13.8 ± 10.7, median: 10) with idiopathic and CHD-PAH. 347 potentially quantifiable proteins were identify and trough SWATH analysis we selected in these patients a panel of nine highly expressed proteins. On the potentially prognostic proteins, a further statistic validation was performed using ANOVA test and subsequently by ROC and Kaplan-Meier curve. Proteins validated in all analysis were haptgloblin (HPT), thyroxine-binding globulin (TGHB) and transgelin-2 (TAGL2). HPT significantly correlates with an increased risk of admissions for HF. TGHB and TAGL2 significantly correlate with an increased risk of life threatening arrhythmias and particularly with a high risk of arrhythmias in CHD group.
Conclusion: HPT, TGHB and TAGL2 identified in patients with PAH correlates with a high risk of arrhythmia and HF hospitalization and might have a use in clinical management and the evaluation of new therapies in children.

Keywords: Pulmonary Hypertension, Proteome Analysis, Children

P-502

Identification of failing fontan patients as potential candidates for venous assist cannula by breath-modulated realtime quantitative blood flow CMR

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Background and Aim: Right heart assist with a venous cannula in combination with a pulsatile assist device is a new bridging option for failing Fontan patients, but identification of suitable patients remains challenging. First fitting of the new devices was investigated. Then a previously published clinical risk score (RS) was applied and compared to non-invasive hemodynamic CMR data.
Methods: Axial 3D whole heart DIXON stacks (TR/TE1/TE2 = 3.8/1.37/2.3 ms) were acquired as part of a routine protocol investigation. Fitting of 3 differently sized cannulas was assessed by measurements of central venous vessel calibers. Real-time phase-contrast cardiovascular magnetic resonance was performed using non–ECG triggered echoplanar imaging (temporal resolution = 24 to 28 ms) in the superior vena cava (SVC)/inferior vena cava (IVC) during regular (RB) and forced breathing (FB). The simultaneously recorded patients' respiratory cycle was divided into expiration (Exp), end-expiration (EExp), inspiration (Insp) and end-inspiration (EInsp) to generate respiratory-dependent stroke volumes (SVs). The imaging data were matched with physiological data and analyzed with home-made software.
Results: Fifty-two Fontan patients could be included in the study (2019-2021). In 24 Fontan patients with preserved (22.4 ± 6.5 ys, 17.5 ± 7.9 ys after Fontan, RS ≤3) and in 22 Fontan patients with failing (16.6 ± 8.6 ys, 13.9 ± 7.4 ys after Fontan, RS>3) Fontan circulation complete flow calculation was possible. In Thirty-six of all (69.2%) no systemic venous abnormalities impeded fitting of cannulas. In preserved Fontans high differences of rel SVi in the IVC during RB could be detected ranging from 59.9% in EInsp to -46.6% in Exp. The effect was increased during FB to 116.2% in Insp and to -114.8% in Exp. Failing Fontan circulation resulted in minor differences of rel SVi in the IVC ranging from 21,2% in Insp to -21.3% in Exp under RB and from -17.0% in Insp to -30.0% in Exp. ROC statistics in absolute IVC SVs results in a cut-off value of 38.3 ml (Sensitivity 90%, $p<0.01$, EInsp) for RB and 41.3 ml (Sensitivity 90%, $p<0.005$, EInsp) for FB.
Conclusion: The newly constructed venous cannulas fitted in a high percentage of Fontan patients. End-inspiratory IVC flow facilitates the identification of possible candidates with failing Fontan circulation and should be included into decision making.

Keywords: Failing Fontan Venous Cannula Realtime Flow

P-503

Post-transplant lymphoproliferative disorder following paediatric heart transplantation: a retrospective review over 30 years in a national centre

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Background and Aim: Of 46 patients who underwent paediatric orthotopic heart transplantation (OHT) through our national shared care transplant service over 31 years, three patients (5%) developed PTLD. All 3 cases of PTLD presented in a cluster over a 6 month period during 2021 despite a lack of obvious change to our post-transplant medical practice.
Methods: Retrospective review.
Results: Case 1: A 4 year-old boy 3 years post OHT for complex congenital heart disease presented with signs and symptoms of

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critical airway obstruction and was initially diagnosed with infective supraglottitis. Following re-presentation and biopsy, this was confirmed as EBV-positive polymorphous PTLD in an unusual site; laryngeal PTLD is rare. Staging demonstrated localised disease. The patient failed standard therapy (rituximab, then R-CHOP) and ultimately was successfully treated with EBV-specific cytotoxic T lymphocytes (CTL). This required elective tracheostomy prior to CTL therapy. The treatment was successful and the patient was decannulated prior to discharge following 4 negative biopsies, the most recent at 6 months following treatment completion.
Case 2: A 7 year-old boy presented one year following OHT for restrictive cardiomyopathy with recurrent intermittent abdominal pain over several months. The patient developed fever and raised inflammatory markers and an abdominal ultrasound demonstrated ileocolic intussusception. Laparotomy and biopsy confirmed EBV-positive monomorphic PTLD acting as a lead point. Resection and stoma formation was performed. Staging demonstrated localised disease to the GI tract. The patient didn't respond to rituximab therapy and converted to cyclophosphamide and prednisolone therapy.
Case 3: A 6-year old girl presented 5 years following OHT for severe hypertrophic cardiomyopathy in the context of PTPN11 Noonan's syndrome with symptoms of loose stool for several months despite normal growth and absence of other symptoms. Infectious stool studies were negative. The patient ultimately underwent endoscopy which confirmed gastric EBV-positive polymorphous PTLD. Staging demonstrated localised disease. The patient didn't respond to rituximab therapy and converted to cyclophosphamide and prednisolone therapy.
Conclusion: PTLD affects 5-10% of patients post paediatric heart transplant and may occur at any time. Clinical features may be subtle or atypical and these cases highlight the importance of extra-vigilance in addition to patients potentially requiring second or even third-line therapies.

Keywords: Paediatric heart transplant; Post-transplant lymphoproliferative disorder; Cytotoxic T Lymphocytes

P-504
Pulmonary hemodynamics before and after heart transplantation in childhood

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Background and Aim: Pulmonary hypertension (PH) in children undergoing heart transplantation (HTx) may limit feasibility before and outcome after HTx. Therefore, we aimed on factors influencing pulmonary hemodynamics in children undergoing HTx during long-term follow up.
Methods: Cross sectional, single center case study analysing pulmonary hemodynamics in all consecutive pediatric patients (< 18 years) undergoing HTX over a 15 year period (2005 to 2019) determined by the Fick principle during cardiac catheterization.
Results: Twenty-three patients (female 15) underwent HTx at a median (IQR) age of 3.9 (0.9-8.2) years resp. body weight of 12.5 (8 – 23.3) kg with a time interval between first clinical signs and HTx of 1.1 (0.4-3.2) years. Indications for HTx included end-stage heart failure due to cardiomyopathy (17/23, 74%), single ventricle congenital heart disease (CHD) with failing Glenn or Fontan physiology (5/23, 22%), and intracardiac tumor (1/23, 4%). Before HTx, pulmonary artery pressure (PAP) was elevated by 26 (18.5-30) mmHg, pulmonary capillary wedge pressure (PCWP) 19 (14-21) mmHg, left ventricular enddiastolic pressure (LVEDP) 17 (13-22) mmHg. Transpulmonary pressure gradient (TPG) was 6.5 (3.5-10) mmHg and pulmonary vascular resistance (Rp) 2.65 WU*m2 (1.87-3.19). After Htx, first invasive hemodynamic evaluation PAP decreased to 20.5 (17-24) mmHg, PCWP 14.5 (10.5-18) mmHg ($p<0.05$), LVEDP 16 (12.5-18) mmHg, TPG 6.5 (4-12) mmHg, Rp 1.49 (1.08-2.74) WU*m2. At last hemodynamic assessment at 4.0 (1.4 – 6) years follow up, pulmonary hemodynamics normalized with PAP 19.5 (17-21) mmHg ($p<0.05$), PCWP 13 (10.5-14.5) mmHg ($p<0.05$), LVEDP 13 (10.5-14) mmHg, TPG 7 (5-9.5) mmHg, Rp 1.58 (1.38-2.19) WU*m2 ($p<0.05$). Of note, in CHD patients PAP increased ($p<0.05$) immediate after HTx at first evaluation and decreased until last follow up ($p<0.05$), while in CMP patients there was a continuous decline of PAP values immediate after HTx ($p<0.05$) and during long term follow up. PH medical management was reduced according to the hemodynamic results.
Conclusion: While PH before HTx is frequent, after HTx the normalization of PH starts during acute postoperative care and continues during long-term follow up. In CHD patients with failing Glenn or Fontan physiology undergoing HTx the normalization of pulmonary hemodynamics needs longer time.

Keywords: Pulmonary hypertension, Heart Transplantation, Failing Glenn, Failing Fontan

Nursing

P-505 / Moderated Poster
Adolescents with CHD. -is their illness perception impacting quality of life?

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Background and Aim: Congenital heart disease (CHD) is a condition with a variation of anatomical lesions. Depending on the CHD severity and its manifestation it may affect everyday life for a young person. Living with a long-term disease may cause a psychosocial burden due to disease-related stress. There is a lack of knowledge about young people's perception of their CHD and whether this as well as CHD severity, age, and sex affect Quality of Life (QOL) The aim was to (i) describe adolescents' QoL and perceptions of their congenital heart disease. (ii) explore to what extent illness perception, CHD severity, age and sex are associated with quality of life (QoL) in adolescents with CHD. β
Methods: Of the 202 adolescents between 14-18 years who participated, 44.1% were male, and the mean age was 15.7 (±1.1). QoL was measured with a linear analogue scale ranged from 0-100 (best imaginable QoL). The Brief Illness Perception Questionnaire (B-IPQ) was used to measure illness perception. B-IPQ has eight

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items, assessing cognitive illness representations and emotional representations range (1-10). Sociodemographic data were collected with the questionnaires. Descriptive data was expressed in mean and standard deviation. Statistical analyses were performed with multivariable regression analyses.
Results: Mean score for self-assessed QoL was 83 (±18.3). The adolescents scored high on Timeline (8.2±3), Personal control (6.9±2.9), Understanding (6.6±2.6), and Treatment control (6.1±3.4). Concerns (2.9±2.4), Consequence (2.8±2.2), Emotional response (2.7±2.5) and Identity (2.6±2.4) was given low scores. The regression analysis displayed after adjusting for CHD severity, age and sex, that perceiving less illness consequence (β-2.0, S.E 0.9), a less emotional response (β-2.7, S.E 0.8) or having a better understanding of the illness (β.9, S.E 0.5), were associated with better QoL.
Conclusion: The findings revealed that adolescents showed an overall good QoL and perceived CHD as a low threat. Further, perceptions of less Illness consequence, less emotional response and having a better understanding were positively associated with QoL. Hence emphasis should be given to support adolescents to increase their understanding of their illness and to provide emotional support.

Keywords: Congenital Heart Disease, Adolescents, Illness perception, Quality of life

P-506
"The only thing i wonder is when i will have surgery again" - everyday life for children with RVOT anomalies during assessment for heart surgery

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Background and Aim: Children with complex right ventricle outflow tract (RVOT) anomalies often require repeated cardiac surgeries, thereby needing repeated thorough preoperative assessments during their lifetime. Since this is assumed to pose a heavy burden on them the aim of this study was to explore how children with RVOT anomalies experience their everyday life during the assessment and one year after the decision on whether to perform a new cardiac surgery.
Methods: Individual interviews were conducted with nine children between 9 to 17 years of age on three occasions from 2014 to 2016. The interviews were analyzed with thematic analysis.
Results: The analysis yielded three themes and eight subthemes. The theme "Me and the heart disease" concerns the children's experiences related to the heart disease. Almost all children described various degree of tiredness and signs of adaptive behavior in their everyday life. The theme "Being me" concerns the children's sense of self, in which their heart disease did not feature. The theme "Being placed in someone else's hands" describes children's experiences during and after the preoperative assessment. Even if fear for having heart surgery was awakened, they could reassure themselves with the thought that no surgery was needed this time, at least until the decision was delivered. Some experienced shortcomings in communication and participation.
Conclusion: Although these children had symptoms from their heart disease, they did not always pay attention to them, presumably by developing adaptive behavioral strategies to manage their heart disease in their everyday life. Traces of an adaptive behavior were also obvious with respect to preoperative assessment. The findings underscore the need for child-centered approach to achieve individualized support based on the child's experiences and to ensure that these children are involved in their own care.

Keywords: Children, Right ventricle outflow tract anomalies, Everyday life, Repeated heart surgery, Thematic Analysis

P-508
Nursing rounds in the paediatric cardiac intensive care unit – an instrument for increasing the quality of care and patient satisfaction

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Background and Aim: The progressive development of medical treatment of paediatric patients in a cardiac intensive care unit (CICU) leads to an increasing complexity of nursing care. The implementation of nursing rounds is intended to ensure the quality of care, increase patient satisfaction and enhance trust between patients and nursing staff. The aim is to establish the nursing round as a fixed element of patient care. It should be individually designed with the involvement of the patients and their families.
Methods: Based on a literature review, a policy for the nursing rounds was developed and introduced. The nurs-ing rounds are performed weekly for patients hospitalized in the CICU for more than seven days. A standardized and comprehensible care planning with nursing interventions and documentation is dis-cussed with several nursing staff members. Information is collected, questions about the patient are clarified and nursing priorities and goals are defined. The nursing documentation is kept up-to-date and nursing interventions are evaluated. After one year, the nursing rounds were evaluated in a nurs-ing expert meeting using the 5-D Model of Appreciative Inquiry and optimized based on the gained experience.
Results: The quality of care has been significantly improved by the implementation of the nursing rounds. The nursing process is continuously applied and interventions are individually adapted to the patient. The exchange between the nursing team results in an optimal integration of resources and knowledge. A holistic care of the patient and his family with the goal of evidence-based nursing can be guaranteed. The integration of the family in the nursing rounds also strengthens their trust and integrity.
Conclusion: Nursing rounds should be introduced as a standardized instrument in every CICU into the clinical rou-tine. This will increase nurses' awareness of the importance of continuity of the nursing care process. At the beginning, nurses may be uncertain. However, this changes with time, as they recognize the positive effects on patient care through the participatory approach. The nursing rounds sustainably increase the quality of nursing care and provide an instrument to meet new nursing challenges in a mindful manner.

Keywords: Nursing rounds, Family centered Care, PICU, Quality of Care

P-509

Exploring mothers' experiences of caring for a child with a single ventricle heart condition. a phenomenological study

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Background and Aim: Medical innovations have ensured children born today, with a single ventricle heart condition, not only survive, but are likely to reach adulthood. However, this improvement comes with costs of morbidity and premature death. The available treatment is complex, including multiple high-risk cardiac by-pass operation, nevertheless remains palliative. As a result, the majority of these children face long-term complications, which impacts not only their life, but also affects the lives of their families. There is a paucity of rigorous research exploring families' experiences with a child with a single ventricle heart condition. Therefore, this study aims to explore families' lived experiences caring for a child with a single ventricle heart condition, the physical and psychosocial impact on their child and their families.
Methods: Underpinned by interpretive phenomenological, five semi- structured interviews were conducted with mothers of a child with a single ventricle heart condition. A purposive sampling frame was developed to recruit participants from UK heart-charities, between March and September 2019. Interviews were audio-recorded and transcribed verbatim by the researcher and analysed utilising Interpretive Phenomenology Analysis.
Results: This study recognises each single ventricle heart condition journey is a unique and complex experience. The diagnosis is life changing and impacts families' lives on many levels. The unique circulation comes, for all represented children, with costs of physical limits impacting their everyday performance as well restricts lifestyle choices. In contrast, psychosocial impacts emerged as prominent outcome of the whole family. Despites, families experience positive outcomes, also cope well when accepting the condition in their life. The complex treatment pathway requires complex support systems, although the findings were able to identifies significant support gaps.
Conclusion: The revealed rich insightful findings and added knowledge demonstrates greater awareness and more holistic support at different levels are required for children with a single ventricle heart condition, whereby a focus on the whole family is indispensable. This enhanced understanding might be a meaningful first step towards overcoming identified burdens and add quality to the palliative life of these children, as well their families.

Keywords: single ventricle heart condition, families' experiences, lived experiences, psychosocial impact, phenomenology

P-510

Family management in families of children with congenital heart disease, down syndrome and both conditions

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Background and Aim: Congenital Heart Disease (CHD) is among one of the most common birth defects. One group of children at increased risk for CHD are children with Down syndrome (DS); between 40 to 50% of children with DS have CHD. The aim of this international collaboration, guided by the Family Management Style Framework, was to examine how families of children with CHD, families of children with DS, and families of children with both conditions compare on the six Family Management Measure (FaMM) scales.
Methods: Parents completed an online survey (available in both Spanish and English) that included the six FaMM scales. Clinicians and group leaders of support groups and foundations in Spain, Ecuador and the United States shared information about the study with eligible families.
Results: 560 parents completed five of the FaMM scales (273 from Spain, 146 from the United States and 141 from Ecuador). The Parental Mutuality scale was completed by the 425 partnered parents. One-way analysis of variance F tests were used to compare means of each FaMM scale across the nine groups: CHD, DS, and both conditions for parents from Spain, Ecuador, and the United States. Means for each scale were ranked across the nine groups to assess patterns of family management. Overall, there was a pattern of positive family management. There were significant differences for five of the FaMM scales. Parents of children with CHD from Spain reported the most positive family management and parents of children with both CHD and DS from the United States reported the least positive family management. At the country level, family management was generally less positive for families from Ecuador.
Conclusion: Management of chronic conditions can be difficult for families, especially if the family member has more than one chronic condition. This is especially true if the condition affects the individual's cognitive ability. More research is needed to understand how social determinants of health influence family management in families of children with CHD, DS, and both conditions. For this study, it is likely that families from Spain and the United States had greater access to resources than families from Ecuador.

Keywords: Family, Congenital Heart Disease, Down Syndrome, Family Management

P-511

Cost-effectiveness of a transition program for adolescents with congenital heart disease: the stepstones project

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Background and Aim: Transition programs for adolescents with chronic conditions have been proposed to ensure continuity of care and independence in adulthood. However, evidence on the health economic impact of transition programs is limited. The aim of this study was to evaluate the cost-effectiveness of a transition program for adolescents with congenital heart disease in comparison to usual care, in a Swedish healthcare setting.

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Methods: A cost-effectiveness evaluation was performed alongside the STEPSTONES randomized controlled trial in two university hospitals in Sweden. Adolescents aged 16 were sampled and followed-up for 2.5 years. Intervention patients (n = 58) received a multi-component intervention in addition to usual care and control patients received only usual care (n = 67). A micro-costing approach was used to identify health resource use in addition to intervention costs for training and staff. Self-reported health status was collected at age 16, 17 and 18. Incremental cost-effectiveness ratios were calculated using a longitudinal regression and non-parametric bootstrap.
Results: Health resource use and costs in usual care differed negligibly between the groups, with the control group having slightly higher usual care costs (m = SEK 2672, 95% CI -29687 – 30703). However, total costs were higher in the intervention group because of the transition program (m = SEK 21,049, 95% CI -5966 – 54,423). Differences in self-reported health status between the intervention and control group were small (m = -0.011, p = 0.57).
Conclusion: The intervention and control groups were similar in health outcomes and total costs differed due to the additional costs of delivering the intervention. Thus, these findings alone cannot provide evidence on whether transition programs should be implemented based on only short-term health economic outcomes. To guide this decision, future research is needed on the long-term effects of transition programs and evaluations that capture benefits in other societal domains for young people living with chronic conditions.

Keywords: Adolescent, Congenital heart defect, Economic evaluation, Randomized controlled trial, Transition of care

P-512

Post discharge surveillance of the surgical site in the cardiac paediatric population using the innovative digital web platform ISLA care

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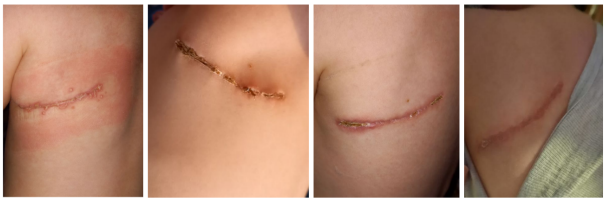
Background and Aim: Surgical Site Infections (SSIs), is a serious post-operative complication associated with prolonged hospital stay, readmission, reintervention and significant financial burden. For the last 5 years our SSIs rate has been between 3-4%. With infections mostly occurring in the post discharge group. We aimed to formalise our post discharge surveillance using the Isla care visual record (https://www.islacare.co.uk/).
Methods: From August 2021 we adopted Isla care web platform for surveillance of the surgical wound. It commences pre discharge with a baseline photo taken on the cardiac ward, this is printed along with customised wound care advice for the patients. The post discharge surveillance was for 30 days. A weekly text with a unique link for wound photo submission was sent to the patient's parents/carer. Monitoring of submissions was performed by dedicated clinical nurse specialists. New submissions are alerted by email, the photo is then reviewed and actioned accordingly. We examined the number/response rate of paediatric cardiac patients recruited into post discharge surgical site surveillance after cardiac surgery between August 2021 and December 2021.
Results: Between August and December 2021 33 patients have been recruited and 99 submission requests generated with 79 responses, which shows an 80% response rate. An increasing response rate is noted from the start of the project with November and December showing a response rate of 89%. 9% of patients who submitted a wound picture were escalated to the surgical team for review. Those patients either required a physical review, start of antibiotics treatment or increased surveillance, for more than 30 days.
Conclusion: Use of a secure system such as Isla care and its ease of use by both clinical staff and parents is an improvement on current processes. Submission at a time convenient to them, without the struggle and upset to daily routines compared to a hospital visit. The use of this platform has allowed us to rapidly respond to changes, escalate care as necessary without the unnecessary burden on families and no impact on clinical care. Further audit will be required to understand why some parents/carer declined to submit a wound picture.

Keywords: surgical site infection, surveillance, wound photo

Wound timeline



From left to right, chronological sequence of wound pictures sent by patient's parents/carer soon after discharge, at 2 weeks and 1 month after discharge. Infection was identified promptly and treated accordingly, surveillance was increased until the wound healed (picture on the right, 7 weeks post discharge)

P-513

Practice development project up-date of a recommendation for positioning after glenn surgery at the children's hospital

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Background and Aim: Hypoplasia of the heart is one of the most severe forms of heart defects, especially when one ventricle is completely missing or underdeveloped. The treatment of these heart defects always consists of palliation, which involves three operations. The Glenn operation is the second operation and takes place in fourth to sixth month of life. The treatment of this heart defect is complex Postoperative management in particular requires specialist knowledge and well-trained professionals. One of these aspects of care is Glenn positioning after Glenn surgery. This has an impact on patient convalescence and postoperative management. The Glenn position consists of a supine position and a 30° upper body elevation. The aim of this positioning is to lower the preload and afterload of the heart, and an increase in cardiac output and supports passive blood flow. The risks of complications are high and therefore, nurses play an important role in the early detection, prevention, and management of these complications. The current Glenn positioning is reviewed for its evidence. Based on these results, an evidence-based recommendation for the positioning will be developed.
Methods: The project was developed using participatory action research, project management and practice development. The

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project organization was interprofessional. A literature search was conducted in various databases and enquiries were sent to various centers in Switzerland and abroad in order to review the evidence for the Glenn positioning.
Results: A recommendation for the up-date Glenn positioning was developed based on a literature review and experts. In addition, a poster and a training concept were prepared for the nursing staff of intensive care unit to be able to train the nursing team in a next step. The recommendation for the revised Glenn positioning was worked out in several steps, whereby the close cooperation with the expert group was very much appreciated and we received many valuable inputs.
Conclusion: Two added values for practice were gained through this project. First, the Glenn material was revised and adapted to the current evidence. Secondly, a training and poster were developed to fill the knowledge gaps in the nursing team regarding the correct use and benefits of Glenn positioning.
Keywords: Glenn position, Glenn surgery, nursing care

Psychosocial

P-514
The frequency of anxiety in pediatric patients with mitral valve prolapse and the effect of mitral valve anatomy on anxiety levels

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Background and Aim: Mitral valve prolapse (MVP) and anxiety are both seen with high frequency in the general population. MVP usually cause non-specific complaints, and these complaints may be accompanied by psychiatric disorders such as anxiety and panic disorder. In this study, we investigated the frequency of anxiety and the effect of mitral valve anatomy and functions on anxiety levels in children diagnosed with MVP.
Methods: We examined a total of 91 children (< 18 years old) in our Pediatric Cardiology Outpatient Clinic between February 2019 - February 2021. Among them, the patient group consisted of 53 pediatric cases diagnosed with MVP by echocardiographic examination and the control group consisted of 38 healthy children with similar age and gender distribution with the patient group and they had normal echocardiography. Beck anxiety questionnaire was applied to both groups. In the echocardiographic examination, type of MVP and the degree of mitral regurgitation (MR) were evaluated in the patient group. Classical MVP groups had a maximal leaflet thickness (MLT) of equal or > 5 mm, and if MLT of less than 5 mm, they were classified as non-classical MVP.
Results: The mean age was 13.5 years and female and male ratio was 1.78 in the patient group. The mean "Beck anxiety score" was 13.4 in cases with MVP and 6.18 in the control group ($p<0.001$). Anxiety was determined in 35 cases (66%) with MVP and 11 cases (28.9%) in the control group ($p<0.001$). No significant effect of gender on anxiety scores were determined in both groups. Anxiety scores were mild in 17, moderate in 11, and severe in 7 of the cases in the patient group. In the control group, mild anxiety was detected in 7 cases and moderate anxiety in 4 cases. The presence and degree of MR did not have a significant effect on anxiety scores. No significant difference was observed between the anxiety scores of classical and non-classical MVP cases ($p>0.05$).(Figure-1)
Conclusion: Anxiety often accompanies in pediatric patients with MVP. The effect of the myxomatous structure of the mitral valve and the presence of MR on anxiety could not be defined in this study.
Keywords: Anxiety, children, Mitral valve prolapse, echocardiography

	The patient with MVP N= 53	Control group N= 38	p
The mean age (years) (SD)	13,45 (3,2)	12,9(3,1)	0.439
Minimum-Maximum	7-18	7-17	
Female/ Male number of cases	34/19	24/14	0.923
(F/M ratio)	(1.78)	(1.71)	
Body mass index z score mean (SD)	-1.04(1.3)	0.08(1.2)	0.001
BECK anxiety score,, mean (SD)	13.4(10.0)	6.2(6.1)	0.001
Anxiety			
Absent N (%)	18(34,0)	27 (71,1)	0.001
Mild	17 (32,1)	7 (18,4)	0.001
Moderate	11 (20,8)	4(10.5)	
Severe	7(13.2)	0(0)	

The demographic characteristics, echocardiographic findings and Beck anxiety scores in the patient group and control group were show

P-515
Acute cardiopathy in children with anorexia nervosa and physical instability: echocardiographic findings and reversibility at MID-term follow-up

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Background and Aim: Cardiac and electrocardiographic changes are present in anorexia nervosa children at diagnosis or during stable disease, and most recover after body-weight treatment. It is unknown if anorexia nervosa children with phisical instability requiring hospitalization present higher cardiovascular-risk profile requiring specific treatment and care. Therefore, the aim of study was to investigate prevalence of cardiovascular abnormalities in those children at admission and to assess the effects of hospital treatment on their reversibility at mid term follow up.
Methods: We retrospectively studied all anorexia nervosa children requiring admission at Paediatric Institute in the period 2015-2019. Anorexia nervosa acute cardiopathy at admission was defined by the presence of at least two of the following clinical FINDINGS: pericardial effusion, mitral regurgitation, bradycardia, mitral billowing, aortic regurgitation, altered LV morphology and ECG abnormalities. Echocardiographic data were compared with those registered at 3-8-month follow-up after treatment and with data from a healthy population.

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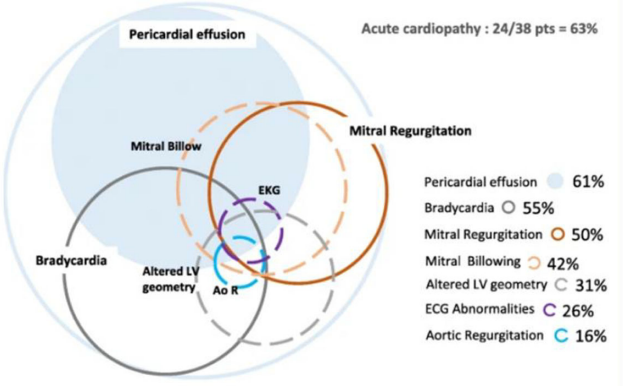
Results: 38 anorexia nervosa children were examined. Between children with phisical instability requiring hospitalization, prevalence of anorexia nervosa cardiopathy at admission was high, 63% (24 patients, figure 1). Pericardial effusion, bradycardia and mitral regurgitation were present together in 26% of patients. Most cardiovascular changes recovered at follow-up. Anorexia nervosa cardiopathy was associated with significantly lower left ventricle end-diastolic diameters and mass, and higher E wave, E/A and E/e' ratios and left ventricle sphericity index values vs healthy population and vs anorexia nervosa children without cardiopathy ($p<0.05$). Left ventricle global longitudinal strain was significantly reduced only in anorexia nervosa cardiopathy patients but recovered, whereas end-diastolic diameters, E/A ratio and sphericity index values remained impaired.
Conclusion: Among anorexia nervosa children requiring hospitalization, those presenting several cardiac findings together express an acute anorexia nervosa cardiopathy which is characterized by worse LV filling, geometry and subclinical myocardial deformation impairment. Despite hospital treatment, in those patients some alterations persist at mid-term follow-up. On going studies are investigating the effects of different cardiac loading manouvres and nutritional therapies on cardiac changes.

Keywords: anorexia nervosa; acute cardiopathy; advanced echocardiography

Fig. 1
A



Prevalence of acute cardiac findings in anorexia nervosa children with physical instability ad admission

Psychosocial

P-516
Application of remote videoconferencing to home monitor and support patients with major congenital heart disease during the COVID-19 pandemic

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Background and Aim: The COVID-19 pandemic presented unique challenges to global healthcare provision. Face-to-face outpatient care was dramatically reduced as a consequence. This study implemented a remote videoconferencing call (VC) service delivered by a mobile app to continue close monitoring of our most vulnerable patients in their home environment. The patient cohort was followed up at a regional paediatric cardiology centre.
Methods: Patient recruitment began in September 2020, concluding in December 2021. Most participants were identified in the newborn/infant period and consisted of a mixture of cyanotic and acyanotic congenital heart disease. All study participants required regular, frequent outpatient monitoring in usual circumstances. Parents/guardians of identified patients received written and verbal explanation of study aims and objectives prior to giving written consent. The videoconferencing interface was delivered by PEXIP Infinity Connect Mobile app and conducted by experienced medical and/or nursing staff. This app was already a well established method of communication within the Regional Paediatric Cardiology Team. Primary outcome measures included admissions to hospital and avoidance of hospital attendances. Clinical proformas including growth parameters and clinical observations was recorded at each virtual appointment. Patient and parent related research data was collected at the first, fourth and eighth appointment. A select number of patients were given home saturation monitors and weight scales.
Results: A total of 32 patients were enrolled. 164 VCs were delivered (patient mean = 5.8). The average age at recruitment was 10.8 weeks. 18 patients had surgical intervention during the study period. There were 11 admissions to hospital directly resulting from the VC; the commonest indication was abnormal oxygen saturations (45%). 33 hospital attendances were avoided; the commonest concern reported by parents was difficulty related to infant feeding (36%).
Conclusion: Qualitative and quantitative measurement tools showed reduction in parental anxiety. The study was well received by participating families. There was prompt identification of unwell children on VCs as well as providing advice to prevent unnecessary hospital attendance. Videoconferencing technology proved very user friendly and proved to be a very valuable adjunct to the provision of good patient care during challenging circumstances.

Keywords: videoconferencing COVID-19 home monitoring complex congenital heart disease

P-517
Impact of a child with congenital heart defect on mother's quality of life

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Background and Aim: Congenital heart defects (CHD) are the most frequent congenital anomalies (1%). Mortality of patients has greatly reduced due to advances in surgery and postoperative care resulting in long term morbidity which represent a burden on parents. The aim of this study was to assess the impact of rising a child with congenital heart defect on maternal quality of life.
Methods: A prospective, longitudinal study included 32 mothers of children with unrepaired CHD evaluated with Impact of a Child with Congenital Anomaly on Parents (ICCAP) questionnaire, developed by dr. Petra Honig-Mazer and her team from Erasmus University in Rotterdam. The ICCAP questionnaire measures six domains: contact with caregivers, social network, partner relationship, state of mind, child acceptance, and fears

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and anxiety. Normality of data was evaluated with Shapiro-Wilk test and Pearson rank test was used for assessing the correlation between variables in Stata version 13. Statistical level of significance was considered $p <= 0.05$.

ICCAP dimensions
Contact with caregivers
Doctors clearly explain things
I have good interaction with nurses
Doctors take enough time to listen to me
I am satisfied about my contacts with doctors
Social network
My friends support me
My colleagues are understanding
People around me support me
My friends help me with practical things
I can share worries with my family
I can share worries with good friends
Partner relationship
I feel my partner sympathizes with me
On important issues I agree with my partner
My partner is someone I can talk to
Generally I am happy with my partner
My relationship with my partner is good
State of mind
I feel sad
I feel angry
I wonder whether I am to blame for my child's CA
I feel guilty
Child acceptance
My child fits into my life
My child is welcome in our family as it is
I am happy with my child
I wish my child was never born
Fears and anxiety
My child faces a difficult life
I expect my child will be able to function well
The CA is/are a heavy burden on my child
I wonder whether my child will ever be healthy
I am very anxious about all the tests on my child
My child is facing a difficult period
My child is the same as other children
I worry a great deal about my child's health
I doubt whether my child will be happy later
I fear about my child's expectations for the future
My child is handicapped
I feel I can't do enough for my child
My child will be able to have a normal life later

Results: The mean age of mothers was 30 years (limits of 16-42 years) when the questionnaire was completed, with a mean age at birth of the child with CHD of 29 years. Majority of mothers (43.3%) had only middle school education with a percent of 62.5 of them living in the country area. The mean age of their children was 11 months (limits of 1-51 months), 81.5% of them without prenatal diagnosis of CHD. The majority of children had siblings, with a mean of 2 children per family (limits of 1-8 children).
For contact with care givers items, maternal education level showed significant positive correlation (0.38). A significant correlation was found between social network items and maternal age (-0.35), maternal education level (0.44) and number of children per family (0.39). State of mind items significantly correlated with number of children per family (-0.44), maternal education level (0.38) and maternal age at child birth (-0.42). Fears and anxiety category turned out to be negatively correlated with number of children per family (-0.38) and child age (-0.37).
Conclusion: ICCAP questionnaire identifies domains in which mothers are at risk and need intervention to obtain a better quality of life for them and for their children.

Keywords: maternal quality of life, congenital heart defect, Impact of a Child with Congenital Anomaly on Parents questionnaire

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Cognitive and social-emotional development in two- to three-year-old children with critical congenital heart disease

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Background and Aim: The overall survival of children born with critical congenital heart disease (CHD) has considerably improved due to improvement of perioperative care and surgical techniques. However, these children are at risk for neurodevelopmental sequelae. Little is known about the early cognitive and behavioral outcome of children with critical CHD. This study aimed to assess the cognitive and social-emotional development of two to three-year-old children with critical CHD and to identify potential risk factors for worse outcomes.
Methods: Seventy-two children, aged two- to three years old, with critical CHD, who underwent open-heart surgery with the use of cardiopulmonary bypass (CPB) within the first six weeks of life, were prospectively included. The Bayley Scales of Infant and Toddler Development, Wechsler Preschool and Primary Scale of Intelligence, Child Behavior Checklist (CBCL), and Teacher Report Form (TRF) were used for assessment of cognitive and behavioral development. Medical records were consulted to gather surgical and clinical data.

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Results: Cognitive scores were within average range in 70.8%, superior to very superior in 22.2%, and below average in 7%. Cognitive index scores were lower in children with single ventricle pathology (SVP, $p = 0.003$), more cardiac procedures ($p = 0.001$), and longer total operating time of surgeries with the use of CPB ($p = 0.01$). For the CBCL questionnaire, Total problem score was in the normal range for 91.3% and for the TRF, 92.7% scored within normal range on Total problems score. CBCL Internalizing problem scores were higher in children with SVP ($p = 0.03$) and in those with multiple surgeries with CPB ($p = 0.02$), whereas higher gestational age at birth was correlated with lower Internalizing problem scores ($p = 0.02$).
Conclusion: Children two- to three years of age with critical CHD mostly develop within normal range in both cognitive and social-emotional domains. However, those with either SVP or who underwent multiple cardiac procedures appear to be most at risk for adverse developmental outcome. Future research should focus on perinatal and perioperative modifiable factors to allow targeted intervention for improvement of developmental outcomes.

Keywords: Critical Congenital Heart Disease, Cognitive Development, Behavioral Development

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Cognitive functioning in identical twins with complex heart disease and chronic hypoxia

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Background and Aim: Lower intelligence and cognitive functioning for cyanotic than for acyanotic children with congenital heart defects (CHD) have been reported, a finding attributed to the degree of hypoxemia. We aimed to assess the relationship between degrees of chronic hypoxia, reduced intelligence quotient (IQ), and cognitive and social functioning in pair identical twins.
Methods: We examined two 14 years old identical twins with CHD and chronic hypoxia. Twin I - situs solitus, levocardia, complete atrioventricular septal defect (AVSD), moderate atrioventricular insufficiency (AVI), and anomalous systemic venous drainage. Pulmonary arterial hypertension (PAH) and Eisenmenger syndrome were contraindications for corrective surgery. Twin II – situs inversus, levocardia, AVSD with common atrium, spontaneous closure of inlet VSD, moderate AVI, anomalous systemic venous drainage. An unsuccessful attempt for corrective surgery was made at the age of 2 years. Persistent systemic PAH was the indication for pulmonary vasodilator therapy (Bosentan) since 2016 in both. Methods for medical evaluation: 6-minute walking test (6MWT), blood-gas analysis; for psychological evaluation: Wechsler test to determine IQ and battery of tests for cognitive and social functioning. The results were compared in 3-year period.
Results: Twin I had higher oxygen saturation, better physical capacity, better results at the IQ test, and fewer problems in cognitive functioning, than Twin II. The finding did not change over time. Twin II, who had more pronounced hypoxemia and poorer physical capacity, shows a lower IQ score, with rapid decline through the years. She also has deterioration in cognitive functioning, memory, concentration of attention, and social dysfunctions.
Conclusion: This pair of identical twins have the same genetic basis, the same social environment but different levels of oxygen saturation, physical activity, learning abilities, and ability to respond to social and cognitive requirements. The findings in Twin II can be attributed to chronic hypoxia and maybe open heart surgery in early childhood. The poor social environment, poor parent's capacity, and neglecting behavior can be the cause as well. The whole family needs systematic psychological training and social support. Further examinations and discussions about the reason for the quick drop of IQ level of the Twin II are needed.

Keywords: cognitive functioning, chronic hypoxia, identical twins, congenital heart disease

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