



Cardiology in the Young

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YIA-1

Macitentan Reverses Early Obstructive Pulmonary Vasculopathy in Rats: Early Intervention in Overcoming the Survivin-mediated Resistance to Apoptosis

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Objectives: We tested the hypothesis that a novel endothelin receptor antagonist macitentan reverses the early and/or late stages of occlusive pulmonary vascular disease in rats.

Methods: Rats with pulmonary arterial hypertension (PAH), which were produced by combined exposure to a vascular endothelial growth factor receptor inhibitor Sugren 5416 and hypobaric hypoxia for 3 weeks (SuHx), were assigned to receive macitentan (30 mg/kg, once daily by oral gavage) or vehicle during 3–5 weeks (early study, n = 40) or during 5–8 weeks (late study, n = 38) after Sugren injection. A baseline SuHx PAH rat group, sacrificed just before treatment initiation, was present in each study to evaluate the reversal of disease during treatment. A P-value of <0.05 was considered to be statistically significant.

Results: Compared with vehicle-treated PAH rats and baseline SuHx PAH rats, the macitentan-treated rats significantly showed decreases of the proportion of occlusive lesions in all small arteries (outer diameter: 15–50 µm) per lung section in the early study (baseline PAH rats: 33.8 ± 4.4%, vehicle-treated PAH rats: 41.5 ± 4.1% and macitentan-treated PAH rats: 17.9 ± 2.8%), a finding consistent with the reversal of right ventricular systolic pressure (control rats: 19.4 ± 1.4 mmHg, baseline PAH rats: 78.3 ± 4.9 mmHg, vehicle-treated PAH rats: 79.5 ± 6.4 mmHg and macitentan-treated PAH rats: 50.3 ± 5.0 mmHg), indices of right ventricular hypertrophy and medial wall thickness. Macitentan ameliorated but did not reverse the proportion of occlusive lesions in the late study. Although macitentan significantly decreased the proportion of Ki67 positive lesions in both studies, macitentan significantly increased the proportion of cleaved caspase 3 positive α smooth muscle actin (αSMA) cells in occlusive

lesions and significantly decreased an anti-apoptotic molecule survivin protein and mRNA expression in lungs and the proportion of survivin positive αSMA cells in occlusive lesions in the early study but not in the late study.

Conclusions: Macitentan reversed early but not late obstructive pulmonary vascular disease in rats. This reversal was associated with the suppression of survivin-related resistance to apoptosis and proliferation of αSMA cells in occlusive lesions. These findings could be mechanistic basis for the efficacy of early treatment and give an insight into later appearance of resistance to treatment for this disorder.

YIA-2

Does reversal of flow in the fetal aortic arch in second trimester aortic stenosis predict hypoplastic left heart syndrome?

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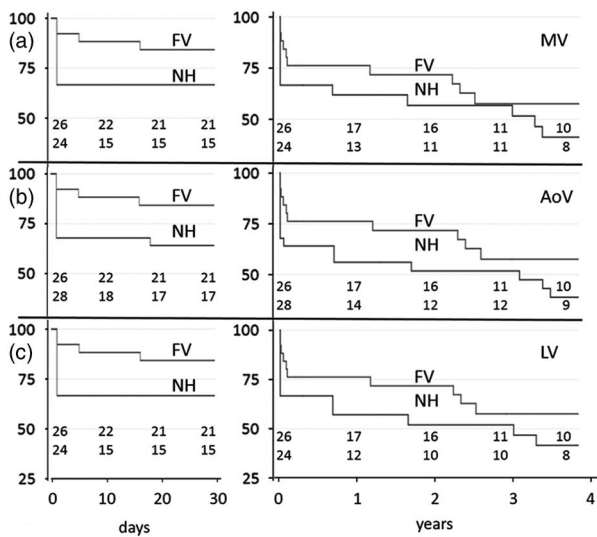


Figure 1. a-c: Kaplan Meier curves showing no significant difference in survival and BV for case control groups (FV = Fetal aortic valvuloplasty, NH = Natural history, matched for \pm one Z-score for MV, AoV, LV inlet Z-scores and retrograde aortic arch flow at 23 \pm 3 gestational weeks at 4 years after birth).

Introduction: Fetal aortic valvuloplasty (FV) has been proposed as an effective therapy to prevent progression from aortic stenosis to hypoplastic left heart syndrome (HLHS). Reversal of aortic arch flow in second trimester is thought predictive of HLHS without FV. We hypothesized that reversed arch flow does not predict HLHS.

Methods: In a retrospective multicenter and multinational study (2005-2012) 214 fetuses with aortic stenosis were enrolled into a hybrid case-control and repeated samples cohort. Liveborn surgical candidates undergoing FV or without FV (natural history, NH) were matched for between \pm one Z-score for mitral valve (MV) and aortic valve (AoV) diameters, left ventricular inlet length (LV) and retrograde arch flow at 23 \pm 3 gestational weeks, producing best match cohorts for each. Outcome measures were survival with biventricular circulation (BV) at four years. We analysed outcomes using Cox proportional hazards regressions and Kaplan Meier curves. **Results:** FV was performed in 67/214, technically successful in 59 (88.0%). FV-related deaths occurred in 7/67 (10.4%). Overall 151/214 (71%) were live-born, including unknown outcomes in 5 and no surgery in 16. Kaplan Meier curves show no significant difference in survival and in BV for case-control groups matched for retrograde arch flow and mitral valve, aortic valve or left ventricle inlet length Z-scores at 4 years after birth (Figure 1 a-c). **Conclusions:** The results from this multicenter study, matching for similar morphology and reversed arch flow in second trimester, showed that those with and without FV had similar BV outcomes and failed to show that reversed arch flow predicts HLHS.

YIA-3

Analysis of the Mechanisms of Intravenous Immunoglobulin-Resistant Kawasaki Disease Using iPSC Cell Technology

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Introduction: Although the treatment of intravenous immunoglobulin (IVIG) significantly resolves inflammation, 10-20% of Kawasaki disease (KD) patients have persistent or recurrent fever after the administration of IVIG, and IVIG-resistant patients have a particularly high risk of developing coronary artery abnormalities. The mechanisms of IVIG-resistant KD have been analyzed using the patients' leukocyte samples. However, vascular endothelial cells (ECs), closely related to the vasculitis of KD, have not been examined in the previous reports. We propose a hypothesis that ECs are mainly involved in the etiology of IVIG-resistance.

Methods: The purpose of this study is to establish new in vitro disease models of vasculitis using induced pluripotent stem cell (iPSC) technology, and clarify the mechanisms of IVIG-resistance in KD. Dermal fibroblasts or T cells from 2 IVIG-resistant and 2 IVIG-responsive KD patients were reprogrammed by episomal vectors encoding Oct3/4, Sox2, Klf4, L-Myc, LIN28, and p53 shRNA. The iPSC lines were then differentiated into ECs and smooth muscle cells (SMCs) by using a previously-reported differentiation method, and the EC and SMC samples were subjected to the microarray analyses.

Results: The KD patient-derived iPSCs could be differentiated into ECs and SMCs. The gene expression profiles were compared between iPSC-derived ECs (iPS-ECs) generated from IVIG-resistant and IVIG-responsive KD patients, and between iPSC-derived SMCs (iPS-SMCs) generated from two group patients. We found the expression of chemokine X, which stimulates migration of monocytes and T-lymphocytes through its receptors, was significantly up-regulated both in iPS-ECs and in iPS-SMCs from IVIG-resistant KD patients compared with those from IVIG-responsive patients. The Principle Component Analysis (PCA) was performed, but the gene expression levels showed no significant differences between the groups. The Gene Set Enrichment Analysis (GSEA) revealed that the gene sets related to IL-6, NRAS (a member of the RAS oncogene family) and breast cancer were up-regulated in iPS-ECs from IVIG-resistant KD patients.

Conclusions: Taking into account that the concentration of IL-6 has been reported to be elevated in acute phase of IVIG-resistant KD, our results suggest that the up-regulation of IL-6 related genes in ECs might be involved in the pathogenesis of IVIG-resistant KD.

YIA-4

Improvement of haemodynamic flow abnormalities after aortic valve replacement in bicuspid aortic valve disease

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Introduction: Bicuspid aortic valve disease (BAV) is associated with dilatation of the proximal aorta and abnormal flow patterns, particularly increased helical flow and changes in the aortic wall shear stress. The aortic dilation may be slowed by aortic valve replacement via normalisation of flow patterns. We assess the effect of different types of aortic valve replacement (AVR) on aortic flow patterns.

Methods: We prospectively enrolled 69 participants: 23 BAV patients with prior AVR (10 mechanical, 6 bioprosthetic, 7 Ross procedure), 23 BAV patients with a native aortic valve and 23 healthy volunteers. All underwent 4D flow cardiovascular magnetic resonance.

Results: The majority of patients with mechanical AVR or Ross showed a normalised flow pattern (70% and 57% respectively) with near normal rotational flow values (7.4 ± 3.9 and 11.0 ± 12.0 mm²/s respectively; normal range -5 to $+11$ mm²/s); and reduced in-plane wall shear stress compared to native BAV (0.13 ± 0.18 N/m² for mechanical AVR vs. 0.37 ± 0.26 N/m² for native BAV, $p < 0.05$). In contrast, all subjects with bioprosthetic AVR showed abnormal flow patterns (mainly marked right-handed helical flow), with similar

rotational flow values to native BAV (25.3 ± 15.0 mm²/s and 20.1 ± 11.0 mm²/s respectively, $p > 0.05$) and similar wall shear stress pattern. Data before and after AVR ($n = 13$) supported these findings: mechanical AVR showed a significant reduction in rotational flow (29.3 ± 15.1 to 7.9 ± 4.2 mm²/s, $p < 0.05$) and in-plane wall shear stress (0.45 ± 0.19 to 0.20 ± 0.12 N/m², $p < 0.05$), whereas these remained unchanged in the bioprosthetic AVR group.

Conclusion: Abnormal flow patterns in BAV are significantly reduced after mechanical AVR or Ross procedure, but remain similar after bioprosthetic AVR. This is the first insight indicating that type of valve replacement may influence post-operative flow patterns, and could have important implications for future aortic growth.

YIA-5

Team Approaching for Pregnancy and Delivery in patients with Severe Coronary Arterial Lesions After Kawasaki Disease

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Objective: Management for pregnant female patients of Kawasaki disease (KD) with severe coronary arterial lesions (CAL) is recently closed up. Pregnancy is thought to have two problems to maintain anti-platelet effect. One is acceleration of coagulation, and the other is increased heparin clearance. Those problems are more serious for KD patients with CAL. The feasibility of continuous intravenous heparin administration (CIVH) for them is necessary to be evaluated.

Method: We experienced 6 deliveries in 4 patients who switched oral aspirin to CIVH of 5-8 unit/kg/hour before delivery. Conferences with cardiologists, obstetricians and pediatricians including neonatologists were done for each case before and/or after their admission. Those medical records were summarized retrospectively.

Results: Age of expecting female KD patients was 28.5 years in average. All patients had left CAL and two had right CAL also. Three of them had ischemic finding by scintigraphy and coronary CT angiogram before pregnancy. No patient was prescribed warfarin. Three of them were prescribed aspirin, but one of them and the other patient were lost to follow-up of our university hospital. They came back to follow-up at our hospital after they were aware of pregnancy. They admitted to our hospital 7 to 14 days before expected date, and spontaneous, vaginal delivery was indicated in 4 deliveries. In two cases decided to carry out Caesarian section at 38th gestational week. All patients discontinued aspirin after admission and at latest 7 days before delivery and started CIVH and restarted aspirin from 24 to 48 hours after deliver. In all cases, target of activated partial thromboplastin time (APTT) was controlled over 50 seconds. All 6 deliveries completed and newborns were all mature with no hemorrhagic complication. No KD patients developed any hemorrhagic or thrombotic complication.

Discussion: APTT control longer than 50 seconds by CIVH is enough to warrant prevention of thrombotic event. Safe upper limit of APTT control without bleeding complication and the pros and cons of pregnancy in KD patients treated with warfarin are still two assignments. Team approaching should be achieved for appropriate management of the expecting patients with CAL after KD.

YIA-6

Outcomes after Protein-Losing Enteropathy in Univentricular Hearts: a Multicenter Study

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Background: Protein-losing enteropathy (PLE) is a rare but severe complication after Fontan surgery in patients with univentricular hearts (UVH). Outcomes are compromised since mortality is high and treatment efficiency appears limited.

Methods: This was a retrospective observational study carried out in sixteen Pediatric Cardiology Centres in France. All UVH patients diagnosed with PLE after Fontan type surgery such as atrio-pulmonary/atrioventricular anastomosis or intracardiac/extracardiac total cavopulmonary connection were included. The study period went from 1988 until 2014.

Results: PLE was diagnosed in thirty-five patients at a median age of 9.7 years. Median delay after Fontan type surgery was 3.6 years. At diagnosis, cardiac catheterization revealed a hemodynamic dysfunction of the Fontan circulation in 63% ($n = 22/35$). Treatment modalities included medical treatment alone (e.g. oral/topical steroids, Calciparine®, sildenafil) in 46% ($n = 16/25$) or combined treatment options (interventional or/and surgical therapy associated to medical treatment) in the other 54% ($n = 19/35$). Treatment was considered to be efficient in case of normalization of albumin level (>30 g/l). Medical treatment alone led to complete recovery in 13% ($n = 2/16$), to transient improvement in 31% ($n = 5/16$) and to no improvement in 56% of patients ($n = 9/16$). No patient treated medically died but two (13%) were finally transplanted (one recovery, one transient improvement). Combined treatment modalities led to 21% recoveries ($n = 4/19$) and to 37% transient improvements ($n = 7/19$). No improvement was noted in 42% of patients ($n = 8/19$). 21% ($n = 4/19$) in this treatment group died. 16% ($n = 3/19$) finally underwent heart transplantation but all three subsequently died (one early death, two late deaths). Five and 10 years survival of the whole cohort were 89.7% (CI95%: $\pm 11.3\%$) and 74.9% (CI95%: $\pm 21.3\%$) respectively. Median follow-up was 4.5 years [0.5-21.7]. At last visit, 75% of survivors ($n = 21/28$) were in functional NYHA class I/II. However, 75% still had patent hypoalbuminemia and 61% received specific medical therapy for PLE.

Conclusions: Despite decreased mortality, PLE remains a significant burden after Fontan surgery since complete recovery is rare and treatment modalities remain unsatisfying. Heart transplantation is an ultimate therapeutic option but carries a high risk. Further studies are needed to develop innovative treatment strategies and improve outcomes.

OC-1

Stenting of the right ventricular outflow compared to modified Blalock-Taussig shunt palliation: Mortality, Morbidity and Re-intervention rate

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Introduction: Stenting of the RVOT is an alternative to surgical creation of a modified Blalock-Taussig shunt (BTS) in selected patients with Fallot type lesions to augment pulmonary blood flow.

Objectives: To compare mortality, procedure related morbidity and re-intervention rate after stenting of RVOT versus BTS for palliation prior to corrective surgery in patients with pulmonary atresia, VSD and confluent pulmonary arteries (PAVSD) or Tetralogy of Fallot-type lesions.

Methods: Retrospective case review study to evaluate 133 patients (86male) with PAVSD or Fallot-type lesions who underwent palliation with either a BTS (n=73) or RVOT stent (n=60) to augment pulmonary blood flow over a 10 year period. Overall mortality until corrective surgery, 30-day mortality, procedure related morbidity (defined as admission rate to PICU, complications during palliation period, ward length of stay) and re-intervention rates were assessed and compared.

Results: There were more patients with non-cardiac co-morbidities (prematurity, IUGR, gut malformations, cerebral abnormalities) in the RVOT stent group (p=0.362). Median days of palliation was shortest in the RVOT stent group 224 (110 - 323) days (p<0.001) based on clinical practice and good PA growth. Despite a lower 30-day mortality in the RVOT stent group 1/60 (1.7%) compared to 5/73 (6.8%) in the BTS group (p=0.221) the overall mortality was similar in both groups with 8.9% in the RVOT stent group and 8.2% in the BTS group (p=0.564). There were 2 non-cardiac deaths in the RVOT stent group. Procedure related admission rate to PICU was lower in the RVOT stent group (22% versus 100%, p<0.001). There were 8 NEC in the BTS group versus 2 NEC in the RVOT stent group (p=0.113). There were a total of 9 BTS thrombosis (12.3%) versus no complete occlusion of RVOT stents (p<0.001). Ward length of stay was shorter for the RVOT stent group (p<0.001). There was a higher catheter re-intervention rate in the RVOT stent group.

Conclusion: RVOT stenting in Fallot type lesions promotes low 30-day mortality (1.7%), lower admission rate to PICU post procedure and shorter ward length of stay compared to BTS palliation to augment pulmonary blood flow.

OC-2

Ablation of idiopathic ventricular tachycardias arising from the sinuses of valsalva in pediatric patients: tips and tricks

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Objective: To optimize the approaches for catheter ablation of idiopathic ventricular arrhythmias arising from sinuses of Valsalva.

Methods: In 2003–2014, 555 children aged 6–17 yo received invasive treatment of VA in one hospital. 96 (17.3%) of them with ventricular premature beats (VPB) or/and ventricular tachycardia (VT) arisen from sinuses of Valsalva had been included into the study. Only conventional method of mapping was used. The catheter position during the applications near the ostium of the coronary arteries was precisely controlled by continuous fluoroscopy and coronarography. The follow-up period was 3 months. Efficacy, complications, time of procedure and effective dose were examined.

Results: According to results of endocardial mapping pts were divided in two groups depending on the risk of potential complications: group I – “low risk zones” with the distance between the origin of VPB/VT and the ostium of the coronary arteries was more than 12 mm; group II – “high risk zones” with distance <12 mm between the origin of VPB/VT and coronary ostium or near to His bundle projection. Transaortic access was chosen based on evaluation of QRS morphology. 86 (90%) pts had been selected into the group I and 10 (10%) pts – into the group II. In group I radiofrequency catheter ablation was performed in all cases. In group II cryoablation was used for safety reasons. Only in 7 pts

cryoablation was performed, in 3 pts earliest activation was found in the ostium of left main coronary artery. In these cases epicardial localization VPB/VT substrate had been recognized and catheter ablation was not performed. No significant difference in time of procedure and effective dose between the groups was found. The efficacy of all procedures achieved up to 100%. There were no complications in both groups.

Conclusions: Transaortic access should be selected without previous mapping of right ventricular and based on evaluation of QRS morphology. Electroanatomic method of mapping is not necessary for these pts. Radiofrequency catheter ablation is appropriate method for “low risk zone”. In “high risk zone” cryoablation should be selected. Continuous coronarography should be performed during ablation for prevention complications in all cases.

OC-3

Mutations in pulmonary arterial hypertension genes in children

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Background: Prevalence of germline mutations in the genes associated with pulmonary arterial hypertension (PAH) in childhood-onset PAH is scarcely known.

Objectives: To determine the prevalence of six known genes for heritable PAH gene (BMPR2, ALK-1, ENG, TBX4, KCNQ3 and EIF2AK4) in children with PAH and to describe the clinical characteristics of children harboring mutations.

Methods: Over a period of six years, 71 index cases were included for genetic analysis of PAH genes: idiopathic PAH (n=36); familial PAH defined as one first degree relative with PAH (either one parent or one sibling; 8 families–10 patients– all children were considered index cases as none mutation had been previously identified in the family before inclusion of the case into the study); pulmonary veno-occlusive disease (PVOD; n=2); type 3 PAH associated with congenital heart disease (coincidental CHD) APAH-CHD (n=13); and type 4 (postoperative) APAH-CHD (n=10). All patients with already known or chromosomal anomaly or identified syndrome were excluded from the study.

Results: We found no mutations in children with type 3 and type 4 APAH-CHD.

Eight mutations were found in 36 children with iPAH (22%): three in BMPR2, three in ALK1, and two in TBX4. No mutations were identified in ENG, or KCNK3. Four mutations were found in the eight familial PAH families (50%): two in BMPR2, one in ALK1, and one in TBX4. In these four familial forms, only one sibling of an index case with a TBX4 mutation was alive with PAH, and had the same mutation. In the three remaining families, the first-degree relatives who had PAH were all dead at inclusion of the index case into our study with no material available for genetic testing. Finally, we identified mutation in EIF2AK4 in the two patients with clinical, hemodynamic and CT features of PVOD. Outcome of children with mutation and without were similar.

Conclusion: Mutations in the known genes for PAH is either exceptional or absent in children with unusual APAH-CHD. Prevalence of PAH genes mutations is less frequent than in the adult forms of iPAH and familial PAH. Genetic status does not predict outcome in our series.

OC-4

Impact of precision prenatal diagnostic of congenital heart diseases on perinatal and long-term management

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Objective: To evaluate the impact of the precision of prenatal diagnosis of congenital heart diseases (CHD) on perinatal and long-term management.

Methods: Over a period of 10 years, 1258 neonates with a prenatally diagnosed CHD and 189 fetal autopsies after termination of pregnancy were included. Changes in CHD diagnosis were classified as totally different, similar but leading to changes in neonatal management, and similar without changes on initial management. The impact on long-term outcome was considered negative if the final diagnosis was a more complex CHD precluding the planned biventricular repair, or if additional surgical interventions were needed, or if the complexity level of the Aristotle score was increased. Conversely, the impact on outcome was considered positive if biventricular repair was possible while not planned prenatally, or if the number of surgical interventions was reduced, or if the complexity level of the Aristotle score was lower.

Results: In live births, the post-natal diagnosis was imprecise in 30.2% of cases (n = 380/1258): completely different in 2.9%, led to changes in initial management in 8%, and did not affect initial management in 19.3%. Imprecision in the prenatal diagnosis had a negative impact on long-term outcome in 62/1258 (4.9%), and a positive impact in 52/1258 (4.1%). In 91% of the cases, no consequence on long-term outcome was observed.

In the fetal autopsy group (mean term 26 weeks, range 10–38), the diagnosis was imprecise in 54.5% of the cases (n = 103/189): completely different in 8.5%, could have led to changes in postnatal management in 14.3%, and with minor differences that would not have led to changes in management in 31.7%. In two cases, correcting the CHD diagnosis could have led to continue pregnancy.

In both groups, the most frequent differences were anomalies of the outflow tract anatomy (43%), and the systemic or pulmonary veins (25%).

Conclusion: Imprecision of prenatal diagnosis of CHD changes early management in 11% of the cases, and impacts long-term outcome in 9% of the cases. Improvement of CHD diagnosis for anatomy of the outflow tract and main veins should help to reduce impact on postnatal management and outcome.

O1-1

Outcome in Paediatric Pulmonary Arterial Hypertension: a Systematic Review and Meta-Analysis

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Introduction: Current data on outcome and prognostic factors in children with pulmonary arterial hypertension (PAH) are derived from small patient series and contradictory findings have been reported. For the development of treatment strategies and guidelines, there is a high unmet need for an evidence-based recapitulation of prognostic factors. Therefore, the aim of this study was to combine the currently available data on prognostic factors in paediatric PAH by a systematic review of the literature and to summarize the prognostic value of currently reported prognostic factors using meta-analysis.

Methods: Medline, EMBASE and The Cochrane Library were searched at April 1st 2014 to identify original studies that described predictors of mortality or lung-transplantation exclusively in children with PAH. Titles and abstracts followed by full-text articles were screened by two independent reviewers. Eligible studies were required to report at least (1) data on mortality in paediatric PAH and (2) variables studied in relation to mortality. Hazard ratio's (HR) and 95% confidence intervals (CI) were extracted or calculated from the papers. For variables studied in at least three non-overlapping cohorts, a combined HR was calculated using random-effects meta-analysis. Heterogeneity was assessed using Cochran's Q-test and the I²-quantity.

Results: Of 1053 primarily identified articles, 25 were eligible for inclusion. In these paediatric studies, a total of 40 candidate prognostic factors was reported. 10 of these 40 candidate prognostic factors were studied in at least 3 cohorts, allowing for 10 separate meta-analyses involving up to 585 children. WHO-functional-class (HR = 2.67, CI = 1.49–4.80, p = 0.001), (N-terminal-pro-) B-type natriuretic peptide (HR = 3.24, CI = 1.76–6.02, p < 0.001), mean right atrial pressure (HR = 1.12, CI = 1.05–1.20, p = 0.001), cardiac index (HR = 0.66, CI = 0.52–0.84, p = 0.001), indexed pulmonary vascular resistance (HR = 1.32, CI = 1.17–1.48, p < 0.001) and acute vasodilator response (HR = 0.27, CI = 0.14–0.54, p < 0.001) were identified as significant prognostic factors in the absence of evidence for heterogeneity (Q-test p > 0.10 and I² ≤ 50% for all identified significant prognostic factors).

Conclusions: This systematic review combined with separate meta-analyses identifies 6 clinical variables that are consistently reported prognostic factors for outcome in paediatric PAH. These variables are useful clinical tools to assess prognosis and should be incorporated in treatment strategies and guidelines for the management of paediatric PAH.

O1-2

RSV hospitalization in winter and summer season stratified by CHD subgroups; a national study

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Introduction: Respiratory tract infections are common and serious CHD is a risk factor for severe illness and hospitalization. Prophylactic treatment with palivizumab, recommended during winter, is known to reduce risks of severe RSV disease. The aim of this study was to compare the calculated relative risk for hospitalization due to RSV infection in winter and summer season in different types of CHD.

Methods: All children born in Sweden and less than two years old, in 2006–2011, were included in the study. Winter was defined as first of November to 30th of April each year. Rates of children with CHD (divided into eight subgroups) and hospitalization caused by RSV were retrieved from the National Inpatient registry. The relative risk of hospitalization due to RSV was calculated comparing each subgroup to other types of CHD and to healthy children.

Results: The relative risk of hospitalization due to RSV infection was increased for all CHD subgroups. For children with univentricular heart defects, systemic-pulmonary shunt defects, other complex CHD and Tetralogy of Fallot, the relative risk of RSV

hospitalization was significantly higher during summer season compared to winter.

Conclusions: National guidelines of prophylactic treatment are recommended only during winter. The cost-benefit of such treatment throughout the entire year is low. We argue that health personnel must be aware of the increased risks of RSV infection for the sickest CHD-children throughout the whole year.

O1-3

Cardiopulmonary Exercise Testing Predicts Outcome in Paediatric Pulmonary Hypertension

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Introduction: Pulmonary hypertension (PH) is associated with exercise intolerance and poor survival. Exercise limitation, measured by 6-minute walking distance tests (6MWD) and cardiopulmonary exercise tests (CPET) has been robustly shown to correlate with disease severity and outcome in adult PH populations. Accordingly these measures have proved invaluable in guiding therapeutic decisions and as outcome measures in clinical trials. Data for their validity in children with PH are lacking. We aimed to analyse the relationship between CPET parameters and outcome in paediatric PH.

Method: A single center retrospective study of PH patients less than 18 years old that had undergone CPET between the dates April 2004 and April 2014. Exclusion from CPET was based upon height <120 cm, or resting symptoms too severe for formal testing. Patients with cardiomyopathy or univentricular physiology were also excluded. All CPET and 6MWD took place within the same 24-hour period. The need for ethical approval was waived.

Results: 93 patients underwent exercise testing. The median (IQR) age was 14.2 years (10.9-15.9), height 152.0 cm (135.0-162.0) and weight 44.0 kg (30.0-53.0). 46 (49.5%) had PH associated with congenital heart disease whilst 37 (39.8%) had idiopathic PH. WHO functional class was 1, 2, 3 and 4 in 16, 38, 38 and 1 patients respectively. Patients experienced significant exercise limitation; mean (SD) percentage predicted 6MWD (6MWD%) 57.7% (15.6%), peak VO₂ (PVO₂) 54.9% (20.7%). Both 6MWD% and PVO₂ correlated with WHO (r^2 0.27 and 0.25 respectively, $p < 0.001$) with a weaker correlation for VE/VCO₂slope (r^2 0.17, $p < 0.001$). Over a median follow-up of 3.53 years (1.62-5.63), 16 patients died and 2 were transplanted. 6MWD% did not predict outcome (HR 0.67, $p = 0.07$) however, the following CPET measures were prognostic; PVO₂, VE/VCO₂ slope; hazard ratio for each standard deviation increase of 0.52, 0.48 and 2.6 respectively.

Conclusion: In this large study we report for the first time the prognostic utility of cardiopulmonary exercise testing in children with pulmonary hypertension. Our findings support the use of CPET in paediatric PH.

O1-4

Re-interventions after the arterial switch operation

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Introduction: Surgical or catheter-based re-interventions after the arterial switch operation (ASO) for transposition of the great arteries (TGA) are reported in 16 to 37% of patients. The objective

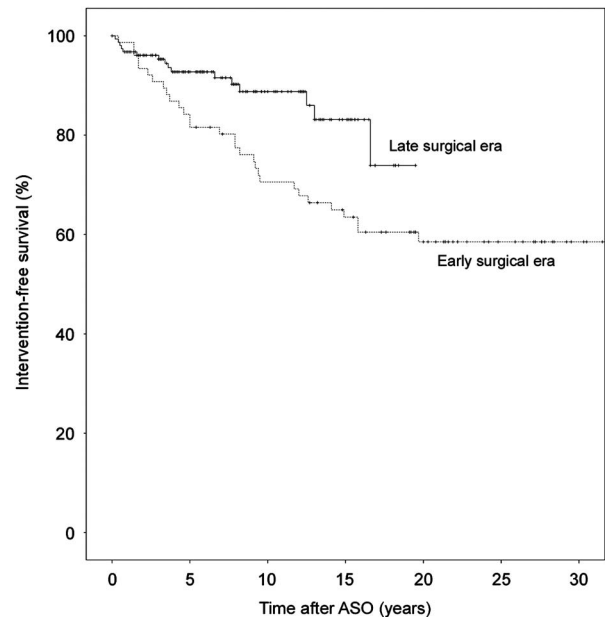


Figure. Kaplan-Meier comparing intervention-free survival between the late (<18 years after ASO) and early surgical era (≥ 18 years after ASO).

of is this study is to evaluate a possible relation between the surgical era and the need for future re-intervention.

Methods: A single institution retrospective cohort study was performed to assess re-intervention after ASO between 1978 and 2013. All patients with simple or complex TGA were included. Medical records were reviewed for re-interventions after initial discharge for ASO. Patients were subdivided in two groups; ≥ 18 years after ASO (early surgical era) and < 18 years after ASO (late surgical era). Event-free survival was estimated by Kaplan-Meier method and differences were analysed using log-rank test.

Results: Mean follow-up was 15.8 years (± 0.62) for 285 patients. Forty-eight re-interventions were performed, 27 (9.5%) surgical and 21 (7.4%) catheter-based interventions. The most frequent indication for re-intervention was pulmonary artery stenosis (N = 33), followed by neo-aortic regurgitation (N = 6), re-coarctation of the aorta (N = 6), left ventricular outflow tract obstruction (N = 2) and right ventricular outflow tract obstruction (N = 1). Overall intervention-free survival at 10, 20 and 30 years was 81.8%, 72.6% and 70.6% respectively. Intervention-free survival for patients of the early surgical era was significantly lower in Kaplan-Meier analysis ($p = 0.009$). This is mainly explained by a decrease in surgical relief of pulmonary stenosis over time. The incidence of catheter-based re-interventions did not increase over time. With Cox regression, younger patients also showed a statistically significant higher intervention-free survival, with hazard ratio = 1.05 per year ($p = 0.01$).

Conclusions: The risk of re-intervention after ASO is inversely related to the surgical era. This improvement is mainly explained by a decrease in surgical re-interventions for pulmonary artery stenosis.

O1-5

Aortic valve prolapse and aortic regurgitation in 2283 patients with isolated ventricular septal defect

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There are many reports concerning aortic valve prolapse and aortic regurgitation associated with subpulmonic ventricular septal

defect, but there are limited data about the natural history of aortic valve prolapse associated with other types of ventricular septal defect. Echocardiography has provided a reliable assessment of the presence and location of ventricular septal defect, aortic valve deformity, and degree of aortic regurgitation noninvasively, even in infancy and early childhood. This study evaluates aortic valve prolapse, onset and progression of aortic regurgitation, and ventricular septal defect location in 2283 patients with ventricular septal defect.

Materials and Methods: The study population consisted of 2283 patients with isolated ventricular septal defect who had been studied at our institution from 1988 to 2014 with transthoracic echocardiography. Ventricular septal defects were classified according to their location and relation to the tricuspid annulus and semilunar valves. Defect size was expressed in terms of the size of the aortic root. SPSS 17 was used for statistical analysis and statistical significance was inferred at $p < 0.05$.

Results: Aortic valve prolapse was detected in 173 (7.6%) of 2283 patients with ventricular septal defect by echocardiography (27 at initial echocardiographic examination and 146 at follow up). Of 173 patients with aortic valve prolapse, aortic regurgitation was detected in 103 (59.5%), resulting in an incidence of aortic regurgitation of 4.5% of ventricular septal defects. The percentage of aortic valve prolapse in muscular outlet ventricular septal defects (12.2%) and perimembranous ventricular septal defects (10.2%) was similar but associated aortic regurgitation was common in muscular outlet defects (12.2%) then in perimembranous defects (6.6%) ($p < 0.05$). One hundred twenty three patients were followed up after aortic valve prolapses appeared. Initially there was no aortic regurgitation in 63 of these patients, trivial aortic regurgitation in 22 and mild aortic regurgitation in 38.

Discussion: We recommend frequent echocardiographic evaluation (every 6 months) for detecting of appearance of aortic regurgitation in patients with perimembranous or muscular outlet ventricular septal defect after aortic valve prolapse develops.

O1-6

Clinical Features, Management and Outcome of Pre- and Postnatally Diagnosed Common arterial trunk: A Comprehensive Single Institution's Experience Since 1990

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Introduction: Common truncus arteriosus (CTA) is associated with significant morbidity and mortality although improvements in management have been made over time. The purpose of this study was to determine outcome, risks associated with mortality and the impact of prenatal detection on survival.

Methods: We performed a retrospective review of CTA cases diagnosed fetally and postnatally at our institution between 1990 and 2014.

Results: There were 192 CTA cases with 61 fetally diagnosed. Of the fetal cases there were 18 terminations of pregnancies, 2 intrauterine deaths and 4 live-births that were not actively managed. One patient diagnosed postnatally was not actively managed. This left 167 patients with intended postnatal care. Mixed truncal valve disease occurred in 41% of cases with moderate or severe regurgitation or stenosis seen in 23% and 18% of cases respectively. Other associations included aortic arch interruption (16%), right aortic arch (26%), pulmonary arterial interruption (16%) and DiGeorge syndrome (24% of tested cases). Including 17 (9%) infants that died prior to surgery, survival of the 167 actively managed patients was 74%, 62% and 56% at 1 month, 1 year and 10 years respectively. Reoperation rate at last follow up was 37%. Multivariable independent risk factors for mortality included interrupted aortic arch [HR 3.7 (2.1-6.6), $p < 0.001$], moderate or severe truncal valve stenosis [HR 3.3 (1.7-6.4), $p < 0.001$] or regurgitation [HR 2.6 (1.5-4.5), $p = 0.001$], hypoplastic or interrupted

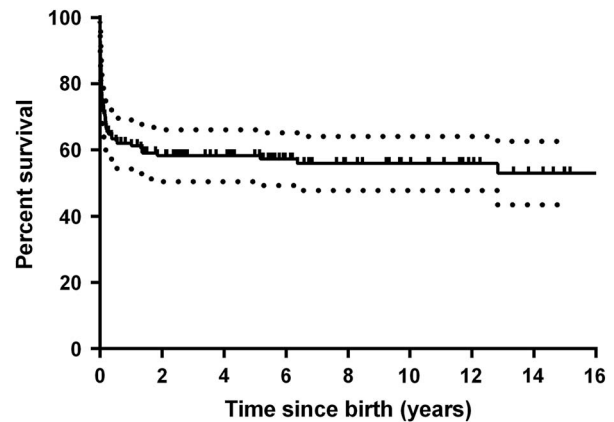


Figure.

pulmonary arteries [HR 2.1 (1.2-3.6), $p = 0.006$], prematurity [HR 1.2 (1.1-1.3), $p = 0.004$], lower preoperative weight [HR 1.6 (1.1-2.4), $p = 0.02$], earlier year of surgery [HR 1.08 (1.03-1.13), $p = 0.002$] and the need for life support preoperatively in the form of inotropes [HR 3.2 (1.6-6.3), $p < 0.001$] or mechanical ventilation [HR 2.1 (1.1-4), $p = 0.03$]. Fetal detection rates improved over time but a fetal diagnosis was not associated with improved survival rates.

Conclusions: The overall outcome of CTA during childhood is poor and comparable to the most severe forms of congenital heart disease. Moderate to severe truncal valve regurgitation or stenosis, interrupted aortic arch and pulmonary artery abnormalities are associated with a worse outcome.

O1-7

Does Prenatal Diagnosis of Transposition of the Great Arteries Improve Survival?

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Introduction: Current evidence suggests prenatal diagnosis of transposition of the great arteries (TGA) improves pre-operative clinical condition and long term outcome. In 2008 the UK National Institute of Clinical Excellence (NICE) published guidelines advising that ventricular outflow tracts should be assessed as part of routine antenatal assessment. This was consolidated in the 2010 Fetal Anomaly Screening Programme guidelines.

Our aim was to establish whether local improvement in prenatal diagnosis of TGA improved early survival rates.

Method: All patients undergoing the arterial switch procedure (ASO) for simple TGA between 2001 and 2014 were identified. Patients with septal defects were included but those with additional congenital heart disease were excluded.

A retrospective review of patient records was performed. Data was gathered regarding demographics, diagnosis, admission to the cardiac centre, hospital stay and long term follow up. The primary outcome was survival 30 days post ASO. A comparison was undertaken between neonates diagnosed postnatally and prenatally. Secondary outcomes included clinical status at time of admission to cardiac centre, timing of septostomy, length of stay and long term comorbidity.

Results: 228 patients with simple TGA +/- septal defects who had the arterial switch procedure were identified during the 14 year period. The rate of prenatal diagnosis has been consistently increasing from 11% prior to 2008 to greater than 60% in the last 2 years. 168 patients were diagnosed postnatally, 60 were diagnosed prenatally. The majority of prenatal diagnoses were made from 2008 onwards (49/60). Overall survival across both patient groups was 95% at 30 days. Survival was 97% in the postnatally diagnosed

group (163/168) versus 93.3% in the prenatally diagnosed group (56/60). Average length of stay was 1 day shorter in prenatally diagnosed neonates (19 versus 20 days).

Conclusions: Regionally our rate of prenatal diagnosis is improving. However this does not currently seem to be reflected in an increased rate of survival. A larger subgroup of prenatally diagnosed neonates would be required to establish significance. We anticipate that further improvement in both prenatal diagnosis and expectant management at time of delivery will begin to demonstrate the benefits of prenatal diagnosis and its impact on survival.

O2-1

How normal is a “normal” heart in fetuses with Down syndrome?

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Introduction: Congenital heart disease is present in 44–56% of fetuses with Down syndrome (DS). There are, however, signs that hearts in DS without apparent structural heart defects, also differ from the normal population. Sonic hedgehog signaling may be involved in the pathogenesis of AVSD in DS, although currently data is limited. We aimed to compare the atrioventricular septum and valves in 3 groups: DS without atrioventricular septal defect (‘DS no-AVSD’), DS with AVSD (‘DS AVSD’) and control hearts.

Methods: The ventricular septum, the atrioventricular- (white arrow) and interventricular (black arrow) –membranous septum and AV valves were examined and measured in histological sections of 15 ‘DS no-AVSD’, 8 ‘DS AVSD’ and 34 ‘control’ hearts (Gestational age 10–22 weeks) In addition, the ventricular septum length was measured on ultrasound images of fetal (6 ‘DS AVSD’, 9 ‘controls’) and infant (10 ‘DS no-AVSD’, 10 ‘DS AVSD’, 10 ‘controls’) hearts.

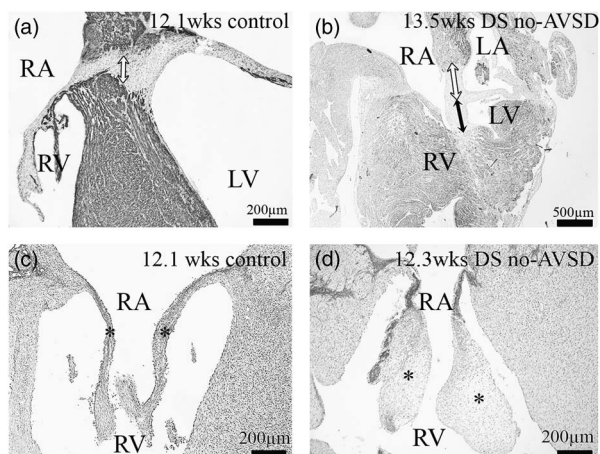


Figure. RA = right atrium, RV = right ventricle, LA = left atrium, LV, left ventricle, white arrow atrioventricular membranous septum, black arrow interventricular membranous septum, * = atrioventricular valves.

Results: The membranous septum volume was 3 times larger in ‘DS no-AVSD’ fetuses compared to control foetuses (panel A–B) and valve dysplasia (panel D asterisk) was frequently (64%) observed. In the DS fetuses with a (complete) AVSD, fibrous tissue was observed at the top of the ventricular septum in 3 cases. The ventricular septum was shorter in patients with DS both with (0.7 times the length of controls, $p=0.001$) and without AVSD (0.78 times the sizes of the control, $p<0.001$). In contrast to controls, in ‘DS no-AVSD’ fetuses, clear expression of Gli1, an effector of sonic hedgehog signaling, was present in the dysplastic plump AV valves and membranous septum in 2 out of 4 cases.

Conclusions: DS no-AVSD hearts are not normal as they have a larger membranous septum, shorter ventricular septum and dysplasia of the AV valves as compared to control hearts. Findings indicate that a careful cardiac follow-up may be warranted in these patients.

O2-2

Inner architecture of the right ventricle: the role of the tricuspid valve

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Background: A major anatomic characteristic of the right ventricle (RV), in addition to the coarseness of the apical trabeculations, is the presence of muscular bands arranged in a semicircular fashion. Recent publications show that, while parietal band and subpulmonary conus represent the embryologic ventriculo-infundibular fold (VIF), the septal band (septomarginal trabeculation), moderator band and anterior papillary muscle of the tricuspid valve (APM) could derive from the muscular tricuspid primordium and from the inlet portion of the heart.

Objectives: To analyze the anatomy of the RV in heart specimens with tricuspid atresia (TA), in order to confirm the hypothesis of different embryologic origins for the muscular bands of the RV.

Material and Methods: We reviewed 54 hearts with TA from the anatomic collection of the French Center of Reference for Complex Congenital Heart Defects: 32 postnatal and 22 fetal hearts. The presence of a VIF, septal band, moderator band was assessed as well as the position of great vessels, the type of TA, muscular (musTA) or membranous (mbTA), the type and patency of the ventricular septal defect (VSD), and the associated lesions.

Results: Forty specimens had ventriculo-arterial concordance, 14 had D-transposition. There were 50 musTA (including 6 without any RV cavity), and 4 mbTA. Among the 48 specimens with a RV cavity, all had a well-developed VIF. A rudimentary septal band (with demonstrable limbs in only 3) was present in 7/44 musTA vs 3/4 mbTA ($p<0.04$), rudimentary moderator band and APM in 2/44 musTA vs 2/4 mbTA ($p<0.03$). A patent VSD was found in 40 hearts, muscular in 90% of musTA, outlet type in 75% of mbTA ($p=0.03$).

Conclusion: Septal band and moderator band were absent in the vast majority of hearts with tricuspid atresia, particularly in musTA, while parietal band and subpulmonary conus are always present. These anatomic findings confirm the hypothesis of a dual embryologic origin for the muscular bands of the RV: the VIF is reminiscent of the inner curvature of the heart, while the septal band, moderator band and APM develop later from the muscular tricuspid primordium, itself developed from the posterior part of the primary fold.

O2-3**Impact of 1st trimester anomaly scan on un-natural history of CHD**

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Prenatal prevalence of congenital heart defects (CHD) is higher in comparison to postnatal prevalence. True incidence is however unknown due to frequent intrauterine demise in early post-conceptual stages in fetuses with severe CHD and associated co-morbidity.

Objective: Analysis of spectrum of CHD, severity of associated abnormalities and outcome of fetuses diagnosed with CHD in first and second trimester screening (1TS and 2TS respectively).

Methods: Total of 510 fetuses diagnosed with CHD (127 in 1TS, 383 in 2TS) undergone ultrasound cardiac and anomaly scan and genetic assessment including karyotype analysis.

Results: Out of 127 fetuses diagnosed with CHD within 1TS (11 + 0 to 13 + 6 weeks of gestation) chromosomal and/or extracardiac anomalies were found in 85(67%) fetuses and CHD with univentricular circulation (UV) in 54(43%). Seven (6%) fetuses died in utero (IUD), pregnancy was terminated (TOP) in 108(85%) and only 12(9%) were born (11 of those with biventricular CHD).

Similarly, out of 383 fetuses in 2TS (14 + 0 to 28 + 0 weeks of gestation) chromosomal and/or extracardiac abnormalities were found in 117(31%) fetuses and CHD with UV circulation in 50(13%). Four (1%) fetuses died in utero, TOP was in 115(30%) and 264(69%) were born (258 with biventricular outcome and 227 without associated co-morbidity).

There were significantly less chromosomal and/or extracardiac abnormalities ($p < 0.0001$), CHD with UV outcome ($p < 0.0001$), IUD ($p = 0.0027$) and TOP ($p < 0.0001$) in the group of CHD diagnosed within 2TS.

The frequency of atrioventricular septal defect ($p = 0.0139$), hypoplastic left heart ($p < 0.0001$), pulmonary atresia ($p < 0.0010$) and tricuspid atresia ($p = 0.0097$) was significantly higher in 1TS, while detection of transposition of great arteries was higher ($p = 0.0033$) in 2TS.

Conclusion: We confirmed that 1TS screening has significant impact on outcome of pregnancies with CHD as more severe forms of CHD and higher co-morbidity result in increase of TOP. In 2TS transposition of great arteries and less severe forms of CHD are more likely to be diagnosed. These fetuses have better postnatal outcome due to more frequent biventricular circulation and less associated co-morbidities. In severe CHD with associated co-morbidity diagnosed in 2TS, families are more keen to continue with pregnancy as opposed to those diagnosed with similar lesions at 1TS.

O2-4**Significance of Social Deprivation and Modifiable Risk Factors in the Aetiology of Congenital Heart Defects**

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Introduction: The incidence of congenital heart disease (CHD) in Wales is two to three times the UK national average

(Wales 10/1000 vs UK 4/1000). The reasons for this increased incidence have not been ascertained. Therefore, we retrospectively studied the frequency and importance of social deprivation and other modifiable risk factors among the patients referred for a fetal cardiac scan to our institution over the past 12 years.

Methods: All patients from South Wales who had a fetal cardiac anomaly scan between 2001 and 2013 were included. The demographics, social deprivation indexes, maternal, familial and fetal exposures to potential risk factors and their correlation with ultrasound findings were evaluated. The results obtained from those with a diagnosis of CHD were compared to a control group with normal scans.

Results: 2701 fetuses had cardiac scans between 2001 and 2013. 751 (27.8%) of these were abnormal. A strong correlation between CHD and social deprivation was identified, with CHD being twice more common in those patients from the areas with a higher level of social deprivation. However, there was no significant difference in smoking rates, maternal medication, alcohol consumption and maternal age between the two groups. Genetic abnormalities were 8-fold more common in those with a fetal diagnosis of CHD compared with the control group of normal scans (12.4% versus 1.6% respectively, OR 9.0 [CI 4.3-19.1] $p < 0.0001$). There was a three-fold increase in the number of patients with insulin-dependent diabetes between those with abnormal and those with normal scans (3.6% versus 1.2% respectively, OR 3.08 [1.21, 7.81], $p = 0.018$).

Conclusions: Our study suggests that social deprivation have a strong association with CHD and common risk factors such as maternal IDDM and genetic abnormalities. These findings highlight the need for more comprehensive prospective data collection to determine the true nature and extend of modifiable risk factors in the aetiology of congenital heart disease.

O2-5**Pulsed Wave Tissue Doppler Imaging in Fetuses with Aortic Stenosis and Evolving Hypoplastic Left Heart Syndrome before and after Fetal Aortic Valvuloplasty**

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Background: Fetal aortic valvuloplasty is technically feasible and can improve filling and reduce afterload of the left ventricle in critical aortic stenosis. In the present study we used tissue Doppler imaging (TDI) to evaluate changes in ventricular function before and after prenatal aortic valvuloplasty.

Methods and Results: Between October 2008 and December 2012, cardiac function was assessed by TDI in 23 fetuses with critical aortic stenosis that underwent fetal valvuloplasty. Mean \pm SD gestational age at intervention was 27.5 ± 3.1 . Of the 23 fetuses, 14 underwent successful postnatal biventricular repair. All fetuses were examined with B-mode, M-mode, conventional Doppler and tissue Doppler imaging before and after intervention.

Before intervention the z-scores of all TDI-derived parameters (except E'/A') and MAPSE were severely abnormal. Post-intervention TDI-E', and TDI-S' but not TDI-A' were significantly higher than pre-intervention. While the E/A ratio remained unchanged, the ratios E'/A' and E/E' changed significantly. Values of MAPSE improved significantly as did LV MPI, but still remained abnormal. Additionally right ventricular A', S' and MPI improved significantly. Post-Intervention z-scores

were correlated with postnatal outcome (i.e. biventricular vs. single-ventricle): a higher left ventricular E' velocity ($r=0.605$, 95%BCa CI [0.333,0.784] $p=0.003$), a higher left ventricular E'/A' ratio ($r=0.492$, 95%BCa CI [0.094,0.889], $p=0.020$), a lower M-MPI ($r=-0.534$, 95%BCa CI [-0.799, -0.174], $p=0.011$) and a lower T-MPI ($r=-0.438$, 95%BCa CI [-0.715,-0.061], $p=0.042$) were significantly correlated with postnatal biventricular outcome. Left ventricular E/E' demonstrated a clear trend ($r=-0.379$, 95%BCa CI [-0.604,-0.048], $p=0.082$).

Conclusion: In critical aortic stenosis with evolving HLHS intrauterine aortic valvuloplasty led to improvement of TDI derived parameters of the LV but also of the RV. Significant improvement of these parameters was seen only in fetuses with later biventricular outcome. Some parameters continued to improve until birth indicating remodeling of the LV. This data supports the concept, that TDI in addition to currently used techniques is able to contribute to a proper patient selection and to the assessment of technical success and is of prognostic value. Therefore it should be part of the initial evaluation as well as of the follow-up examinations.

O2-6

Fetal pulmonary valvuloplasty – how sufficient it is?

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Objective: Evaluation of fetuses diagnosed in our unit with pulmonary atresia or critical stenosis and intact ventricular septum (PA/PS&IVS) between 1995–2010 (group1) showed poor outcome. Basing on this results prospective study was undertaken to establish indications and results of fetal pulmonary valvuloplasty (FPV) between 2011–2014 (group2).

Method: Review the data of 59 fetuses with PA/PS&IVS. Group1 (28fetuses) was presented previously. Data and outcome of group2 (31fetuses) was analyzed. 5 underwent FPV.

Results: The mean age of diagnosis in group1 was 27 ± 6 weeks, for group2 25 ± 6 weeks. Previous evaluations of group1 showed survival rate of 46% and no biventricular (BV) repair. Out of 59 fetuses, 25 were diagnosed before 24 weeks, 6 had abnormal karyotype. 5 were terminated, just one with isolated heart defect. In 5 fetuses who had FPV, RV outflow tract was patent and TV annulus was just slightly smaller, but RV major diameter was decreased. Severe TR with high velocity was in 4, in one it was absent and sinusoids were suspected. All FPV were successful. In one fetus PA closed completely just before delivery, so radio-frequency was performed in a neonate. In all 5 neonates TV annulus was within normal limits and right ventricle enlarged significantly. One neonate died due to post-operative complications. The size of TV and RV in this neonate was within normal limits, PA was patent with 50 mmHg RV-PA gradient. Other 4 most likely will have biventricular circulation (BV). Out of rest 26 fetuses, FPV would probably be possible in 8 fetuses. It was discussed in 3 couples, who refused. 1 family decided to terminate the pregnancy. In 5 the diagnosis was too late for FPV. None of those 7 who were life-born had BV circulation. None of fetuses whose anatomy was not suitable for FPV had BV repair, there were 2 IUUD and 3 neonatal death.

Conclusions: The prognosis for fetuses with PA/PS&IVS after fetal intervention is much better than in those with natural history. FPV is possible in fetuses with patent RVOT, good size of pulmonary valve and branches. More precise criteria for FPV are necessary. Fetuses with hypoplastic RV and atretic RVOT had poor prognosis.

O2-7

Improved Fetal Detection of Simple Transposition of the Great Arteries Results in Better Clinical and Surgical Outcomes

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Introduction: The reported antenatal detection rate of transposition of the great arteries (TGA) is notoriously low (25%). Therefore the influence of antenatal detection on the surgical outcomes of TGA could not be accurately determined. The current inclusion of outflow tract view to the routine 20 week foetal anomaly screening protocols may improve the detection rate of TGA resulting in better neonatal and surgical outcomes.

Methods: All children who were diagnosed with simple TGA (with or without VSD, but no other cardiac abnormality) at UHW between 1998 and 2013 were included in this study. The antenatal-postnatal diagnosis, clinical and surgical data were retrospectively reviewed; the fetal, neonatal and surgical outcomes were evaluated.

Results: There were 79 patients; 58 diagnosed postnatally, 21 antenatally. 36 had isolated TGA and 43 had an additional VSD. Antenatal detection rate improved from 0% to 75% in the last 7 years. All patients in antenatal detected group were delivered at term, but there were eight preterm deliveries were in postnatal group. 40% of cases diagnosed postnatal were unwell at presentation; 15% required inotropes and 80% ventilatory support. The need for prostin infusion following delivery was 68% and balloon septostomy was required in 60% of the cases before surgery. Major postoperative complications were more common in postnatally diagnosed patients (26% vs 10%, $p < 0.01$). One death was recorded in the antenatal group but all other 8 deaths were in the postnatal group. Preoperative mortality rate was 10% and operative mortality was 1.2% with no death being recorded after 2008 which coincided with the adoption of outflow tract view into the routine cardiac anomaly screening planes in Wales.

Conclusions: Inclusion of outflow tract view represents a major leap in antenatal detection of TGA. Increased antenatal detection rates of TGA has resulted in better cardiovascular status at presentation, more favourable clinical outcomes with lower postoperative complications and virtually eliminated mortality rates in our cohort.

O3-1

Diabolo stents in various positions: an innovative way to reduce vessels size or to stabilize stents

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Background: Stents are usually used to open vessels or to maintain their patency. We describe the use of stents to reduce the diameter of vessels. By using 2 stents with different properties, we have created self-made diabolo stents in various positions.

Material and Methods: All patients with diabolo stent placement were reviewed.

Results: 7 patients were eligible. 4 patients had percutaneous valve insertion in structures larger than 24-mm (tricuspid valve, $n=2$;

pulmonary valve, n = 1; Inferior Vena Cava, n = 1). Reduction was made by the use of a short EV3 LD mega (S17-26, EV3, France) fitted over a CP covered stent (8Z39, Numed, Canada), the 2 stents being crimped and deployed on a large balloon creating a landing zone. Melody deployment was feasible. Two patients had Blalock-Taussig stenting to treat important cyanosis. Procedure was complicated by pulmonary overflow. In one patient, reduction of the Blalock was done with a diabolito sent: a coronary stent Biotronic 4*18 mounted on an Atrium V12 10*38-mm. The second patient who also had a Glenn shunt had an endovascular PA band with a diabolito stent placed in the right pulmonary artery (a Tsunami 5*12-mm on a covered CP8Z34 crimped on a 18-mm BIB). Because of persisting symptoms: the patient had PA disconnection: endovascular band was closed with a muscular VSD plug using the shape of the diabolito stent. A patient with Eisenmenger syndrome and cor triatriatum sinister had percutaneous creation of an ASD. The ASD was then stented to ensure patency using the diabolito shape technique.

Conclusions: we report the used of the combination of 2 stents to create diabolito stents in various position with good results.

O3-2

BeGrow™ - a new stent with special design for growing vessels: First results of a preclinical animal study

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Background: There still does not exist an ideal treatment solution for stenotic pediatric vessels with growth potential. The presented cobalt-chromium-stent (BeGrow™, Bentley Innomed GmbH, Germany) with a designated diameter of 5-6 mm can be introduced and implanted via a 4 Fr sheath. It allows redilation up to 12 mm with preserved stent integrity and stent length. Beyond this diameter it can be opened/dilated with the help of predetermined circular breaking points. Therewith this stent allows a thorough interventional treatment approach into adulthood.

Methods and Results: Within the animal study 11 pigs with a body weight of 4-5 kg received in each case 3 BeGrow™ stents into the abdominal aorta via a transfemoral vessel access. Implantation procedure was successful and uneventful in all. Within the following 3-4 months the stents were redilated and finally opened in repeated procedures up to a vessel diameter of 12-18 mm. At each step of the procedure some of the stents together with the circumferential aortic vessel wall were analysed histologically. They did not show any signs of inflammation or thrombus formation. The results will be presented in detail.

Summary: The BeGrow™ stent can be implanted safely and straightforwardly via a 4 French sheath. Repeated redilations without stent shortening are possible. Beyond 12 mm stent diameter an opening of the stents can be achieved under preserved vessel configuration. A clinical multicentre trial is in preparation.

O3-3

Tricuspid regurgitation does not impact right ventricular remodeling after percutaneous pulmonary valve implantation

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The aim of the study was to investigate the impact of tricuspid regurgitation (TR) on right ventricular size after percutaneous pulmonary valve implantation (PPVI). We compared patients with pulmonary stenosis (PS) and TR (PS/TR+ group) to patients with PS and without TR (PS/TR- group) as well as patients with pulmonary regurgitation (PR) with and without TR (PR/TR+ and PR/TR- groups), respectively.

Method: In a retrospective study among all patients treated by PPVI (n = 173) those with moderate or severe TR were selected. Indication for PPVI was PS (n = 11) and PR (n = 9). For all patients included in the study, matched pairs were selected considering cardiac diagnoses, age at implantation, functional state and number of previous surgeries. We analyzed hemodynamic parameters by CMR, the degree of TR by echo and data of exercise testing in all patients before PPVI and 6 months after.

Results: None of the patients had significant post-procedural PS or PR. Regardless of the presence of TR the RV size decreased significantly in all groups (mean end diastolic volume index before and after PPVI in ml/m²: PS/TR+ group: 113 vs. 95 (p = 0.01); PS/TR- group: 87 vs. 74 (p = 0.01); PR/TR+ group: 133 vs. 112 (p = 0.02) and PR/TR- group: 116 vs. 94 (p = 0.008); mean end systolic volume index before and after PPVI in ml/m²: PS/TR+ group: 62 vs. 45 (p = 0.02); PS/TR- group: 39 vs. 29 (p = 0.02); PR/TR+ group: 85 vs. 70 (p = 0.05); PR/TR- group: 59 vs. 45 (p = 0.01)). There was no statistically significant difference in the reduction of RV Volume indexes between PS/TR+ and PS/TR- groups and PR/TR+ and PR/TR- groups, respectively. The degree of TR improved in 72% of the PS/TR+ group (p = 0.002) and 77% of the PR/TR+ group (p = 0.001). The improvement in exercise capacity after PPVI did not differ significantly in any of the groups.

Conclusion: Tricuspid regurgitation does not impact right ventricular remodeling after percutaneous pulmonary valve implantation, neither in patients with prevailing PS nor in those with PR.

O3-4

Reversible Pulmonary Artery Banding for Left Ventricular-DCM with preserved RV function: feasibility of percutaneous PA-debanding

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Background: Dilated cardiomyopathy (DCM) in childhood has a considerable morbidity, mortality and high incidence of heart transplantation (HTX). Recently we published our initial experience with reversible pulmonary artery banding (rPAB) as an additional strategy in young children with LV-DCM and preserved-RV-function instead HTX. Purpose of the study is to demonstrate indication and feasibility of percutaneous PA-debanding as part of concomitant therapy.

Methods: Since April 2006, 24 children (age < 3 yrs) with LV-DCM were treated by rPAB considering defined entry criteria; 18 received only off-pump rPAB without any associated open-heart surgery. For creating a balloon-dilatable, reversible PAB, the bands were secured with 6-0 polypropylene sutures, for growing-in by double suture technique. All pts were closely monitored. In case of a too tiny rPAB with reduced RV-function, including moderate tricuspid regurgitation, secondary rise of BNP serum levels, pts were admitted for partial or total PA-debanding by transcatheter ballooning. Catheter procedures were performed under gentle sedation. After hemodynamic assessment and angiographic delineation of the PAB, high-pressure balloons were used for PA-debanding

followed by hemodynamic re-assessment. The pts were discharged home after clinical and echocardiography re-evaluation.

Results: Until now, 20 of 22 pts were partially de-banded without procedural mortality. Of these 20 pts 8 underwent a complete debanding (defined as a residual gradient < 20 mmHg). The mean interval between rPAB and first de-banding was 440 + /-235 days. Mean pressure gradient (dp) across the PAB before de-banding was 64 + /- 17; after ballooning 36 + /-40 mmHg. BNP level before de-banding were 260 + -289 and afterwards 58 + -66. Two LV-NC pts deteriorated several months after complete debanding and died later, one on Berlin-Heart. Two pts have not yet met the criteria for PA-de-banding. Two, on rPAB non-responding patients, one with LV-EFE (Endocard Fibroelastosis) and one with LV-NC needed HTX, which was successfully performed in both.

Conclusion: rPAB is a promising, safe and feasible procedure, including the concomitant de-banding strategy, which is based on surgical double stitch technique; it offers the possibility of a gradual PA-de-banding addressing the individual growth and need of the young patient.

O3-5

Percutaneous Treatment of Residual Lesions in Postoperative Pediatric Cardiac Surgery Infants receiving Extracorporeal Membrane Oxygenation support

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Introduction and Objective: Residual lesions in cardiac surgery patients receiving ECMO support may result in incomplete recovery of cardiac function. The aim of this study was to examine the incidence and clinical outcomes of postoperative residual lesions of pediatric cardiac surgery patients who received ECMO support.

Methods: A retrospective observational study was undertaken to collect the pediatric cardiac surgery patients who received ECMO support within 14 days of surgery between 2003-2014. A hemodynamically significant cardiac lesion that required intervention for successful decannulation was defined as a residual lesion. Demographic data, complexity of the disease, surgical data, indications for ECMO assistance, echocardiographic findings, and outcomes of cardiac catheterization outcomes were studied. Evaluation of residual lesions based on the duration of ECMO support, interventions performed, and clinical outcomes were also examined.

Results: Residual lesions were evaluated by catheterization in 75 of 100 postoperative patients placed on ECMO. The indications for ECMO were: off CBP (38%), low cardiac output (32%), cardiac arrest requiring cardiopulmonary resuscitation (26%) and arrhythmia (2,7%). Residual lesions were detected in 52 patients (69%), predominantly in branch pulmonary arteries (n = 8), aortic arch (n = 14), shunts (n = 8) and coronary arteries (n = 6). There was unexpected diagnostic information not foreseen by echocardiography in 33 (63%). Percutaneous intervention was performed in 44 patients (59%), 16 during the first 3 days on ECMO support. Early intervention improved the rate of decannulation and ECMO duration compared with later intervention (71% versus 35% and 6 days versus 9 days, p = 0.037). In those who received surgical reintervention, 11 patients, the rate of decannulation was 36%.

Conclusions: In our experience residual lesions are present in about half of patients requiring ECMO support after cardiac surgery. All postoperative pediatric cardiac unable to wean off ECMO

successfully should be evaluated actively to find residual lesions. Earlier detection of residual lesions and percutaneous intervention are associated with improve clinical outcome.

O3-6

Multiple pretesting provides for the Melody valved stent an adequate landing tube with minimal recompression or fracture

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Introduction: Long term function of the Melody valved stent depends on conduit integrity; stent fracture and recompression which has been reported to exceed > 20% in first year. Pretesting adds stiffness to the scaffold, reduces the stress amplitude and therefore may delay or avoid metal fatigue and fracture.

Patients and Methods: A/in vitro: stents in various combinations of CP stent (Numed, NY, USA) which is also the Melody stent, and Andrastent XXL (Andramed, Reutlingen, DE) at 22 mm were submitted to pressure tests to quantify stiffness.

B/patient implantation: all patients treated by Melody PPVI; systematic follow-up with dedicated database. Doppler velocity across the RVOT stent tube to assess tube and valve function. Chest X-ray at 6 months and annually to look for stent recompression or fractures.

Results: A/deploying stents into each other significantly increased stiffness; for similar lengths the CP stent was slightly stiffer.

B/131 patients had the RVOT presented in 2006-2014; mean age 19.9 years (3.8 – 81,6); follow-up 2.8 years (31 days – 8 years). Pre-stenting evolved significantly: stents were implanted until the outflow tract became a rigid non-restrictive tube without relative motion nor wringing. In 33 patients without a previous surgical conduit a single open cell bare stent was used. In patients with a surgical conduit, 1 stent was used for pre-stenting in 98 patients, 2 stents in 21 patients, 3 stents in 6 patients and in 2 patients up to 4 stents were implanted prior to the implantation of the valved stent. The residual gradient after PPVI was 20 mmHg (8-53) Vmax 2.1 cm/s (1.4-3.6); the final diameter was 22 (18-24). During follow-up no relevant increase in peak RVOT velocity (+0,2 m/sec at 3 y, 0,5 m/s at 5 y). During follow-up of mean 2.8 years, we observed stent fractures in 6 patients, anterior recompression in 3 pts; an increase in gradient was seen in only 1 patient (+25 mmHg).

Discussion: Adequate pretesting of the RVOT before revalvulation offers good stent support which nearly abolishes Melody recompression or fracture.

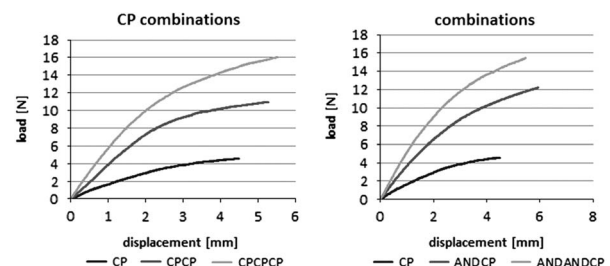


Figure.

O3-7

Stenting of the arterial duct in infants with functional univentricular heart: Single center experience in 68 patients

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Introduction: Ductal stenting has emerged as a non-surgical alternative to surgical aorto-pulmonary shunt in patients with duct-dependent or decreased pulmonary blood flow. This study reports our experience with duct stenting in 68 patients with functional univentricular heart (FUH).

Method: We retrospectively analyzed 68 infants who had FUH in 136 patients underwent cardiac catheterization for duct stenting in our institute between 2004 and 2014.

Results: Median age was 20 days (3 days–8 months) and median weight was 3.4 (2.7 – 6.8) kg in 68 patients. 26 had pulmonary atresia with intact ventricular septum, 15 had tricuspid atresia or severe hypoplasia, 10 had unbalanced complete AVSD, 10 had double/single inlet ventricle and 7 had miscellaneous type of FUH. Ductus was arising from descending aorta in 43, arcus aorta, (vertical) in 16, innominate artery (atypical) in 6 and bilateral in 3. Implantation was successful in 66 of 68 (97%), unsuccessful due to acute ductal constriction in two. Implantation performed retrograde in 53, antegrade in 11 and both (bilateral duct stenting) in one. Oxygen saturation increased from $70 \pm 7.6\%$ to $87 \pm 4.6\%$, immediate after the procedure. In one patient stent was migrated to descending aorta and underwent to surgery. One patient died after successful stent implantation probably due to pulmonary overflow. The follow-up period ranged from 3 months to 10 years (median 64 months). 48 infants reached to Glenn anastomosis without surgical intervention. Fontan completion was achieved in 19 of them. However, aorto-pulmonary shunt was required in 11 infants after 3 days to 7 months of the procedure, 2 were lost to follow up. Three patients died without intervention 4 days–6 months later during follow-up. The deaths were not related to the procedure. Stent redilation was performed in 14 patients due to decreasing of oxygen saturation in 3 to 10 months.

Conclusion: Stenting of the duct in infants with FUH is effective and safe alternative option as a bridge to second stage palliation. Mortality rate is comparable even better to conventional surgical shunt in FUH. Additional advantages of duct stenting to surgery are shortening hospital stay, eliminating problems of thoracotomy and reducing the number of operations.

O4-1

Right Ventriculo-arterial Coupling In Repaired Fattot Patients With Pulmonary Valve Regurgitation Before and After Pulmonary Valve Implantation: A CMR Study

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Background: Right ventricular-pulmonary arterial coupling plays an important role in the occurrence of right ventricular failure. Ventricular-arterial coupling is defined as Ea/Emax. Ea is the effective arterial elastance and an index of the post-load and includes vascular resistances, vessel compliance, vascular impedance, systolic and diastolic time intervals. Emax is the maximal systolic ventricular elastance. This is a load-independent index. Optimal ventricular arterial coupling is when Ea/Emax = 1.

Aim: To evaluate by cardiac magnetic resonance (CMR) the right ventricular-arterial coupling before and after surgical or transcatheter

pulmonary valve implantation in subjects who underwent surgery for tetralogy of Fallot and have pulmonary valve regurgitation.

Patients and Methods: We evaluated 34 patients (age 23 ± 21 years; BSA 1.61 ± 0.5) treated for tetralogy of Fallot who have pulmonary valve regurgitation and/or stenosis and underwent pulmonary valve implantation (surgical or transcatheter). CMR studies were performed before and 12 months after pulmonary valve implantation. The volumes and stroke volumes were adjusted for body surface area.

Results: Fifteen subjects were submitted to a surgical procedure while 19 underwent a transcatheter approach. All subjects completed the pre-operative and the 12 months follow-up evaluations. The Ea/Emax changed significantly between the pre-operative and the post-operative period (pre 1.42 ± 1.06 versus post 1.02 ± 0.43 ; $p = 0.027$). No relationships were found with age at procedure, age at evaluation, sex and group of treatment.

Conclusions: In our series procedures of pulmonary valve implantation normalize the right ventricular-pulmonary artery coupling at 12 months of follow-up.

O4-2

Cardiac magnetic resonance assessment of right ventricular performance after stage 2 palliation for hypoplastic left heart syndrome: a dual center study comparing the Norwood and Hybrid approach

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Objectives: Stage 1 palliation of hypoplastic left heart syndrome (HLHS) can be achieved either by the Norwood (NW) procedure or the Hybrid (HY) approach. These procedures result in different hemodynamic profiles that may have an impact on ventricular performance in the long term. Accordingly, the aim of this study was to compare CMR data of HLHS patients after stage 2 who underwent either a NW or the HY strategy in two European centres.

Methods: HLHS patients after stage 2 palliation who underwent an initial classic NW operation using a BT shunt ($n = 38$, mean age 2.4 ± 0.7 years) or the HY approach consisting of bilateral pulmonary artery banding and arterial duct stenting ($n = 38$, mean age 2.1 ± 1.0 years, $p = 0.12$) were included. The CMR protocol consisted of cine assessment of right ventricular (RV) size and function. Feature tracking analysis was performed to quantify RV strain and intraventricular synchrony parameters.

Results: NW patients had significantly larger RV enddiastolic volumes (91 ± 24 vs 84 ± 33 ml/m²; $p = 0.03$) and higher indexed stroke volumes (53 ± 12 vs 47 ± 12 ml/m², $p = 0.01$) compared to the HY group whereas global pump function (ejection fraction 59 ± 10 vs $58 \pm 9\%$; $p = 0.71$) and cardiac output (4.8 ± 1.2 vs 4.9 ± 1.2 l/min/m², $p = 0.27$) did not differ between the two groups. In the HY group RV circumferential (-15.1 ± 6.2 vs $-18.4 \pm 5.8\%$, $p = 0.01$) and radial strain (13.1 ± 7.8 vs $21.1 \pm 9.7\%$, $p < 0.001$) was lower and intraventricular dyssynchrony significantly increased (88 ± 48 vs 51 ± 24 ms, $p < 0.001$ and 97 ± 59 vs 59 ± 49 ms, $p < 0.001$). No difference was found in

longitudinal strain (-14.3 ± 5.4 vs $-16.5 \pm 5.7\%$, $p=0.08$) and dyssynchrony (85.4 ± 31.9 vs 81.6 ± 38.1 ms, $p=0.78$).

Conclusions: The initial palliative strategy in HLHS patients has significant effects on ventricular size and performance at mid-term follow-up. Future studies comparing the two approaches are necessary to assess whether the observed differences determine functional outcome in the long-term.

O4-3

Volume and Function of the Right Ventricle in Ebstein Anomaly: Knowledge-based Three Dimensional Reconstruction of Echocardiographic Images Compared to Cardiac Magnetic Resonance

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Objectives: Quantification of right ventricular (RV) volume in Ebstein anomaly is challenging, given the different degrees of displacement of the tricuspid valve (TV). Echocardiographic knowledge-based 3D reconstruction (3DR) of RV volumes has been validated in normal and dilated RV. We sought to assess feasibility of 3DR in Ebstein patients and to compare the RV volumes with volumes obtained by cardiac magnetic resonance (CMR).

Methods: Ebstein patients with and without TV reconstruction were prospectively recruited in 4 centres and underwent 3DR and CMR in our core lab on the same day. Patients with pacemaker or Fontan palliation were excluded. Knowledge-based 3DR was performed with Ventripoint™ Diagnostics, USA. RV volumes were calculated using 3DR and the summation disc method on short axis images (saCMR) and on axial images (axCMR) by two independent experienced observers. Intermodality differences were assessed using correlation coefficient and Bland-Altman analysis and expressed as % mean difference \pm standard deviation and 95% limits of agreement.

Results: Eighteen patients underwent 3DR and CMR examination. 3DR was feasible in 78% of the patients, image quality being insufficient in 4/18. In 14 patients significant correlation was present for all RV volumes among all imaging modalities. RVEDV in 3DR correlated best with saCMR with a mean difference of $0.6 \pm 5.4\%$ ($-11/10$). Agreement between 3DR and axCMR was $4.8 \pm 9.4\%$ ($-23/13.6$), between axCMR and saCMR $4.2 \pm 6.8\%$ ($-9/17$). For RVESV agreement was good between 3DR and saCMR $-1.9 \pm 9.4\%$ ($-20/16$), weaker between 3DR and axCMR was $6.5 \pm 13\%$ ($-32/19$), and between both CMR methods $4.6 \pm 14\%$ ($-24/33$). Reasonable agreement was found for RV EF% between 3DR and saCMR $0.9 \pm 5.1\%$ ($-9/11$). In contrast wide limits of agreement were observed between 3DR and axCMR $1.7 \pm 8.9\%$ ($-16/19$) and between saCMR and axCMR $-0.9 \pm 9.7\%$ ($-19/18$).

Conclusions: 3DR echocardiography is feasible in Ebstein anomaly patients. 3DR RVEDV measurements show good agreement with saCMR and axCMR. Limits of agreement for RVESV and RV EF% are wider, but still comparable with published ranges obtained by CMR. Since volumes obtained by different techniques are not interchangeable, we recommend to use consistently the same modality for follow up of Ebstein anomaly patients.

O4-4

Disturbed Intra-cardiac Flow Organization after AtrioVentricular Septal Defect Correction as assessed with 4DFlow MRI and Quantitative Particle Tracing

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Introduction: The normal pattern of left ventricular (LV) inflow and ejection affects the efficiency of cardiac pumping performance. Patients after atrioventricular septal defect (AVSD) correction have an abnormal atrioventricular valve shape and an altered inflow direction. Whether this influences blood flow efficiency and long term follow-up is unknown. We aimed to quantitatively describe the LV blood flow pattern using 4-dimensional velocity-encoded cardiac magnetic resonance imaging (4DFlow MRI) and particle tracing in healthy volunteers and corrected AVSD patients.

Methods: 32 patients (25 ± 14 years) and 30 healthy volunteers (26 ± 12 years) underwent 4DFlow MRI. At end-diastole the LV was evenly filled with particles and subsequently tracked by backward and forward particle tracing to analyse the LV multi-componential flow as introduced by Eriksson [JCMR, 2010] discriminating 1. direct flow entering and leaving LV within one cycle, 2. retained flow entering during diastole but remaining in LV during next systole, 3. delayed ejected flow already in LV before diastole and leaves LV during systole 4. residual volume, 5. Regurgitant flow. The path of inflowing particles (i.e. direct and retained flow) during diastole was evaluated using the 16-segment LV cavity model.

Results: Patients showed a smaller percentage of direct flow compared to controls (Figure, * indicating $p < 0.05$). In patients versus controls more inflow was observed in the basal inferior segment ($22 \pm 11\%$ versus $17 \pm 5\%$, $p=0.005$), with less direct flow but more retained inflow. In patients, more inflow reached into the mid-ventricular level compared to controls ($68 \pm 13\%$ versus $58 \pm 9\%$, $p < 0.001$), most notably retained inflow in infero-lateral and antero-lateral segments. Subsequently, in patients more

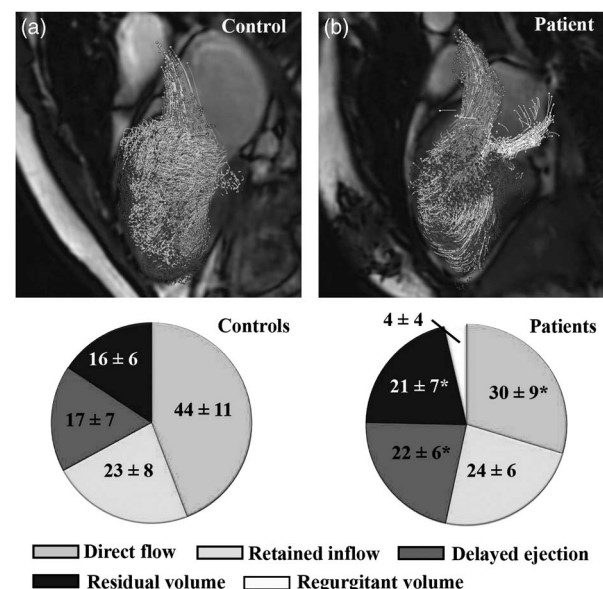


Figure.

(mostly retained) inflow reached into the apex ($23 \pm 13\%$ versus $14 \pm 7\%$, $p < 0.001$), which correlated with early peak filling velocity ($r = 0.637$, $p < 0.001$). Patients with a corrected complete or intermediate AVSD presented with less direct flow ($24 \pm 8\%$ versus $33 \pm 8\%$, $p = 0.003$) and more apical inflow ($30 \pm 14\%$ versus $18 \pm 12\%$, $p = 0.014$) compared to corrected partial AVSD. **Conclusions:** Multi-component particle tracing combined with 16-segment analysis quantitatively demonstrated altered LV filling and ejection patterns after AVSD correction, with less direct flow and more (retained) inflow in the apical and lateral LV cavity segments, which may contribute to a decreased cardiac pumping efficiency.

O4-5

Altered Vortex Ring formation in the Left Ventricle in Patients after AtrioVentricular Septal Defect Correction using 4DFlow MRI and Vortex Core Analysis

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Introduction: Vortex formation in the left ventricle (LV) is suggested to contribute to efficient blood pumping and altered vortex formation is associated with diastolic dysfunction. Patients after atrioventricular septal defect (AVSD) correction have abnormal valve morphology and inflow patterns [Calkoen et al. JMRI 2014], which may disturb normal vortex formation. We aimed to analyze vortex ring formation in AVSD-corrected patients compared to healthy controls.

Methods: 32 patients (age 26 ± 12 years) and 30 healthy controls (25 ± 14 years) were included. Whole-heart 4DFlow MRI was performed and the Lambda2 method was used to depict the cores of three-dimensional vortex ring structures in the velocity field in the LV at peak early filling phase. Subsequently, if a vortex ring was present the 3D position (circumferential, longitudinal and radial coordinates) and orientation of the vortex rings relative to the LV long-axis was defined and the circularity index was calculated to quantify the shape. Through-plane flow over the LAVV and the Vortex Formation Time (VFT) was quantified to analyze the relation of vortex flow with the inflow jet.

Results: Absence of a vortex ring core during E-peak (controls 0% versus patients 19%, $p = 0.015$) was more frequently reported in patients and accompanied by a high VFT (controls 2.4 ± 0.6 versus $5.1-7.8$ in patients without vortex ring) and in another two patients with abnormal valve anatomy. In 81% of the patients a

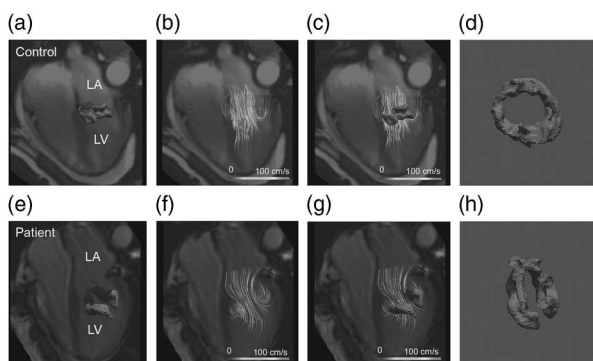


Figure 1. Vortex formation in the LV correlates well with streamlines (colours indicate velocity) and have a more tilted orientation and elliptical shape in patients.

Table 1.

	Controls (N = 30)	Patients (N = 26)	p
Circumferential	$90 \pm 26^\circ$	$70 \pm 21^\circ$	0.003
Longitudinal	0.19 ± 0.04	0.23 ± 0.07	0.015
Radial	0.26 ± 0.07	0.33 ± 0.08	0.001
Orientation	$71 \pm 9^\circ$	$50 \pm 20^\circ$	< 0.001
Circularity Index	0.80 ± 0.08	0.70 ± 0.13	0.002

vortex core was present, but this was more anterior and apical positioned, closer to the ventricular wall, with a more elliptical shape (Figure 1H) and oblique orientation (Figure 1G) (Table 1) and related to blood inflow direction ($r = 0.41$ $p = 0.037$) and LAVV inflow characteristics.

Conclusions: This study confirms in vivo using 4DFlow MRI, the influence of an abnormal LAVV and inflow on 3D vortex ring formation during LV filling. The quantitative comparison revealed altered vortex formation in corrected-AVSD patients compared to controls. Future studies are required to investigate the effect of absence or altered vortex ring formation on energy loss.

O4-6

Biventricular response to pulmonary artery banding in children with dilated cardiomyopathy assessed by cardiac magnetic resonance strain analysis

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Objectives: Pulmonary artery banding (PAB) has been proposed as a novel therapy in infants with severe left sided dilated cardiomyopathy (DCM). Although potential mechanisms leading to improvement of left ventricular function have been suggested, the response of the right ventricle (RV) to increased afterload and its impact on left ventricular (LV) remodelling remains unknown. Our study aimed to assess changes in biventricular properties after PAB in severe left sided DCM using cardiac magnetic resonance (CMR) feature tracking (FT) analysis.

Methods: Thirteen patients who underwent central PAB due to severe DCM were evaluated using standard cine CMR measurements of cardiac size, mass and function. CMR-FT software was used to quantify biventricular longitudinal (LS), circumferential (CS) and radial (RS) strain including intraventricular synchrony before (mean age 5.8 ± 4.0 months) and after PAB (mean age 19.8 ± 11.3 months). **Results:** LV enddiastolic (153 ± 54 to 65 ± 11 ml/m², $p < 0.001$) and endsystolic volumes (116 ± 52 to 32 ± 11 ml/m², $p < 0.001$) decreased and LV ejection fraction (18 ± 7 to $53 \pm 12\%$, $p < 0.0001$) increased significantly after PAB. LV myocardial mass dropped significantly from 103 ± 29 to 71 ± 33 g/m² ($p = 0.002$). LV-LS (-5.0 ± 3.4 to $-8.2 \pm 4.5\%$; $p = 0.03$), LV-CS (-2.8 ± 2.1 to $-10.7 \pm 3.5\%$, $p < 0.001$) and LV-RS (7.0 ± 6.7 to $17.8 \pm 9.2\%$, $p < 0.001$) increased and intraventricular dyssynchrony diminished significantly. RV size and function (EF 54 ± 12 to $55 \pm 12\%$, $p = 0.69$) did not change while RV-LS (-7.0 ± 3.6 to $-11.9 \pm 5.5\%$, $p = 0.01$) and RV-CS (-4.2 ± 4.3 to $-8.1 \pm 3.3\%$, $p = 0.02$) showed significant increment.

Conclusions: In infants with left sided DCM and severely depressed function, PAB induces a rise in RV strain thereby preserving RV dimensions and function. Recovery of LV size and function is accompanied by improved intraventricular synchrony.

O4-7

Cardiac index during routine cardiovascular magnetic resonance under general anaesthesia

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Objectives: Cardiac index (CI) can be measured non-invasively by cardiovascular magnetic resonance (CMR)[1]. Young children need some form of sedation during CMR. However, only scarce information is available of CI during anesthesia. Therefore, the aim of this study was to determine CI levels during routine standard CMR of patients with congenital and acquired heart disease under general anesthesia (GA) and compare them with CI of patients without GA and CI of healthy individuals.

Methods: To assess the impact of GA on CI were retrospectively reviewed one hundred twenty seven measurements of CI and EF in 75 patients with congenital and acquired heart disease, who underwent a routine clinical CMR at our institution over a 7-year period and have a CI and LVEF measurements in the study. As control 15 young healthy volunteers were selected.

The entire cohort was divided into three groups: 43 measurements were done in 38 patients with congenital and acquired heart disease with GA, (median age 3.6(0-40) yrs.); 69 measurements in 37 patients with congenital and acquired heart disease without anesthesia, (10(1-17) yrs.); and 15 measurements in 15 healthy controls without anesthesia, (13(7-19) yrs.).

Results: CI was significantly lower in the GA group compared with patients without GA (2.8 ± 0.9 l/min/m² vs. 3.6 ± 0.8 l/min/m² $p < 0.0001$) and controls (2.8 ± 0.9 l/min/m² vs. 4.0 ± 0.7 l/min/m², $p < 0.0003$).

There was no statistically significant difference between pts. without GA and controls ($p = 0.223$). There was no statistically significant difference in LVEF between groups. There was no correlation between the CI and LVEF.

Conclusions: CI is significantly lower during any form of anesthesia than without anesthesia. Therefore hemodynamic parameters (e.g. ventricular volumes) under anesthesia may not represent true hemodynamics under resting conditions. Further studies need to determine acceptable levels of CI during any form of anesthesia.

O5-1

Comparison of neurodevelopmental outcome at 1 and 4 years in children undergoing Hybrid and Norwood procedures for hypoplastic left heart syndrome

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Background: Data on long-term neurodevelopmental outcome of children treated for hypoplastic left heart syndrome (HLHS) comparing Hybrid and Norwood procedure are needed.

Methods: Neurodevelopmental outcome assessment at 1 and 4 years of age of infants undergoing Hybrid (2006-2008) and Norwood procedure (2004-2008) for HLHS after implementation of both treatment options in a single centre setting. Assessment included at 1 year of age, psychomotor (PDI) and mental developmental index (MDI) of Bayley Scales of Infant Development III, at 4 years of age the Wechsler Primary Preschool Intelligence Scale - III, and Movement-ABC 2. Furthermore, we compared outcome of early (within first year after implementation) vs. late period (after first year).

Results: Thirty-one (18 males) infants were treated with Hybrid (n = 13) and Norwood (n = 18) procedure. Twenty children survived until age of 1 year. Motor (PDI) and cognitive (MDI) outcome at 1 year were significantly impaired compared to the norm [PDI 57(49-99), $p < 0.001$; MDI 91(65-109), $p = 0.002$], but not different comparing Hybrid (n = 9) and Norwood (n = 11) procedure [PDI: Norwood 56.5(49-81) vs. Hybrid 65(50-99), $p = 0.18$; MDI: Norwood 93(65-109) vs. Hybrid 88(71-102), $P = 1.0$] (1). At 4 years, 16 of 20 children were reevaluated (late death n = 1, lost to follow up n = 3). Overall, both cognitive and motor performance was poorer than the norm [IQ: 89(76-116), $p = 0.02$; motor outcome: $p = 0.002$], but not different between Hybrid (n = 7) and Norwood (n = 9) procedure [IQ: Norwood 92(80-104) vs. Hybrid 88(76-116), $p = 1.0$; motor outcome: $p > 0.8$]. However, cognitive performance of children treated in the early learning period was significantly lower compared to children treated in the late period independent of performed procedure [IQ early period 87(76-101) vs. late period 96(80-116), $p = 0.03$]. Motor outcome was comparable between early versus late treatment period ($p > 0.8$).

Conclusions: Overall, cognitive and motor functions are impaired at 1 and 4 years of age in children with HLHS. As described for the 1 year outcome, at 4 years of age there are no evident differences between Hybrid and Norwood procedure, but we could observe that an institutional learning curve affects cognitive outcome.

Reference:

(1) Knirsch W et al., Eur J Cardiothorac Surg 2012

O5-2

Perinatal Risk Factors for Mortality after Norwood Palliation of Hypoplastic Left Heart Syndrome

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Background: Since 1993, pediatric cardiology in Sweden has been centralized to Lund and Gothenburg, which together serve a population of nearly 10 million people. We sought to determine the impact of several perinatal variables on survival in patients with HLHS.

Methods: A retrospective survey of 90 consecutive HLHS cases who underwent stage I Norwood palliation (S1P) between 1993 and 2013 at our center. Data on prenatal diagnosis (PND), birth characteristics, preoperative variables, type of shunt at S1P (Blalock-Taussig (BT) or Sano (S)) and early postoperative variables were collected from the hospital's database. Death occurring between S1P and stage 2 palliation (interstage I mortality; ISM-I), between stage 2 and 3 (ISM-II) and overall mortality were chosen as outcome variables.

Results: During the study period, there was a relatively steep decline in ISM-I from nearly 60% in the beginning of the 1990s to a plateau < 10% during the last decade. PND did not improve survival but was associated with earlier S1P ($p = 0.001$) and with less preoperative tricuspid regurgitation (TR; $p = 0.04$). Those who underwent S1P of the Norwood procedure after the 1st week of life had higher risk for death within the first month of life (Odds Ratio (OR) 7.2, $p = 0.002$). Restrictive atrial communication ($p = 0.005$; OR 6.5) was associated with increased ISM-I, while TR increased risk for ISM-II ($p = 0.02$, OR 2.4). Norwood procedure with BT shunt was exclusively performed during 1993-2002, and was linked to higher ISM in patients with aortic atresia-mitral stenosis (AA-MS; $p = 0.06$, OR 2.8). During 2003-2013, in which the majority received S shunt, the only predictors for ISM-I were prematurity and low birth weight ($p < 0.05$ and OR > 5 for both).

Conclusion: Lower preoperative weight and restrictive atrial communication remain important risk factors for mortality, whereas Norwood palliation within the 1st week of life improves survival. Patients with AA-MS appear to be at higher risk for ISM-I during Norwood surgery with BT shunt, but no such trend could be observed in those palliated with Sano shunt.

O5-3

Prognostic value of serum biomarkers of cerebral injury to assess neurological outcome after in-hospital cardiopulmonary resuscitation in pediatric patients

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Objectives: Neurological morbidity and mortality is considerable after in-hospital pediatric cardiopulmonary resuscitation (CPR) and early assessment of cerebral injury severity is helpful to guide post-CPR therapy. While an association of the cerebral injury serum biomarkers neuron-specific enolase (NSE) and protein S100b with neurological outcome after CPR has been documented in adults, their prognostic value in children has not been comparably well studied.

Methods: A retrospective analysis of patients admitted to our pediatric cardiac intensive care unit during the last 3 years identified 32 children (congenital heart disease n = 28, neonatal asphyxia n = 4) who required in-hospital CPR and had subsequent serial measurements of NSE and S100b. Neurological outcome at discharge was classified using the pediatric cerebral performance category score (PCPC). Favorable outcome was defined as $\Delta PCPC \leq 1$ and unfavorable outcome as $\Delta PCPC > 1$. Serum levels of NSE and S100b were analyzed towards their ability to predict post-CPR neurological outcome.

Results: Survival to discharge was 69% (n = 22). In 6 patients neurological assessment was not performed due to early death from non-cerebral cause (heart failure n = 4, sepsis n = 2), they were excluded from further analysis. Of the remaining 26 patients, 18 (69%) had a favorable neurological outcome. Both NSE and S100b were significantly higher in patients with unfavorable neurological outcome at 24 hours (NSE $128.9 \pm 25.9 \mu\text{g/l}$ vs. $53.6 \pm 11.1 \mu\text{g/l}$, $p = 0.003$; S100b $2.74 \pm 1.32 \mu\text{g/l}$ vs. $0.51 \pm 0.15 \mu\text{g/l}$, $p = 0.03$) and 48 hours (NSE $139.7 \pm 26.4 \mu\text{g/l}$ vs. $50.2 \pm 14.9 \mu\text{g/l}$, $p = 0.02$; S100b $6.26 \pm 3.97 \mu\text{g/l}$ vs. $0.26 \pm 0.06 \mu\text{g/l}$, $p = 0.02$) after CPR. Area under the curve in receiver operating characteristic curves was 0.89 and 0.87 for NSE and 0.80 and 0.91 for S100b at 24 and 48 hours, respectively. However, cut-off values for NSE and S100b with the best specificity of 100% to predict adverse neurological outcome showed only poor sensitivity (28.6% and 33.3% for NSE and 28.6% and 71.4% for S100b at 24 and 48 hours, respectively).

Conclusions: Serum levels of cerebral injury biomarkers NSE and S100b have the potential to predict adverse neurological outcome at an early stage after pediatric CPR and thus may prove useful to guide therapeutic decisions and parental counseling. However, larger prospective studies are needed to comprehensively evaluate diagnostic accuracy and determine reliable cut-off values.

O5-4

Failing bidirectional cavo-pulmonary anastomosis: Partial take down as possible way out strategy?

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Introduction: After bidirectional cavo-pulmonary anastomosis (BCPA), severe life threatening, therapy resistant hypoxemia is rare and affected patients need ECMO or surgical rescue therapy. Data about incidence and outcome of those therapeutic bail-out strategies, however, are rare and clinical pathways not published. We analyzed the results of our way out strategies in failing BCPA. **Method:** We screened the patient records of all 219 patients who underwent BCPA between 2006 and 2014 at our institution and selected those who suffered from severe therapy resistant hypoxemia, defined as repeated arterial oxygen saturations below 55% with consecutive lactate acidosis. We report the therapeutic strategy and the outcome of these patients with a special focus on partial take down surgery by an additional unilateral systemic-pulmonary shunt and functional separation of the right and left pulmonary arteries by ligation or clip.

Results: Out of 219 patients after BCPA, eleven (5%) showed severe therapy resistant hypoxemia. In 4 patients severe hypoxemia developed early after the end of bypass (median 2.8 h; 0.5 – 6 h) and in 7 patients late after surgery (median 19 d; 4 – 63 d). In 6 of the 11 affected patients, severe hypoxemia led to ECMO, in 2 cases the patients died in multi-organ failure without further surgical treatment. Thus, in total, 9 of 11 patients received a partial take down. Six of these 9 patients survived but 3 remained respirator dependent and died after 16, 90, and 150 days, respectively. These three patients had developed severe hypoxemia late after BCPA. Thus, 6 of 9 patients (67%) with partial take down survived until discharge at home. Fontan completion was performed in 2 patients successfully, 3 patients are waiting for completion but one further patient died at home with tracheostoma and intermittent respiratory support 3 years after surgery.

Conclusion: Partial take down by unilateral Glenn and systemic-pulmonary shunt as a way out strategy in severe therapy resistant hypoxemia after BCPA, is possible in selected patients. The overall mortality in failing BCPA, however, is considerable.

O5-5

Biventricular Repair in Aortic Atresia or Severe Left Ventricular Outflow Tract Obstruction – Yasui Operation

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Introduction: The Yasui operation is a rare surgical procedure, which may be employed to perform biventricular repair in patients with severe LVOTO as well as AA with VSD. We analyze the results of this complex procedure.

Methods: During 2013/2014 YO was performed in five consecutive patients (age: 8 days to 7 months, weight: 3.0 to 6.6 kg with LVOTO (n = 1) or AA (n = 4) and hypoplastic (n = 4) or interrupted aortic arch (IAA n = 1) each with a sufficiently sized left ventricle (mitral valve z-score ranged from -0.82 to +0.26). In two children the YO followed an initial PA banding.

Results: For LVOTO reconstruction Damus-Kaye-Stansel or Norwood procedure was combined with modified reconstruction of the aortic arch using autologous material in the dorsal part to preserve growth potential. The Rastelli procedure was performed using Contegra® 12 mm for reconstruction of the right ventricular outflow tract. Cross-clamp time was 44 to 73 min, time on respirator 4–9 days and hospital stay 13 to 18 days. Complications: One case of junctional ectopic tachycardia and two cases of chylothorax, but no further complications were observed. The median LV ejection fraction at discharge was 67% (58 to 83%) and

remained stable (EF > 55%) during midterm follow-up (1 to 14 months).

In one infant - after initial bilateral PA-banding - residual bilateral pulmonary stenosis was treated interventionaly one month after the YO. All children showed good somatic development (increase in weight percentiles from +5% to +20%).

Conclusions: The Yasui operation may be employed to create biventricular anatomy in young infants with severe LVOTO or AA with VSD and sufficient LV as a primary or staged procedure safely and with excellent outcome.

O5-6

Results and Long-term Follow-up for Double-Outlet Right Ventricle with Biventricular Repair

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Objective: To analyze surgical results in children with double outlet right ventricle (DORV) undergoing biventricular repair and to assess risk factors for mortality and reoperation.

Methods: Between 1993 and 2011, 482 consecutive patients presenting with DORV and 2 adequately sized ventricles. 433 patients (biventricular repair) were included into the study at two centers forming the National Referral Center. 49 patients were excluded (18 patients without surgical strategy, 31 patients with univentricular strategy).

Results: DORV were classified as DORV with subaortic (or doubly committed) ventricular septal defect (VSD) associated with right ventricular outflow tract obstruction (RVOTO) in 33% (n = 141), with subaortic (or doubly committed) VSD without RVOTO in 30% (n = 130), with subpulmonary VSD (Taussig-Bing Anomaly) in 32% (n = 139), with non-committed VSD in 5% (n = 23). Three types of repairs were performed: 1) intraventricular baffle repair (IVR), n = 149 (34%); 2) IVR with RVOT enlargement, n = 163 (38%); 3) IVR with arterial switch, n = 121 (28%). 135 patients (31%) had undergone prior palliative procedures. Early mortality was 7.4% and early cardiac reoperation was 6%. Actuarial survival rate at 10 years was 86.2%, and freedom from reoperation at 10 years was 61.8%. Median follow-up was 5.7 + /- 1.1 years. In the long term, reoperation and mortality was significantly more frequent for DORV with ncVSC (p < 0.01). The type of repair did not influence reoperation (p = 0.20) or mortality (p = 0.27). In multivariate analyses, the factors that influenced reoperation were: left ventricular outflow tract obstruction (p < 0.05), duration of cardiopulmonary bypass procedures (p < 0.01). The factors that influenced survival were: restrictive VSD (p < 0.01), anatomical abnormality of coronary (p < 0.05), duration of cardiopulmonary bypass procedures (p < 0.01), early cardiac reoperation (p < 0.01).

Conclusion: Factors affecting the prognosis of DORV are anatomical and surgical factors. However, there is no difference between the main types of biventricular surgical strategy.

O5-7

Surgical Outcome in Paediatric Patients with Ebstein's Anomaly: a Nation-wide Multicentre Study

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Objectives: To review outcome of a Dutch nation-wide cohort of paediatric patients with Ebstein's anomaly (EA) treated with surgery in childhood (0-18 years). We focus on the intention to treat as biventricular, 1.5 ventricle or univentricular repair, the unplanned reoperations and survival rates in this distinct group of EA patients.

Methods: Records of all paediatric EA patients born between 1980 and 2013 were reviewed. Clinical data including demographics, intended type of operation, intraoperative procedures and post-operative outcomes were studied.

Results: Of the 176 included EA patients, 112 were not operated during childhood, whereas the other 64 patients underwent a total of 110 operations. Median time of follow-up after diagnosis of this surgical subgroup was 115 months (range 0 - 216 months). Thirty (47%) patients required surgery within the first year of life, yet 18/64 already within the neonatal period. Intention to treat was biventricular (n = 37, 58%), 1.5 ventricle (n = 5, 8%) or univentricular (n = 22, 34%) repair. Of the 18 patients requiring surgery during the neonatal period, in 12 patients primarily a univentricular strategy was started, in the other six a biventricular repair was intended. 13/64 Patients required one or more unplanned reoperations (8, 2 and 3 patients in resp. the biventricular, 1.5 ventricle and univentricular group). Ten patients died of low cardiac output syndrome, hypoxemia with ventricular fibrillation, pulmonary hypertension or unknown cause, all within 33 days after surgery. Three patients died in the neonatal period, all but two patients died within the first year of life. The 1-, 5- and 10-years survival rates from time of diagnosis are 88%, 84% and 84% respectively. Univentricular repair was significantly associated with death (p = 0,004), operation in the neonatal period was not (p = 0.78).

Conclusions: While reporting on surgical interventions and their outcomes in patients with Ebstein's anomaly it is essential to differentiate between patients under the age of 18 years and adult patients because they represent a different spectrum of the anomaly. In paediatric patients biventricular repair is often not feasible. Mortality is related to intentional univentricular repair.

O6-1

Exercise Performance in Young Patients with Complete Atrio-ventricular Block: the Relevance of Synchronous AV Pacing

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Introduction: There are currently many pacing strategies for young patients with CAVB. The most frequent policy is to attempt a dual chamber system when possible. However, there is a group of patients that are still functioning with a non-synchronous ventricular pacing, raising the question of the ideal timing and circumstances to upgrade their systems. To solve this interrogation, we investigated the exercise performance of a group of children and young adults with CAVB and DDD pacemakers in both single and dual chamber pacing modalities.

Methods: Fifteen patients performed maximal exercise stress testing (EST) after programming VVIR or DDD modes with two hours of interval in a double blind protocol of study.

Results: Compared to VVIR pacing, DDD pacing resulted in: 1- Increase in the peak VO₂ (27,28 +/-6,40 ml O₂/kg/min to 29,65 +/-6,34 ml O₂/kg/min). 2-A longer test (9,73 +/-2,05 minutes to 10,82 +/-1,73 minutes) (Refer to Figure 1 for point 1 and 2). 3-A major increase in the heart rate achieved during peak exercise (VVIR 125,86 +/-15,36 bpm to 173,93 +/-12,97 beats per minute). 4- Decreased systemic non-invasive arterial blood pressured measured at maximal exercise (163,73 +/-26,44 mmHg to 150,06 +/-22,25 mmHg). 5- Higher maximal workload (172,66 +/-81,73 Watts to 199,73 +/-95,43). 6- Prolongation of the anaerobic threshold timing (7,69 +/-1,77 minutes to 9,64 +/-1,70 minutes) (Refer to Table 1 for point 3 to 6). 7- Better self-rate performance perception in all the patients (estimated by a numerical scale from 1 to 10).

Conclusions: Synchronous AV pacing contributes to increase both the exercise performance and the performance perception in 100% of the patients. This difference contributes to create a sense of "fitness" with repercussions in the overall health, self-esteem and life quality and encourages youngster to practice sports. Our experience tends to favour upgrading patients' systems to dual chamber before reaching the adolescent years, even if the centre policy is to prolong as long as possible the epicardial site in order to avoid long years of right ventricular pacing.

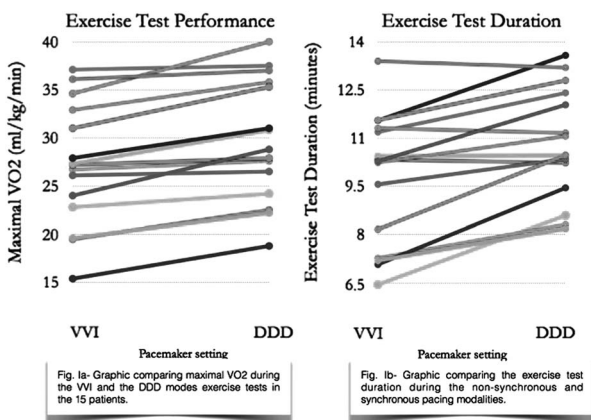


Figure 1.

Table.

	Peak HR		Peak SBP		Watts		AT	
	VVI	DDD	VVI	DDD	VVI	DDD	VVI	DDD
91	150	126	132	108	137	6,27	11,01	
136	160	183	172	229	249	11,56	13,40	
118	185	155	145	135	145	5,45	7,54	
151	180	156	153	116	182	8,21	9,18	
110	186	172	168	235	239	9,57	9,50	
144	179	226	187	364	430	9,50	11,44	
134	190	172	175	180	213	8,00	8,07	
123	171	140	141	89	114	7,19	10,03	
133	160	153	143	184	181	7,58	9,08	
103	165	129	113	78	98	8,06	10,08	
131	171	156	142	158	178	6,29	10,19	
129	159	180	143	140	146	7,32	10,27	
129	185	201	183	314	389	9,29	10,57	
130	193	149	124	127	162	5,07	7,36	
126	175	158	130	133	133	6,02	7,00	
Media	125,86	173,93	163,73	150,06	172,63	199,73	7,69	9,64
SD	15,36	12,97	26,44	22,25	81,73	95,43	1,77	1,70
P	<0.01		<0.01		<0.01		<0.01	

O6-2

Prognosis of radiofrequency catheter ablation in patients with twin atrioventricular nodes before TCPC procedure

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Background: Twin atrioventricular nodes (tAVNs) are sometimes associated with complex congenital heart disease such as heterotaxy syndrome and can be a cause of AV reentrant tachycardia (AVRT). Therefore, electrophysiological evaluation is important for total-cavo pulmonary connection (TCPC) candidates because catheter access to the atrium and AV valve is restricted after TCPC, and prophylactic catheter ablation (CA) is occasionally performed to inducible AVRT. On the other hand, few investigators have reported that ablating one of tAVNs might cause interventricular dyssynchrony and systemic ventricular dysfunction. The aim of this retrospective study is to investigate clinical results of prophylactic CA of unilateral AVN in TCPC candidates.

Methods: Electrophysiologic study was performed in 10 TCPC candidates (median age; 3 years) who had undergone Glenn operation and associated with common AV valve. Nine patients were heterotaxy syndrome (asplenia in 6 and polysplenia in 3). Spontaneous narrow QRS tachycardia had been demonstrated at the palliative operation and/or diagnostic catheterization in all patients.

Results: Two different QRS complexes without preexcitation were observed and two distinct His bundle electrograms were recorded at the anterior aspect and the posterior aspect of the common AV valve in all 10 patients. AVRT involving tAVNs was also induced in all patients and then we decided to perform CA of unilateral AVN. Successful RFCA of one AVN which showed recessive anterograde conduction was achieved and no further AVRT was inducible in all cases. The QRS duration was 80 ± 10 ms (63-90 ms) before CA and 85 ± 12 ms (68-95 ms) after CA. The ventricular ejection fraction was 62 ± 7% (55-68%) before CA and 60 ± 6% (53-69%) after CA.

Conclusion: Prophylactic CA of unilateral AVN for TCPC candidates with inducible AVRT involving tAVNs may be one therapeutic option. Iatrogenic ventricular dyssynchrony after CA was never observed in our experience.

O6-3

3D transcatheter cryoablation of slow pathway in the definitive treatment of atrio-ventricular nodal reentry tachycardia in children: acute success obtained by a single focus cryolesion predicts a better outcome

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Introduction: Transcatheter cryoablation is an effective technique for the treatment of atrio-ventricular nodal reentry tachycardia (AVNRT) in children. Slow pathway conduction can be abolished by a single focus cryoablation or a linear cryo-lesion in the mid-septum performed from the ventricular side to the atrial side. Nevertheless, AVNRT may recur after a successful procedure. In this setting, 3-D mapping systems, besides reducing radiation exposure, can increase accuracy of a cryoablation, allowing to perform a real "focal cryoablation", defined as a single-point cryo-application plus one or more cryo-bonus on the same spot, or a real "high density linear cryo-lesion", defined as a linear lesion without any gap. At this regard, in this study, the outcome of "focal cryo-lesion" and "high-density linear cryo-lesion", performed using EnSite Velocity™ 3D Cardiac Mapping System, in eliminating slow pathways conduction, was compared.

Methods: We retrospectively reviewed the outcome of 67 consecutive pediatric patients (mean age 12.5 years; range: 5.4 to 17.8 years) who underwent cryoablation for AVNRT at our Institution from July 2013 to September 2014 using 3D Mapping System. Cryomapping was performed in the target site and when positive (i.e., no AH jump or non-inducibility of tachyarrhythmia), temperature was further lowered to create a permanent lesion. If a permanent result was not achieved after at least 3 focal cryoablations, a high-density linear lesion was performed delivering multiple (minimum of 4) and overlapped cryolesions from the ventricular side of the tricuspid annulus to the atrial side.

Results: The acute success rate was 100%: 35/67 (52.2%) with focal cryoablation and 32/67 (47.8%) with high-density linear lesion. There were no permanent complications. During the follow-up (mean 9.8 months), AVNRT recurrence rate was 0% (0/35) in patients treated by a focal cryoablation and 12.5% (4/32 $P < 0.05$) in those treated by a high-density linear lesion.

Conclusions: Our data seem suggest that the long-term success of 3D mapping-guided cryoablation for AVNRT in children are related to the cryoablation protocol (focal or high-density linear lesion) used to achieve the definitive elimination of the slow pathway conduction. In this setting a successful result obtained by a focal cryoablation seems to predict a better outcome.

O6-4

Catheter ablation of intraatrial reentry tachycardia in patients with congenital heart disease - efficacy and safety

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Introduction: Prevalence of intraatrial reentry tachycardia (IART) in patients with congenital heart disease (CHD) is significant during long-term follow-up and is associated with increased morbidity and mortality. Besides antiarrhythmic medication, catheter ablation has become the treatment strategy of choice.

Purpose: Single center study on efficacy and safety of IART ablation in a large cohort of patients with CHD over more than a decade.

Patients and Methods: In a total of 128 patients (25 children < 18 years) with CHD (88 patients with complex CHD) and IART, 197 ablation procedures were performed between 01/2003 and 11/2014. Median age at the first procedure was 32.4 (range 1.3 – 70) years. Mapping was performed with the non-contact mapping system and the NavX system, respectively. Radiofrequency current was delivered with irrigated tip catheters (7 F, 4 mm tip) in a temperature control mode (45 °C, 30 – 50 W, 45 seconds).

Results: After the first procedure, acute success was 78.1% (75.0% in patients with complex CHD and 85.0% in patients without complex CHD, n.s.). 45 patients underwent multiple ablation procedures. Follow up data were available from 118 patients. 76 patients (64.4%) were free from IART recurrence over a median of 21.5 (range 1 – 123) months. Median fluoroscopy time was 22.1 (IQR 15.0 – 23.7) min and median procedure duration was 260.0 (IQR 209.5 – 330.0) min. Major complications occurred in 4 patients. In 3 patients groin vessel damage required surgical revision. In one patient, stenosis of the right coronary artery after cavotricuspid isthmus ablation required angioplasty and stent implantation. There was no death and no permanent AV block. Complication rate was not related to age or complexity of CHD.

Summary and conclusions: Overall success of catheter ablation of IART in patients with CHD was 78.1% and was lower when compared with patients without CHD. A substantial number of patients needed additional procedures. Ablation procedures were safe with a low complication rate. New technologies as contact force measurement may increase efficacy of catheter ablation in this population.

O6-5

Resynchronization for chronically failing sub-pulmonary right ventricle in patients with congenital heart disease

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Objectives: Resynchronization of the sub-pulmonary right ventricle (RV CRT) is a potential therapeutic option for dyssynchronous RV cardiomyopathy.

Methods: Nine pts (age < 18 years, median = 6.8 years) with complete right bundle branch block and RV dysfunction by MRI and/or echocardiography after surgical repair of congenital heart disease were intended to be treated by RV CRT using atrial-synchronized RV free wall pacing in complete fusion with spontaneous activation.

Results: Temporary pacing was used to assess acute RV CRT effect as compared to spontaneous ventricular activation and carried significant decrease in QRS duration, septal to lateral RV mechanical delay by 2D strain (SL delay) with abolition of the pathological septal flash and increase in RV and left ventricular filling time (RVFT and LVFT, expressed as % cycle length) and pulmonary artery velocity time integral (VTI), see Table. Permanent RV CRT was applied in 2/9 pts (tetralogy of Fallot and absent

Table.

	QRS [ms]	Septal flash [N]	SL delay [ms]	RVFT [% CL]	LVFT [% CL]	VTI [cm]
Before	137 (32)	6/9	61 (19)	46.6 (6.7)	46.8 (5.6)	19.6 (10.1)
After	104 (13)	0/9	2 (36)	54.9 (10.2)	50.0 (5.1)	21.9 (11.4)
P	= 0.025	= 0.009	< 0.001	= 0.008	= 0.009	= 0.010

mean (SD)

pulmonary valve syndrome, resp.) with clinical symptoms of RV failure and major effect of temporary resynchronisation. Ventricular pacing leads were inserted at the site of late RV free wall activation (q-RV = 70 and 100% of QRS duration, resp.). Major lasting improvement in RV function, mechanical synchrony and functional class was achieved in both pts over a follow-up of 14 and 37 months, resp.

Conclusions: Electromechanical dyssynchrony may play a significant role in sub-pulmonary RV failure development and can be effectively corrected by RV CRT to achieve both acute and long-term improvement of RV function. (Supported by MH CZ - DRO, University Hospital Motol, Prague, Czech Republic 00064203).

O6-6

LV severe systolic dysfunction caused by LV free wall epicardial pacing

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Introduction: Long term right ventricular pacing induces left ventricular (LV) dyssynchrony that might evolve to LV remodelling and severe systolic dysfunction. Recent evidence suggests that pacing from the LV apex or LV free wall could prevent such deterioration. We present 2 cases of LV free wall pacing that caused a severe and rapidly progressing reversed pattern of LV dyssynchrony.

Methods: Retrospective study describing 2 neonates with complete heart block (CHB) who underwent LV free wall epicardial permanent pacing in our institution since 2012.

Results: Cardiac diagnosis included one patient with congenital complete heart block (case 1) and a postoperative complete heart block in a patient with transposition of the great arteries and VSD that underwent an arterial switch operation (ASO) and VSD closure (case 2). The baseline ECG and echocardiogram parameters are summarised in the Table 1 and 2. Both cases underwent an elective implant of a DDD pacemaker, via left thoracotomy with the ventricular lead placed on the LV free wall at 1 month of age, in Case 1 and 20 days old in Case2, 14 days after ASO. Both cases were discharged 5 days after implant with good LV systolic function.

6 months post implant, both infants presented with failure to thrive and signs of heart failure. ECG and ECHO data are summarised in Table 1 and 2. 2D strain analysis demonstrated the presence of a reversed pattern of LV dyssynchrony, with very early activation of the lateral segments of the LV compared with apical and septal segments. 2D Echo analysis demonstrated a rapid

deterioration of LV systolic function that correlated also with QRS prolongation of more than 30 ms over 6 months time. Upgrade to CRT-P was performed via mid-sternotomy with an implant of a new RV apical bipolar ventricular lead. The ECG and ECHO parameters 6 months post-CRT, showed a significant improvement of LV systolic function, LV reverse remodelling and shortening of QRS duration with normalization of LV synchrony in both cases.

Conclusions: LV free wall pacing might cause reversed pattern of LV dyssynchrony that can rapidly progress to severe systolic dysfunction and heart failure. Implant of LV epicardial leads should aim for LV apex segments.

O6-7

Thin, robust and selective: good acute and chronic performance of the worldwide thinnest bipolar pacing lead in patients with congenital heart defects

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Introduction: Transvenous cardiac pacing has become an increasingly applied therapeutic modality in congenital heart disease (CHD). In this highly selected entity patient age, size and anatomic variations pose extraordinary challenges to endocardial pacing which may be addressed by a new type of lead with unique design features. The lumenless SelectSecure™ model 3830 (Medtronic) has an outer diameter of only 1.4 mm (4.1 Fr), making it the world’s thinnest bipolar electrode. We report our acute and longterm experience with this fixed-screw, catheter-delivered pacing lead in a heterogeneous CHD population.

Methods: The majority of procedures (83%) were performed in conscious sedation. Venous access was obtained via subclavian puncture. Leads were either placed with steerable catheters (8.4 Fr) or in patients <20 kg with modified pre-shaped catheters (7 Fr) without the use of additional outer sheaths.

Results: 71 leads were successfully implanted in 47 patients with median age of 14 y (2-70) and median weight of 53 kg (10-114). 9 patients weighted below 20 kg. 78% of patients had previous CHD surgery. Follow up (FU) was 4.58 y (2 d-7.5 y). 93% of ventricular leads were placed in septal or RVOT position. Initial pacing thresholds were low. One early (1.5 mo) and two late dislocations (13 and 45 mo) occurred – all in the same patient. 6 electrodes were completely explanted in 4 patients, all during late FU and with sole traction force. All non-dislocated leads remained functional with only 2 showing moderately elevated PT’s >2 V. 7 patients dropped out due to deaths unrelated to pacemaker issues.

Conclusions: The 3830 pacing lead can be successfully implanted in the CHD population with good acute and longterm performance. With modified delivery techniques the access size can be downsized to 7 Fr to take full potential of the very slim design – even for patients below 20 kg. The catheter-delivery allows for visualization of the target region in patients with challenging anatomy and selective-site placement. The unique tensile strength allows improved extraction

Table 1. ECG parameters

	ECG baseline		ECG day + 1 LV paced		ECG pre CRT		ECG 6 months post CRT	
	HR	QRS (ms)	HR	QRS (ms)	HR	QRS (ms)	HR	QRS (ms)
Case 1	50	67	150	114	120	150	120	100
Case 2	47	87	120	123	130	147	119	122

Table 2. Echocardiographic parameters

	ECHO Baseline				ECHO pre CRT				ECHO 6 months post CRT			
	LVE Dd (Z score)	LVES d (Z score)	EF (%)	SPM WD (ms)	LVED d (Z score)	LVES d (Z score)	EF (%)	SPM WD (ms)	LVED d (Z score)	LVES d (Z score)	EF (%)	SPM WD (ms)
Case 1	2.2	1.8	66	17	3	2.2	39	65	1.8	1.6	63	37
Case 2	1.3	1.2	60	27	2.5	2	34	58	2	1.5	57	40

Table.

	Implantation		Last Follow up	
	Threshold	Sensing	Threshold	Sensing
atrial n = 43	0.7 ± 0.3 V (0.3-2.7)	3.05 ± 1.68 mV (1.4-10.0)	0.75 ± 0.38 V (0.25-2.4)	3.0 ± 1.72 mV (0.18-6.6)
ventricle n = 28	0.5 ± 0.22 V (0.25-1.3)	13.3 ± 6.76 mV (2.8-30.0)	1.0 ± 0.49 V (0.25-2.25)	12 ± 10.28 mV (1.8-31)

and thus facilitates growth adaptation. Unfortunately, the lead has not yet been labeled MRI conditional, which would be mandatory for a more widespread use.

O7-1

Intrinsic function of the systemic right versus the left ventricle of univentricular hearts in the Fontan circulation

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Intrinsic function of the systemic right versus the left ventricle of univentricular hearts in the Fontan circulation.

Introduction: The single right ventricle (RV) is thought to be less suitable to support the Fontan circulation compared to the left ventricle (LV). We sought to compare intrinsic systolic and diastolic function of the single RV and LV taking into account arterial elastance as a measure of afterload.

Methods: 81 single ventricle patients (59 RV with HLHS and 22 LV) were studied 5.6 years after Fontan operation using the conductance catheter technique.

Results: Ejection fraction (EF) of the single LV was higher compared to the RV. Arterial elastance (E_a) and intrinsic systolic function (end systolic elastance, E_{es} : preload recruitable stroke work, PRSW) were higher in the RV. Diastolic stiffness (end diastolic elastance, E_{ed}) of the RV was also higher (see table, mean \pm SD).

Conclusions: Intrinsic systolic function of the systemic RV is increased compared to the LV. This can be interpreted as a physiologic adaptation to elevated arterial elastance after reconstruction of the aorta during the Norwood operation in HLHS patients. Diastolic function of the single RV is already reduced early after Fontan completion, which is of concern for the long term outcome of these patients.

Table.

	RV	LV	P
EF, %	61 \pm 11	71 \pm 8	< 0.001
E_a , mmHg/ml	3.0 \pm 0.96	2.0 \pm 0.86	0.0001
E_{es} , mmHg/ml	3.4 \pm 2.5	1.8 \pm 1.3	0.002
PRSW, mmHg x ml	61.8 \pm 21.6	49.1 \pm 16.7	0.02
E_{ed} , mmHg/ml	0.60 \pm 0.37	0.32 \pm 0.18	0.0001

O7-2

Functional parameters of the Fontan circulation reflected in diffusion-weighted imaging of the liver

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Introduction: Patients with a Fontan circulation tend to develop liver fibrosis, cirrhosis and even hepatocellular carcinoma. Diffusion-weighted imaging (DWI) is an upcoming magnetic resonance technique for detecting and staging hepatic fibrosis and cirrhosis. We hypothesize that the liver reflects functional aspects of the Fontan circulation and that DWI provides a new non-invasive tool to evaluate the liver in time.

Methods: In a cross-sectional study, Fontan patients ($n = 59$) were evaluated by liver DWI. The association between apparent diffusion coefficients (ADC) and patient characteristics, laboratory measurements and functional aspects of the Fontan circulation (NYHA class, maximum oxygen uptake during exercise and cardiac index) was assessed.

Results: Liver ADC values were low ($0.82 \times 10^{-3} \pm 0.11 \times 10^{-3} \text{ mm}^2/\text{s}$) compared with literature values for healthy volunteers and correlated significantly with calculated liver fibrosis/cirrhosis scores (Fib-4 score, $p = 0.019$; AST/ALT ratio, $p = 0.009$), antithrombin ($p = 0.006$) and gamma-glutamyl transferase ($p = 0.031$). Furthermore, ADC values correlated negatively with follow-up duration ($p < 0.001$) and positively with functional aspects of the Fontan circulation (cardiac index, $p = 0.019$). No correlation between ADC values and exercise testing was found. In multivariate analyses, duration of the Fontan circulation was the strongest predictor of ADC values.

Conclusions: Evidence of fibrosis, measured by decreased ADC values, was present in Fontan patients and associated with the duration of the Fontan operation and functional aspects of the Fontan circulation. The DWI provides a safe instrument for liver monitoring in Fontan patients.

O7-3

The intra-cardiac blood flow kinetic energetics of the systemic right ventricle in patients with hypoplastic left heart syndrome

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The impact of diastolic flow energetics on cardiac functions remains unknown in patients with hypoplastic left heart syndrome (HLHS), however the many hemodynamic parameters of chamber and wall kinetics were reported to affect the cardiac performance in HLHS.

We investigated the intra-cardiac blood flow kinetic energy loss (EL) of systemic right ventricle in 15 patients with HLHS after Fontan procedure (mean age at 7.7 y), by using a novel imaging modality of Vector Flow Mapping (VFM) (Hitachi-ALOKA Ltd), compared to those of the left ventricle (LV) in 16 normal healthy children (mean age at 8.9 y).

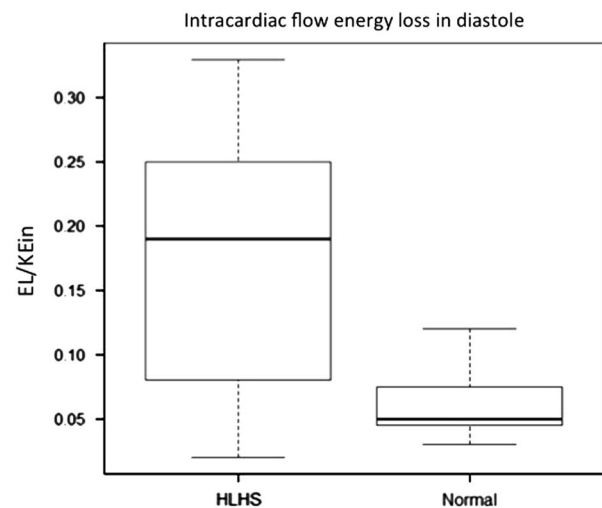


Figure 1.

Method: We first recorded three-chamber view of the systemic ventricle with color Doppler echocardiogram by Prosound F75 with VFM imaging mode. Then we calculated the blood flow kinetic energy loss (EL) from the reconstructed velocity vector components transformed into Cartesian coordinate system as previously reported by Itatani K. *Jpn J Appl. Phys* 2013;52:07HF16). The EL data were indexed by measuring a ratio of EL to the inflow kinetic energy (KEin) through systemic atrioventricular valve in diastole.

Result: EL/KEin was significantly increased in HLHS (0.17 ± 0.1) than in N (0.06 ± 0.03) as Fig. 1. ($p < 0.005$), although fractional area change was not significantly different in HLHS (0.42 ± 0.08) from in N (0.49 ± 0.04).

Conclusion: The systemic right ventricle of HLHS loses more flow kinetic energy in diastole than LV in N, which may contribute to cardiac performance in a long term besides the changes of other chamber and wall kinetics.

O7-4

Echocardiography and right ventricular function: Validation of functional criteria compared to in-vivo and ex-vivo contractility parameters

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Introduction: Right ventricular (RV) dysfunction is a major determinant of long-term survival in congenital heart diseases. Early echocardiographic detection of RV failure is mandatory, but recent parameters need to be validated. Objectives were to: (1) validate standard and strain echocardiographic parameters for evaluation of RV systolic function, compared to hemodynamic parameters; (2) assess the accuracy of these parameters for early detection of RV failure.

Methods: Combined RV overload as observed in repaired tetralogy of Fallot was surgically reproduced in 2-month-old piglets ($n = 6$). Age-matched piglets were used as controls ($n = 4$). RV function was evaluated at baseline and 4 months of follow-up by standard and strain echocardiographic parameters, compared to hemodynamic (conductance catheter). Sarcomere shortening and calcium transients were recorded in RV isolated myocytes (IonOptix). Contractile reserve was assessed by in-vivo (dobutamine $5 \mu\text{g}/\text{kg}$) and ex-vivo (isoprenaline 100 nM) β -adrenergic stimulation.

Results: 4 months after surgery, hemodynamic RV ejection fraction (FEVD) was significantly decreased (29.7% [26.2-34] vs 42.9% [40.7-48.6], $p < 0.01$), and inotropic responses to dobutamine were attenuated (contractile reserve $\Delta\text{Emax} = 51\%$ vs 193% for controls). On echocardiography FAC, TAPSE, S' peak and RV free wall longitudinal strain rate were significantly decreased and correlated with FEVD. Strain rate and S' peak were correlated with ΔEmax ($r = 0.75$ and 0.78 , $p < 0.05$). Isolated RV myocytes from operated animals exhibited hypertrophy, decreased sarcomere shortening peak in response to isoprenaline ($\Delta L = 7.8 \pm 2.8\%$ vs $10.7 \pm 2.9\%$, $p < 0.05$), and increased spontaneous calcium waves suggesting perturbations of calcium homeostasis.

Conclusion: In this model, both standard and strain echocardiographic parameters allowed the detection of early impairments of RV function and cardiac reserve, which are associated with cardiac excitation-contraction coupling alterations.

O7-5

Optical Coherence Tomography (OCT) in Patients with Regressed Coronary Aneurysm after Kawasaki Disease

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Background: Kawasaki Disease (KD) is an acute self-limited vasculitis of childhood. Coronary artery aneurysms (CAAN) are a serious complication of KD, and can lead to ischemic heart disease, myocardial infarction and sudden cardiac death. Regression of CAA occurs in 50% of the cases on follow up. Actual imaging techniques often describe these segments as normal, whereas studies have shown significant endothelial and smooth muscle dysfunction.

Method: KD patients scheduled for routine coronary angiography underwent OCT imaging between March 2013 and August 2014. The aim of the study was to compare microstructural coronary changes in coronary artery segments with no history of CAAN to segments with regressed CAAN, and segments with persistent CAAN.

Results: OCT was performed on 18 patients at 12.4 ± 5.5 years, 9.0 ± 5.1 years following onset of KD. Overall, 14/18 (77.7%) had a history of CAAN. Of those, 7/14 (50.0%) had regressed CAAN at time of OCT. Data was analyzed according to echocardiographic and angiographic progress of CAAN segments, with 18 segments having persistent CAAN, 11 regressed CAA and 13 with no history of segmental CAAN. All segments with persistent and regressed CAAN had significant intimal hyperplasia, compared to 1 with no history of segmental CAAN ($p < 0.001$). The importance of the intimal hyperplasia also differs according to the coronary artery status, amounting $442.9 \pm 176.7 \mu\text{m}$ for segments with persistent CAAN compared to $265.3 \pm 115.9 \mu\text{m}$ for segments with regressed CAAN and $88.8 \pm 93.3 \mu\text{m}$ for segments with no history of CAAN ($p < 0.001$). Also, partial disappearance of the media was found significantly more often in segments with persistent and regressed

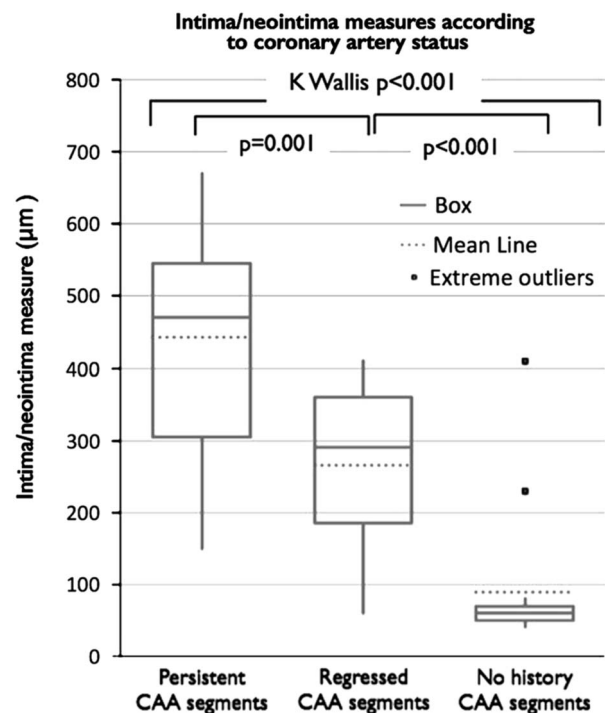


Figure.

CAA, compared to those with no history of CAA, in 15/18 (83.3%), 6/11 (54.5%) and 1/13 (7.7%) respectively ($p < 0.001$).

Conclusion: Despite normal angiographic features, regressed CAAN segments displayed significant microstructural changes, similarly to segments with persistent CAAN. The clinical significance of those changes is yet to be determined, but may present an increased risk of adverse coronary events. These findings demonstrate the potential incremental diagnostic value of OCT in the evaluation of patients following KD.

O7-6

Aortic coarctation pressure gradient prediction using a computational-fluid-dynamic model: Validation against invasive pressure catheterization at rest and pharmacological stress

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Introduction: Even after successful early repair of Aortic Coarctation (AoCo), life expectancy is still markedly reduced due to long term complications (hypertension) and often invasive diagnostic catheter investigations may be required to evaluate the pressure gradient (PG) across the aorta at rest, or unmask such gradients under pharmacological stress to mimic physical exercise. The objective of this research is to know if a MRI based computational fluid

dynamics (CFD) model protocol can accurately predict the pressure gradient in patients with AoCo.

Methods: The study population included 7 cases with aortic coarctation (mean \pm standard deviation; age 19.4 ± 4.6 years, weight 71.9 ± 17.1 kg), who had a previous combined MRI (CE-MRA and 2D CINE-PC) and cardiac catheterization study performed in rest and isoprenaline stress conditions. The 3D CE-MRA data was used to create geometric solid models of the aorta which were then discretized using a tetrahedral mesh generation program (MeshSim, Simmetrix, Clifton Park, NY). The 2D PC-MRI data were used to determine flow waveforms and distribution and to estimate the stiffness of the Aorta. The numerical method solved the Navier-Stokes equations for the flow of an incompressible Newtonian fluid within a deformable domain using a stabilized finite element formulation. Simulations were performed for rest and stress conditions. We compared the PG mean (PG mean-mean) and peak to peak (PG peak-peak) value of the pressure gradient obtained between Catheter and CFD using Bland-Altman and Wilcoxon Test.

Results: We have an average of PG mean-mean 2.85 ± 2.47 mmHg for the catheterization and 2.76 ± 1.64 mmHg for the simulation. For the PG peak-peak, we have an average between all cases of 10.36 ± 6.54 mmHg for the catheterization and 9.77 ± 6.39 mmHg for the simulation. In stress conditions we obtained an average of PG mean-mean of 12.59 ± 8.61 mmHg for the catheterization and 11.25 ± 7.60 mmHg for the simulation. The average PG peak-peak was 52.71 ± 22.11 mmHg for the catheterization and 37.38 ± 21.64 mmHg for the simulation. There were no significant differences between the catheterization and CFD except when comparing the PG peak-peak at stress.

Conclusions: The pressure gradients obtained at rest conditions were in good agreement with the ones obtained from catheterization demonstrating that we can use a systematic method to predict non-invasively pressure gradients using CFD simulation based on cardiovascular MRI.

O7-7

Transthoracic echocardiographic reference ranges for left ventricular, left atrial and aortic root M-mode dimensions in preterm infants

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Objectives: To derive reference ranges for left ventricular, left atrial and aortic root M-mode measurements in preterm infants. M-mode measurement is routine in transthoracic echocardiography. Reference ranges are well defined in adults and children. Calculation of Z scores (number of standard deviations from the mean) in children permits recognition of normal or abnormal change with growth over time. Available data in preterm infants is limited to small numbers analysed in categories rather than as continuous variables.

Methods: Retrospective audit of echocardiograms performed in preterm infants for clinical indications. All echocardiographic studies performed in Monash Newborn, between January 2005 and December 2010 were identified. Offline measurement of M mode variables, i.e. left ventricular systolic and diastolic dimensions (LVEDD and LVESD), left atrial and aortic root, and the wall thickness was performed. Infants with congenital heart disease or greater than a small patent ductus arteriosus were excluded. Sub-optimal studies were also excluded. Analysis was limited to infants with a birth or current weight ≤ 3.5 kg and ≤ 45 completed weeks

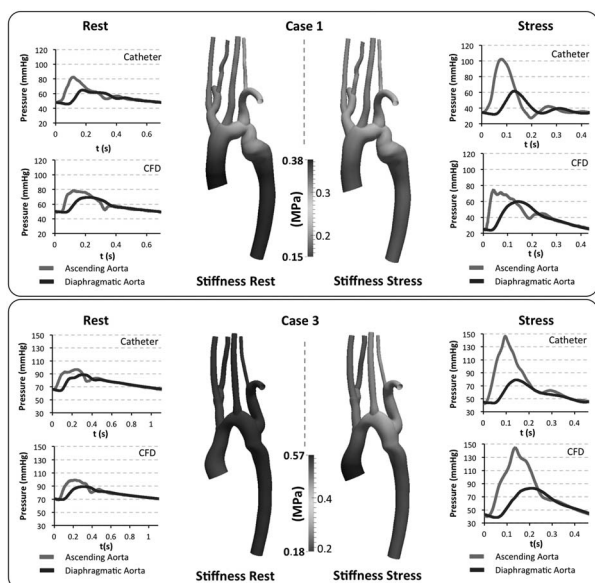


Figure.

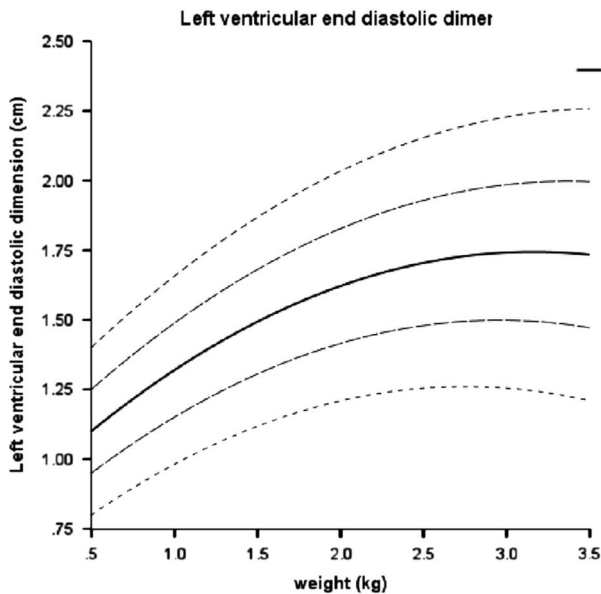


Figure.

of corrected gestational age (cGA) at the time of echocardiography. Regression curves were modelled with respect to weight and cGA and reference curves for ± 1 and ± 2 standard deviations calculated using the method of Altman (Statistics in Medicine 1993). This method allows for standard deviations which vary with weight or cGA.

Results: Two hundred and eighty studies were identified, mean corrected gestational age at study 32.5 ± 3.7 weeks (range 24 – 45 weeks), mean weight of the study group was 1.63 ± 0.7 kg (range 0.5 – 3.5 kg). Regression curves with standard deviations were modelled. Quadratic equations yielded the best fit for all parameters. Left ventricular systolic and diastolic dimensions increased with weight and cGA. Standard deviation was variable with both increasing weight and cGA, with respect to all the parameters measured. Graphical representation for LVEDD is presented below.

Conclusions: These data will permit calculation of z scores for left ventricular, left atrial and aortic root dimensions in preterm infants facilitating easier recognition of abnormalities and better description of change over time.

O8-1

Novel serumbiomarkers in early stages of hypertrophic cardiomyopathy in the young

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Introduction: Hypertrophic cardiomyopathy (HCM) is the most common monogenic cardiac disorder and the leading cause of sudden cardiac death in the young. Although in a majority of HCM cases there are gene mutations coding for sarcomere proteins, the clinical course is difficult to predict, as these mutations do not show any clear relationship to the degree of myocardial hypertrophy. Hence identification of early markers for this disease

is important. The aim of this study was to investigate novel serum biomarkers reflecting myocardial remodeling, microfibrosis and coronary endotheliopathy and cardiac magnetic resonance (CMR) in young presymptomatic HCM patients and HCM-risk individuals.

Methods: A cohort of 102 participants (mean age 15,9 years) consisting of HCM patients (n = 21), HCM-risk individuals (n = 16), healthy controls (n = 52) and young athletes (n = 13) were included in this study. All subjects underwent cardiac ultrasound (conventional and tissue Doppler imaging) and serum analysis for Myostatin, Cathepsin S, Endostatin, type I collagen degradation marker (ICTP), Matrix Metalloproteinase (MMP) 9, vascular (VCAM) and intercellular adhesion molecules (ICAM). In a subset of the study population (18 HCM, 9 HCM-risk, 9 controls including 4 athletes), myocardial perfusion was measured at rest and after adenosine vasodilation on cardiac magnetic resonance.

Results: The mitral annulus E/e' by tissue Doppler was decreased in both the HCM-risk and HCM group ($p < 0.05$), whereas global perfusion during adenosine was decreased only in the HCM group ($p < 0.05$) compared to other groups. MMP-9 ($p = 0.01$), VCAM ($p = 0.04$), Cathepsin S ($p = 0.008$) and Endostatin ($p < 0.0001$) were all increased in the HCM group compared to other groups. Both Cathepsin S and Endostatin showed weak correlation to left ventricular mass and E/e' ($p < 0.05$, $r > 0.3$ for both). Myostatin was decreased and ICAM was increased in the HCM-risk group ($p < 0.01$). ICAM correlated with myocardial perfusion during adenosine stress ($p = 0.04$, $r = 0.4$).

Conclusion: To the best of our knowledge, this is the first study to suggest early onset changes in biomarkers of myoblast regulation, endothelial function and matrix remodeling in young presymptomatic HCM patients and in HCM-risk individuals.

O8-2

Flow speckle tracking provides new information of complex blood flow in congenital heart disease

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Introduction: Blood flow patterns in the developing heart have been proposed to play a significant role in cardiac morphogenesis, and most congenital heart diseases (CHD) present with altered blood flow. There is a growing interest in understanding the detailed flow behavior in CHD, and this is now possible using high-end

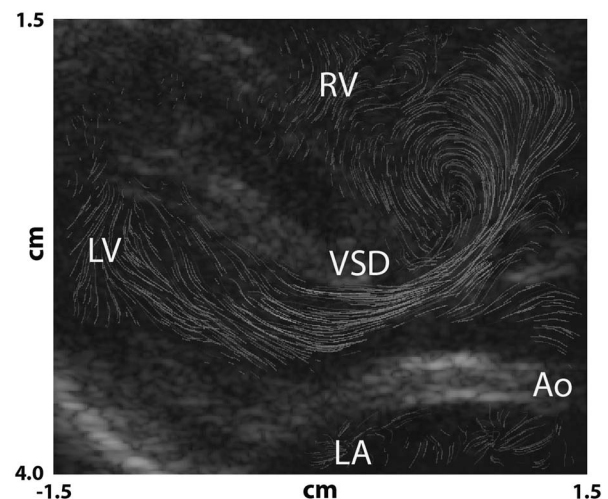


Figure.

ultrasound imaging equipment. In this feasibility study we demonstrate the potential of flow speckle tracking, a next-generation ultrasound imaging method which is able to visualize and analyze complex flow patterns without the need for contrast agents. We studied blood flow patterns in neonates with different types of CHD.

Methods: The examinations were performed using linear array transducers where a broad unfocused wave was transmitted and multiple image lines were generated simultaneously. Thus, the acquisition time was lowered and substantially higher frame rates were achieved. With frame rates in the kHz range, it is possible to utilize pattern-matching techniques to quantify the movement of the blood speckle. In this way, the velocity and direction of the blood flow can be calculated and shown as arrows or streamlines over the color-Doppler images, thereby highlighting areas of complex flow as shown in the image below.

Results: We subsequently examined 35 neonates with different CHD ranging from simple septal defects and valvular stenoses, to more complex heart defects and cardiomyopathies. In spite of the wider footprint of a linear probe, it was possible to obtain images from all standard views. With this new technique, we visualized and quantified vortex formations, for example in the right ventricle near ventricular septal defects (image below), circular flow near valvular stenoses and altered flow dynamics in ventricles with reduced function.

Conclusions: Flow speckle tracking provides new information of blood flow not available in the traditional color-Doppler images. To be able to detect and quantify complex flow patterns such as vortices in CHD, may be of clinical importance, since it is assumed that vortices play an important role in cardiac function. We hope that improved cardiovascular flow imaging can improve the understanding of both physiology and pathology in CHD. Further studies will explore if the method may improve diagnostics.

O8-3

Mutation Analysis of Pediatric Cardiomyopathies using Targeted Next Generation Sequencing: Identification of 7 Novel Mutations in Cardiomyopathy-Associated Genes

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Introduction: Pediatric cardiomyopathies are a heterogeneous group of disorders affecting the heart muscle and most of them being monogenic. A number of genes are known to cause cardiomyopathies. Clinical forms include hypertrophic cardiomyopathy (HCM), dilated cardiomyopathy (DCM), restrictive left ventricular cardiomyopathy (RLVC), arrhythmogenic cardiomyopathy, and left ventricular non-compaction. To date more than 50 genes have been defined associated with cardiomyopathies. Recently Next Generation Sequencing (NGS) allows the evaluation of large number of genes at an affordable cost. In this study we aimed to identify the genetic defects underlying pediatric cardiomyopathies using Targeted Next Generation Sequencing including 46 cardiomyopathy-associated genes.

Methods: Ten pediatric patients with cardiomyopathies (6 DCM, 2 HCM and 2 RLVC) were recruited from Pediatric Cardiology Subunit, Department of Pediatrics, Medical School Hospital, Ege University. A next-generation panel covering 46 cardiomyopathy-associated genes (ABCC9, ACTC1, ACTN2, ANKRD1, CASQ2, CAV3, CRYAB, CSRP3, CTF1, DES, DSC2, DSG2, DSP, DTNA, EMD, FHL2, GLA, JUP, LAMA4, LAMP2, LDB3,

LMNA, MYBPC3, MYH6, MYH7, MYL2, MYL3, MYLK2, MYOZ2, NEXN, PKP2, PLN, PRKAG2, RBM20, RYR2, SGCD, TAZ, TCAP, TMEM43, TNNC1, TNNI3, TNNT2, TPM1, TTN, TTR, VCL) were performed on these pediatric cardiomyopathy patients.

Results: A total of 7 mutations in 5 different genes were identified in 10 patients. All mutations were novel. Among these mutations five (C15003Y in TTN gene, IVS2 + 1 G > A in TPM1, I100T in TNNT2 gene, E115G in NEXN gene and R696L in MYB6 gene) were clearly disease causing while two mutations (K20028D and P29869S mutations in TTN gene) were considered to require further familial or functional studies for evaluating their functional effects. All novel mutations detected in this study were heterozygous with the exception of homozygous C15003Y mutation in TTN gene, which was detected in a patient born to consanguineous parents.

Conclusions: Targeted next-generation sequencing is an efficient, rapid and cost-effective technique for detecting mutations in genetically heterogeneous diseases such as cardiomyopathies. Identifying mutations in cardiomyopathy patients will help cardiologists to predict unforeseen difficulties, allowing detection of the relative risks and genotype-phenotype correlation.

O8-4

Biventricular Regional Fibrosis and Function During Chronic Right Ventricular Pressure Overload in a Rabbit Model of Pulmonary Artery Banding

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Introduction: Death and morbidity in pulmonary arterial hypertension are often due to right ventricular (RV) failure. Right ventricular failure is frequently associated with co-existing left ventricular (LV) dysfunction. However, biventricular fibrosis, remodeling and function during continuously elevated right ventricular afterload are poorly described.

Objective: To study RV and LV regional fibrosis in elevated right ventricular overload and its functional implications.

Methods: Elevated RV afterload was induced by progressive pulmonary artery banding in rabbits (PAB group, n = 14). Sham operated controls had the same operation without pulmonary artery banding (sham group, n = 6).

Results: RV and LV collagen content was increased with PAB compared to sham-operated controls. In the LV, increased fibrosis was seen in the free wall and especially at the septum and septal

Table.

[Median (range)]	Sham (n = 6)	PAB (n = 14)	p-value
Fibrosis			
RV free wall (%)	10.4 (6.4 to 16)	13.2 (5.4 to 41.7)	0.001
RV hinge point (%)	10.2 (4.8 to 20.6)	13.4 (4.8 to 32.6)	0.0005
Septum (%)	6.1 (3.1 to 8.3)	9.2 (3.2 to 25.8)	<0.0001
LV free wall (%)	7.2 (5 to 9.8)	8.4 (3.9 to 21.5)	<0.0001
LV hinge point (%)	8 (3.3 to 12.5)	10.4 (4.7 to 27.6)	<0.0001
LV Longitudinal Strain/SR			
Mean Strain	-11 (-18 to -9)	-10 (-15 to -7)	0.287
Mean SR (/second)	-2.93 (-5.9 to -1.8)	-1.9 (-4.2 to -0.7)	<0.0001
RV Longitudinal Strain/SR			
Mean RV Strain	-19 (-26 to -13)	-9.5 (-19 to -5)	0.009
Mean RV SR (/second)	-3.9 (-6.2 to 4.13)	-1.7 (-4.1 to -0.8)	<0.0001
Conductance			
RV Tau (/second)	7 (4 to 20)	28 (17 to 35)	0.011
LV Tau (/second)	24 (12 to 35)	20 (13 to 49)	0.978
RV Pes (mmHg)	18 (13 to 18)	27 (21 to 34)	0.009
LV Pes (mmHg)	45 (44 to 77)	45 (35 to 74)	0.467

hinge points (Table). RV function was worse in PAB; as seen by worsened RV Tau and TAPSE and reduced RV longitudinal strain and strain rate (SR). LV myocardial function assessed by longitudinal strain at the apical segment and the mean LV longitudinal strain rate were worse in the PAB group compared to shams.

LV ejection fraction, myocardial performance index and conductance catheter parameters were similar in PAB vs. shams.

In the RV these changes were associated with up-regulation of fibrosis signaling via pSMAD3. In both the RV and LV protein levels of apoptosis related enzymes (Caspase 3 and 8) were up regulated, whereas the fetal gene program showed reduced α -myocyte heavy chain protein levels linked with heart failure.

Conclusions: In increased RV overload, extensive fibrosis is found in all areas of the heart associated with up-regulation of protein levels of fibrosis signaling and apoptosis related enzymes. While the right ventricular function showed both an overall worsening with increased workload, and local longitudinal strain or strain rate in the right and left ventricle deteriorated.

O8-5

Slices of human neonatal and infantile myocardium: a novel experimental platform to study (patho) physiological properties at a cellular level

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Introduction: The current understanding of the neonatal and infantile human heart function at a cellular level has mainly been interpolated from experimental studies on animal and adult human heart tissue. This is mainly due to limited access to tissue and lack of suitable preparation techniques. For example, action potentials have not yet been recorded from neonatal and young patients' myocardium. To gain deeper insight into the physiological and pharmacological properties of young myocardium, we established a technique for the preparation of vital tissue slices from biopsies of patients with hypoplastic left heart syndrome (HLHS) and Tetralogy of Fallot (TOF).

Methods: The study has been approved by the local ethics committee and consent was obtained from all patients' parents. Tissue excised for implementation of a Sano shunt or during TOF correction was transferred into ice-cooled modified Tyrode's solution, transferred to our laboratory and embedded into low-melting agarose. 300 μ m thick slices were prepared using a vibrator. After induction of calcium tolerance and re-warming, slices were electrically stimulated and action potentials (AP) were recorded.

Results: Slices were obtained from 4 HLHS patients (mean age: 11.8 ± 7.0 days) and 4 TOF patients (mean age 178.7 ± 16.8 days). AP amplitude and maximal upstroke velocity (V_{max}) were higher in TOF (Amplitude: 110.5 ± 7.8 mV, $n = 24$ vs. 79.2 ± 19.3 mV, $n = 9$, $p = 0.001$, V_{max} : 79.2 ± 12.4 V/s vs. 118.5 ± 30 V/s, $p < 0.05$). Action potential duration was longer in TOF (APD₂₀: 137.0 ± 23.0 ms vs. 105.0 ± 36.3 ms, $p = 0.01$, APD₅₀: 252.4 ± 49.2 ms vs. 215.2 ± 75.2 ms, n.s., APD₉₀: 336.3 ± 65.0 ms vs. 296.9 ± 75.5 ms, n.s.). AP duration showed a strong frequency dependence in both groups (APD₅₀ at 1 Hz: 296 ± 86.5 ms, at 2 Hz: 252.4 ± 49.2 ms, at 3 Hz: 197.4 ± 39.8 ms, $p < 0.001$).

Conclusions: Vital heart slices can be prepared from patient biopsies during surgery for congenital heart disease and are a suitable

platform for physiological measurements at the cellular and multicellular level. This technique can be used for further physiological and pharmacological studies.

O8-6

Delayed brain maturation correlates with motor impairment at one year of age in children with severe congenital heart disease

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Objective: Neonates with severe congenital heart disease (CHD) are at risk for impaired neurodevelopmental outcome. Focal white matter injuries and small strokes can be detected on cerebral MRI before and after open heart-surgery. In addition, brain maturation may be delayed. It is unclear, how these changes relate to later neurodevelopmental outcome. We aimed on the relationship between brain maturation in the neonatal period and neurodevelopmental outcome at one year of age.

Design/Methods: We recruited 30 term-born neonates with severe CHD (d-transposition of the great arteries, single ventricle physiology, interrupted aortic arch). They underwent cerebral MRI before (median age 6 days, 1-12 days) and after (median age 26 days, 13-52 days) open-heart surgery (median age 15 days, range 7-62). At one year of age neurodevelopmental outcome was evaluated with the Bayley Scales of Infant Development III (BSID III). We also recruited 20 healthy term-born neonates. They underwent cerebral MRI at a median age of 23 days (range 13-33) Cerebral MRIs were analyzed using a standardized brain maturation score (TMS) (1).

Results: The median TMS of the preoperative MRI of the patients was 12 (10-14.5), of the postoperative MRI 13.3 (10.5-15.5). Controls had a higher median TMS of 13.4 (11-16), although not statistically significant when controlled for gestational age. The Cognitive Composite Score (CCS) of the BSID III at age one year was 104 (60-125), the Language Composite Score (LCS) 91 (65-132) and the Motor Composite Score (MCS) 90 (46-130). CCS and MCS were significantly lower compared to controls. In patients with severe CHD, the preoperative TMS correlated significantly with the BSID III Motor Composite Score ($R = 0.6$, $p = 0.003$, Spearman Rho bivariate correlation) at one year of age, but not with other developmental domains. In the controls, the preoperative TMS did not correlate with any outcome parameter.

Conclusions: Our results suggest that patients with delayed brain maturation before surgery are at higher risk for later motor impairment. These results offer new insights into possible mechanisms of brain injury and developmental impairments in this population.

Reference:

(1) Childs A et al. Am J Neuroradiol 2001

O8-7

Neurodevelopmental outcome in Fontan patients – is the Norwood procedure a prognostic factor?

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Objective: Patients with single ventricle physiology are prone to neurodevelopmental abnormalities. Complex surgery within

the neonatal period may be a risk factor for brain injury and neurodevelopmental impairment. Neurocognitive outcome at pre-school age was compared between Fontan patients who underwent a Norwood procedure and other single ventricle patients who did not require complex neonatal surgery.

Methods: Verbal IQ, performance IQ and full scale IQ were evaluated with the Hannover-Wechsler-Intelligence scale. The German "Kognitiver Entwicklungstest für das Kindergartenalter" (KET-KID), which is composed of a global scale for cognitive development, a verbal and nonverbal scale, was applied to assess cognitive functions. To identify potential risk factors for adverse neurodevelopmental outcome, patient and procedural variables were evaluated.

Results: Neurocognitive assessment was completed in 90 Fontan patients at a median age of 4.1 (3.5–7.1) years. 62 underwent a Norwood procedure, among the 28 remaining patients 16 had placement of a systemic-to-pulmonary artery shunt, 10 underwent pulmonary artery banding and 2 did not require any intervention in the neonatal period. Overall, IQ-scores and percentile ranks of the KET-KID were in the normal range and, except for the KET-KID verbal scale, did not differ between patients who underwent a Norwood procedure and those who did not (verbal IQ: 96 ± 14 vs. 92 ± 11 , $p=0.095$; performance IQ: 93 ± 10 vs. 92 ± 10 , $p=0.408$; full scale IQ: 93 ± 12 vs. 91 ± 10 , $p=0.177$; KET-KID global: 42 ± 29 vs. 33 ± 30 , $p=0.102$; KET-KID verbal: 47 ± 30 vs. 31 ± 30 , $p=0.016$; KET-KID nonverbal: 38 ± 27 vs. 37 ± 26 , $p=0.385$). Full scale IQ was below average in 14 (23%) of the Norwood patients compared to 10 (36%) of the remaining cases ($p=0.192$). Global KET-KID scores were below average in 17 (27%) and 10 (36%) patients ($p=0.427$), respectively. Gestational age, lower birth weight, smaller head circumference and increasing number of complications during staged palliation were associated with worse neurocognitive test results.

Conclusion: Neurocognitive outcomes of Fontan patients at pre-school age were in the normal range, but performance was lower compared to population norms. Surprisingly, the Norwood procedure was not associated with neurocognitive test results.

O9-1

Impact of pediatric heart transplant program on the outcome of dilated cardiomyopathy

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Introduction: Pediatric dilated cardiomyopathy (DCM) is a multifactorial, progressive disease. Heart transplantation (Htx) is an acceptable treatment option for children with end-stage heart failure resulting from DCM. Impact of introduction of a pediatric Htx program (2007) was analyzed.

Patients and Methods: 82 children (female/male:35/47, age at time of diagnosis:0–17 yrs, weight:2–105 kg) with DCM (LVDD > 2 SD and/or LV-FS < 30%) were enrolled between 2000 and 2014 in our center. Tachycardia-induced (6), pacemaker-related (4), anthracycline-induced (2) and muscular dystrophy associated (1) cardiomyopathies were excluded. During follow-up medical treatment, time from diagnosis to end-stage heart failure (HF) need for VAD therapy and htx results were examined.

Results: Myocarditis was confirmed in 10 cases (8 viral, 2 auto-immun, 10/69, 14.5%), 9 (90%), of them recovered, Htx was necessary in 1 case. Idiopathic DCM was diagnosed in 59 cases, 13 of them were familial (13/59, 22%). In the later group diagnosis was established at younger age (4.1 yrs versus 5.6 yrs). Medical treatment included ACEi (57/59, 97%), beta-blocker (44/59,

75%), aldosterone receptor antagonist (41/59, 69%), furosemide (50/59, 85%), digoxin (34/59, 58%) and anticoagulation (31/59, 52%). Antiarrhythmic drug and ICD implantation were rarely used (amiodarone 4/59, 7%, ICD 7/59, 12%). All pts with end-stage HF before the Htx program died. After 2007 27 pts required htx with/without VAD. Time from diagnosis to end-stage HF was 2.3 yrs (0–12 yrs) regarding all 27 idiopathic cases. Despite introducing Htx program, 4 pts died while waiting (4/27, 15%). 15 children were transplanted without VAD therapy (15/27, 55%), in 8 cases VAD therapy was necessary (8/27, 29%). 2 children died during VAD treatment (2/8), 6 were successfully transplanted (6/8). Among the total of 21 Htx patients 2 pts died in the post-transplant period.

Conclusions: 1. Before the introduction of Htx program all pts with end-stage HF died. 2. The progression of pediatric DCM is rapid. 3. Need for ICD implantation is very rare in childhood. 4. Since the introduction of Htx program almost all pts with end-stage HF were successfully transplanted. 5. VAD treatment is a necessary component of any pediatric Htx program. 6. Mortality after htx is acceptable.

O9-2

RV-Function after Senning-Procedure – a Conductance Study

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Aim: Declining RV-function is a major problem late after atrial switch repair in patients with d-transposition of the great arteries (d-TGA). The adequate therapeutic strategy remains unclear. Load independent indices of RV-function may be potentially helpful in decision making.

Methods: Between 2011–2013 16 patients with d-TGA underwent spiroergometry, cardiac MRI and invasive conductance studies 27.1 ± 6.7 years (21.3 – 46.1 years) after atrial switch repair.

Results: All patients have been in NYHA-Class I–II, they reached step 6 – 11 (median 8) during exercise test with O_2 -uptake of 30.1 ± 5.8 ml/min/kg (22.7 – 45.5 ml/min/kg). The mean end-diastolic RV-volume amounted to 110 ± 22 ml/m² (78 – 156 ml/m²), the endsystolic RV-volume to 66 ± 19 ml/m² (41 – 110 ml/m²), stroke volume to 44 ± 9 ml/m² (32 – 67 ml/m²) and cardiac index to 3.2 ± 0.8 ml/min/m² (2.1 – 4.6 ml/min/m²). The conductance study showed an increase of endsystolic elastance (Ees), enddiastolic stiffness (Eed) as well as arterial elastance (Ea) during dobutamin stress test (Ees from 0.80 ± 0.44 to 1.89 ± 0.72 mmHg/ml, Eed from 0.11 ± 0.07 to 0.13 ± 0.15 mmHg/ml, Ea from 0.97 ± 0.29 to 1.4 ± 0.45 mmHg/ml). A diastolic dyssynchrony ($36 \pm 6\%$) mainly in the apical segments was found in all patients, a systolic dyssynchrony ($22 \pm 10\%$) in 8 of 16 patients.

Conclusion: Although there is a remarkable reserve of contractility, all indices of RV-function are in the range of a failing left ventricle. Perhaps the course of RV-function could be better estimated by the extent of dyssynchrony, which started as disturbed diastolic function and may end as disturbed systolic function.

O9-3

Outcome after surgical repair of Ebstein's anomaly in adulthood

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Objective: To review single-institutional early and mid-term results of surgical repair of Ebstein's anomaly in adults.

Methods: The records (functional and echocardiographic parameters, surgical techniques, adverse events) of consecutive pts undergoing repair of Ebstein's anomaly from 2005 to 2014 were retrospectively reviewed with a median follow-up of 2.8 yrs.

Results: A total of 42 pts were operated at a median age of 33 (range 18–67) yrs with 6/42 having had a preceding cardiac procedure. Tricuspid valve plasty (TVP) and tricuspid valve replacement (TVR) were performed in 20 and 21 pts, resp. Additionally bidirectional cavo-pulmonary anastomosis was performed in 8 pts, an arrhythmia procedure in 17, mitral valve repair in 2 and coronary artery bypass graft in 1 pat. Four pts were re-operated (reTVR in 3 and TVR after TVP in 1 pat) yielding a reoperation-free survival of 82.6% at 5 years. There were 2 early and 2 late deaths with a 5-years survival probability of 83.1%. In pts undergoing TVP the proportion of significant tricuspid regurgitation (>2nd degree) decreased from 20/20 to 6/20 after repair ($P < 0.001$). Functional classification improved significantly with 27/42 pts having had marked or severe limitation (NYHA class > II) before surgery and only 6/42 at last follow-up ($P < 0.001$).

Conclusions: Surgical treatment of Ebstein's anomaly in adulthood can be performed with acceptable early and late mortality and freedom from re-operations. Significant improvement in both tricuspid valve function and functional capacity is achieved in the majority. Associated arrhythmia procedures are frequently needed. (supported by MH CZ – DRO, University Hospital Motol, Prague, Czech Republic 00064203 and by MH CZ – DRO, Nemocnice Na Homolce – NNH, 00023884).

O9-4

Maternal and fetal outcomes of pregnancy with Fontan circulation: a multicentric study of 59 pregnancies

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Objectives: Despite serious long-term sequelae, more and more women with Fontan palliation reach childbearing age. However there is paucity of data on the mid to long-term pregnancy outcomes and management of this condition. We sought to determine maternal and fetal outcomes of pregnancy in women with Fontan palliation.

Methods: This multicentric, retrospective study included women with Fontan circulation followed in 13 French centers from January 2000 to June 2014. All pregnancies were reviewed, including miscarriages, abortions, premature and term births. We reviewed maternal and fetal outcomes.

Results: Thirty-seven patients had 59 pregnancies. Mean age was 27 ± 5 years at first pregnancy. There were 16 miscarriages (27%) and 36 live births with 1 twin pregnancy. Cardiac events occurred in 10 (17%) pregnancies, with no maternal death. The most common cardiac complication was atrial arrhythmia, which occurred in 3 women. Prior atrial arrhythmia was a significant predictor of atrial arrhythmia during pregnancy ($p = 0.03$). Hematological complications including thromboembolic/hemorrhagic events occurred in 5 women antepartum, and 4 women postpartum. There was a high incidence of prematurity ($n = 25/36$, 69%), with a mean gestational age of 34 weeks and a mean birth weight of 1980 g (10th percentile). Anticoagulation therapy, regardless of medication or dose, was significantly associated with neonatal events ($p < 0.01$). In contrast there were more live births in women receiving low molecular weight heparin. After a median follow-up of 24 months, there was no significant worsening of clinical status/cardiac function/worsening thromboembolic disease noted.

Conclusions: Women can successfully complete pregnancy with Fontan circulation. Although there is an increase in cardiac and hematological events during pregnancy, there is no long-term impairment due to the pregnancy of the single ventricle function or the Fontan circulation, and there is no maternal death. At least prophylactic anticoagulation is recommended during pregnancy and postpartum period to prevent thromboembolic complications.

O9-5

Riociguat treatment for pulmonary arterial hypertension associated with congenital heart disease: a subgroup analysis from the PATENT studies

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Introduction: Riociguat treatment has shown beneficial effects in patients with pulmonary arterial hypertension (PAH) in a randomized, placebo-controlled Phase III trial (PATENT-1) and its open-label long-term extension (PATENT-2). Here we investigated the safety and efficacy of riociguat in the subgroup of patients with persistent or recurrent PAH following complete surgical repair of congenital heart disease (CHD) in PATENT-1 and PATENT-2.

Methods: In PATENT-1, treatment-naïve or pretreated patients with PAH received either placebo, riociguat up to 2.5 mg three-times daily (tid) (2.5 mg–maximum group), or riociguat up to 1.5 mg tid (1.5 mg–maximum group; exploratory) for 12 weeks. Patients completing PATENT-1 without ongoing drug-related serious adverse events (SAEs) were eligible to enter PATENT-2, in which all patients received open-label treatment with riociguat. **Results:** There were 35 patients with persistent or recurrent PAH after complete surgical repair of CHD in PATENT-1, of whom 33 (94%) entered PATENT-2. At PATENT-1 baseline, 57% of patients with PAH-CHD were treatment-naïve, most had atrial or ventricular septal defects (40% and 34%, respectively), all were in WHO FC II or III (60% and 40%, respectively), and mean time since last corrective surgery was 16.8 years. At Week 12 of PATENT-1, patients in the riociguat 2.5 mg–maximum group showed greater improvements from baseline in 6MWD (primary endpoint) and a range of secondary efficacy endpoints compared with patients receiving placebo (Table 1). Improvements in efficacy endpoints were also seen in the riociguat 1.5 mg–maximum group. Four (11%) patients reported SAEs during PATENT-1; none were considered drug-related. One patient died owing to right ventricular failure and worsening PAH (riociguat 1.5 mg–maximum group; not considered drug-related) and two patients discontinued treatment; one owing to supraventricular tachycardia and hypotension (riociguat 2.5 mg–maximum group; considered drug-related) and one owing to pneumothorax (placebo group; not considered drug-related). In the PATENT-2 open-label extension (median treatment duration 139 weeks), the improvements in 6MWD and WHO FC observed in PATENT-1 persisted for up to 2 years.

Conclusions: In PATENT-1 and PATENT-2, riociguat improved a range of clinical outcomes in patients with persistent or recurrent PAH after complete surgical repair of CHD.

Table 1. Change from baseline to Week 12 in PATENT-1 primary and secondary efficacy endpoints in patients with persistent or recurrent PAH after complete surgical repair of CHD (observed values).

	Riociguat 2.5 mg- maximum		Riociguat 1.5 mg- maximum		Placebo	
	n	Change from baseline	n	Change from baseline	n	Change from baseline
6MWD (m)	13	+39 ± 60	7	+43 ± 54	12	0 ± 42
Pulmonary vascular resistance (dyn·s·cm ⁻⁵)	13	-250 ± 410	7	-126 ± 368	11	-66 ± 632
NT-proBNP (pg/mL)	12	-164 ± 317	6	-872 ± 1147	12	-46 ± 697
WHO FC, improved/stabilized/ worsened (%)*	14	21/79/0	7	29/71/0	12	8/83/8
Borg dyspnea score	13	-0.3 ± 1.3	7	-0.8 ± 0.8	12	-0.1 ± 2.4
EuroQol Group 5-Dimension self- report questionnaire score	14	+0.03 ± 0.18	7	+0.09 ± 0.14	12	-0.05 ± 0.22
Living with pulmonary hypertension score	14	-8.0 ± 15.9	7	-13.7 ± 13.2	12	-0.1 ± 15.8

Data are mean ± SD unless otherwise indicated.

*Percentages may not add up to 100% due to rounding.

O9-6

Matrix Metalloproteinase-2 Is Associated With Cardiac Function in Adult Congenital Heart Disease

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Introduction: Matrix metalloproteinases (MMPs) and their tissue inhibitors (TIMPs) are a family of extracellular proteolytic enzymes, which play a key role in diverse extracellular matrix remodelling processes. In acquired heart disease, changes in MMP profiles have been associated with diastolic dysfunction, left ventricular (LV) hypertrophy and right ventricular (RV) dilatation. However in adult congenital heart disease (CHD), MMP profiles have never been described.

Methods: In this cross-sectional study, serum MMP-2, MMP-3, MMP-9, TIMP-1 values and echocardiographic measurements were prospectively obtained in consecutive adults with stable CHD seen at the outpatient clinic. Exercise testing results were collected if performed as routine clinical care. Log transformed biomarker values were linearly regressed against echocardiographic and exercise data, adjusted for age, gender, body surface area (BSA) and diagnosis.

Results: We included 425 patients (median age 33 [26-42] years, 57% male, 91% in NYHA class I) with tetralogy of Fallot

Table. ¹⁰log MMP-2 values related to clinical variables (n = 425); *p < 0.05, **p < 0.01.

Clinical variable (exercise testing, echocardiography or blood biomarker)	Mean change in clinical variable per 10-fold increase in MMP-2	p-value
Maximum heart rate (% of target)	-5.21	0.039*
Maximum workload (% of target)	-12.06	0.006**
LV end-diastolic dimension (mm)	-0.7	0.399
Aortic sinus diameter (mm)	1.2	0.182
Interventricular septum (mm)	0.0	0.995
RV end-diastolic basal dimension (mm)	2.3	0.039*
RV end-diastolic area (cm ²)	3.28	0.025*
LV ejection fraction (%)	-1.80	0.368
LV systolic dysfunction (eyeballing 0-1)	0.9	0.065
RV fractional area change (%)	-3.81	0.035*
RV systolic dysfunction (eyeballing 0-1)	0.8	0.034*
TAPSE (mm)	0.3	0.676
LV E/A ratio	0.03	0.745
LV deceleration time (ms)	-22.6	0.008**
¹⁰ log NT-proBNP (pmol/L)	0.143	0.010**

(n=164), congenital aortic stenosis (n=115), systemic RV (n=78), aortic coarctation (n=37), arterial switch operation (n=18) and other (n=13). MMP and TIMP values did not substantially differ among these diagnoses. Associations between MMP-2 values and clinical variables are tabulated. MMP-3 was only related to LV deceleration time (β = -22.5, p = 0.025); no associations were found for MMP-9 and TIMP-1 values.

Conclusions: In our diverse cohort of adults with CHD, increased MMP-2 was independently related to decreased exercise capacity, RV dilatation, decreased RV systolic function, decreased LV diastolic function and increased NT-proBNP. Our data therefore suggest that MMP-2 may serve as a new biomarker in this setting, in contrast to MMP-3, MMP-9 and TIMP-1. Further research is needed to determine the diagnostic and prognostic value of MMP-2 in clinical practice.

O9-7

Impact of type of repair on exercise capacity in patients with atrioventricular discordance

Does the anatomical repair have the advantage over the functional repair? Ohuchi H., Negishi J., Noritake K., Sasaki O., Hayama H., Taniguchi Y., Miyazaki A., Yamada O., Kagisaki K., Ichikawa H.

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Introduction: Anatomical repair (AR), i.e., double switch operation, has been applied to patients with atrioventricular discordance (AVD) due to possible disadvantages of the morphological right ventricle and tricuspid valve for the systemic circulation. However, there has been no data demonstrating clear clinical benefits of AR over conventional functional repair (FR).

Objectives: This study was to find determinant factors of exercise capacity in AVD patients and to see whether AR was advantageous over FR in terms of the exercise pathophysiology.

Method and Results: We measured peak oxygen uptake (PVO2: % of normal) in 99 AVD patients (42 after AR, 37 after FR, 20 corrected transposition of the great arteries without repair [cTGA]) during cardiopulmonary exercise testing (CPX). Of those, serial CPXs were performed to determine a rate of change in PVO2 (dPVO2/year) in 30 AR, 22 FR and 10 cTGA patients with an interval of 10 ± 5 years. Although PVO2 (%) tended to be higher in the AR than the FR group (67 ± 19% vs. 58 ± 19%, p = 0.067), those PVO2 were lower than that (85 ± 13%) of the cTGA (p < 0.01). In all subjects, age at CPX, a number of cardiac surgeries, use of diuretics and heart rate reserve (HRR), instead of the type of repair, independently determined PVO2 (p < 0.05 to 0.0001). In the serial study, the first to latest values of PVO2 for the AR, FR, and cTGA was 65 ± 18 to 67 ± 17, 62 ± 16 to 59 ± 18, 90 ± 11 to 90 ± 15, respectively (p < 0.0001) and there was no difference in the dPVO2/year among the 3 groups (p = 0.80). Male gender and change in HRR, rather than type of repair, were independently associated with the dPVO2/year (p < 0.05) and HRR was independently determined by a number of cardiac surgeries and pacemaker implantation (p < 0.01).

Conclusions: AR for AVD patients had a marginal benefit on the exercise capacity and its follow-up change. Greater number of cardiac surgeries, lower HRR, and use of diuretics, rather than the type of repair, were major determinants of lower exercise capacity in AVD patients.

O10-1

The German National Register for Congenital Heart

Defects: A clinical register that can serve as an important basis for studies on rare diseases

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Introduction: Congenital heart defects (CHD) represent the most frequent congenital disease in humans. They are characterised by a high variability of different heart malformations, of which each single group is extremely rare. Due to this, multicenter research is necessary to achieve significant results. For this purpose, a nationwide clinical register has been initiated by the German societies of paediatric cardiology, cardiology and heart surgery.

Methods: The National Register for Congenital Heart Defects is an individual-related clinical register featuring a broad informed consent given by its participants. Congenital and acquired cardiac diagnoses, as well as courses of disease, operations and catheter based interventions are recorded on the basis of medical reports. Coding takes place according to the International Paediatric and Congenital Cardiac Code. A central database system facilitates contacting patients and relatives for surveys and studies at any time. The range of diagnoses of currently 32,079 living patients is presented here.

Results: The average age is 15.6 years (SD = 13.1, minimum < 1 year; maximum 94 years). The sex distribution is well-balanced (16,182 [50.4%] male; 15,897 [49.6%] female). The distribution of main diagnoses approximates prevalence data as assumed according to available literature. Thus, representative studies are feasible.

Among the secondary diagnoses, chromosome anomalies such as Down's syndrome, Turner syndrome and DiGeorge syndrome stand out. Preterm delivery and Eisenmenger's syndrome can be cited as significant concomitant features.

Conclusions: The clinical register comprises data on a variety of patients with CHD. These data can be used as a basis for epidemiological, clinical and genetic studies. Due to its considerable extent and thus the option to reach unusually high patient numbers for even rare diagnoses, the register also represents a unique platform for research projects beyond Germany.

Table. Secondary diagnoses and concomitant features of recorded cases of CHD

Main cardiac diagnosis*	n = total	VSD	ASD	TOF	AV	CoA	SV	PV	AVSD	TGA	PAD	HLHS	Other
	32,079	9,281	5,021	2,669	2,229	2,143	1,992	1,709	1,836	1,462	1,070	463	2,404
Trisomy 21	1,369	352	129	53	7	11	8	3	753	0	30	0	23
Turner syndrome	51	2	6	0	13	23	0	0	2	0	0	2	3
DiGeorge syndrome	214	28	4	88	2	38	46	3	2	0	0	0	3
Premature infant	1,372	431	296	106	45	66	64	93	33	35	117	15	71
Eisenmenger's syndrome	132	52	1	10	0	4	19	1	29	2	10	0	4

*VSD = ventricular septal defect; ASD = atrial septal defect; TOF = tetralogy of Fallot; AV = aortic valve malformation; CoA = coarctation of the aorta; SV = single ventricle; PV = pulmonary valve malformation; AVSD = atrio-ventricular septal defect; TGA = transposition of the great arteries; PAD = persistent arterial duct; HLHS = hypoplastic left heart syndrome

O10-2

The changing epidemiology and outcome in hypoplastic left heart syndrome in Sweden. Results of a national cohort study 1993-2013

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Introduction: In Sweden pediatric cardiac surgery was centralized in 1993 to Lund and Gothenburg and Norwood surgery for HLHS has been offered in both centers since 1993. Since then postoperative survival and prenatal detection rate has gradually increased.

Objective: To describe the changing epidemiology and outcome in hypoplastic left heart syndrome (HLHS) on a national level.

Methods: This is a retrospective national cohort study covering all of Sweden. Registry data were collected on all patients with HLHS undergoing Norwood surgery 1993-2013 and on all live-borns with HLHS for the period 1997-2010. The prevalence at 18 weeks gestation was calculated from postnatal and fetal registry data from the western part of Sweden and used as an estimate on the national level. A diagnosis of HLHS required all of the following: 1/mitral atresia and/or aortic atresia, 2/hypoplasia of the ascending aorta, 3/severe hypoplasia of the left ventricle, 4/no additional cardiac defects. No cases were lost to follow-up.

Results: 139 patients with HLHS had Norwood surgery 1993-2013. 79/139 (57%) were alive in August 2014. Transplantation-free survival increased for each 5 year period ($p = 0.0011$) and the 5-year postoperative survival probability for patients born 2008-2013 was 88% for boys and 73% for girls. Significant risk factors for death were lower birth weight ($p = 0.0023$) and female sex ($p = 0.0013$).

The prevalence at 18 weeks gestation was estimated at 20/100,000. Based on this there were 280 fetuses with HLHS in Sweden 1997-2010, 141 were born alive and 95 were operated. The prenatal detection rate increased from 37/90 (41%) 1997-2001 to 69/89 (78%) 2007-2010, the proportion who were born alive decreased from 61/90 (68%) to 29/89 (33%) while the proportion of live-borns undergoing surgery increased from 38/61 (62%) to 24/29 (83%). The proportion alive in August 2014 was 15/90 (17%) 1997-2001 and 19/89 (21%) 2007-2010.

Conclusion: The increasing proportion of terminations because of HLHS is counterbalanced by an increasing proportion of live-borns undergoing surgery and by an improving postoperative survival resulting in a largely unchanged number of survivors. These results may have implications for prenatal counselling and for planning of postnatal resource utilization.

O10-3

Serial non-invasive blood pressure measurements in neonates of mothers with early-onset preeclampsia: a case control study

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Introduction: Preeclampsia is associated with increased long-term risk of hypertension and cardiovascular disease in the mother and child. Early onset preeclampsia is considered to be a consequence of fetal disorder, has distinct cardiac and vascular characteristics and may thus represent a separate disease entity. Early onset

preeclampsia is diagnosed before 34 weeks of gestation and carries the greatest risk of long-term cardiovascular sequelae. However, there is paucity of data concerning blood pressure (BP) values in the offspring during the neonatal period.

Methods: BP was measured with an oscillometric monitor at birth, daily during the first week of life and then weekly up to discharge from the Neonatal Unit in 68 neonates of mothers with early-onset preeclampsia and in 68 neonates of normotensive mothers matched for gender, gestational age and twinning.

Results: The two groups did not differ regarding maternal smoking status, diabetes or antenatal corticosteroid exposure, whereas offspring of mothers with early-onset preeclampsia had lower mean birth weight SDS-scores (-0.78 ± 0.77 versus -0.32 ± 0.86 , $p = 0.001$). Repeated measures ANOVA showed that both early onset preeclampsia and postnatal age, as expected, had a significant effect on systolic and diastolic BP during the first 4 weeks of life after adjusting for birth weight SDS-score ($p = 0.003$ and $p < 0.0001$, respectively). Paired comparisons showed no difference between the groups in systolic and diastolic BP at birth and the 1st day of life, whereas neonates of preeclamptic mothers had higher systolic and diastolic BP values at all other time points that BP was measured.

Conclusion: Our findings are in line with other studies that have reported a positive association between hypertensive disorders of pregnancy and offspring BP. We demonstrated that neonates whose mothers developed early onset preeclampsia have increased BP values and documented that these differences are detectable since the first days of life, prior to exposure to postnatal environmental factors that are likely to influence BP. The lack of difference in BP measurements during the first 24 hours of age may be attributed to the adaptation process to extrauterine life or maternal medication exposure.

O10-4

Feasibility of pulse oximetry screening for critical congenital heart defects after home births and early discharge in the Netherlands: a prospective study

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Introduction: Studies have shown evidence for universal neonatal screening for critical congenital heart defects (CCHD) with pulse oximetry (PO). However, the feasibility of CCHD screening after homebirth is unknown. We assess the feasibility of PO screening in the Netherlands, where there is a high percentage of homebirths and early discharge after delivery in hospital.

Methods: From October 2013 a feasibility study was performed in the Leiden region. At home or in hospital pre and post ductal SpO₂ are measured ≥ 1 hour after birth in term low-risk infants using Nellcor PO [Figure 1]. The measurement is repeated at day 2 or 3. Infants with positive screenings are assessed at the pediatric department and echocardiography is performed in case of persistent abnormal SpO₂. An acceptability questionnaire was sent to the mothers and screeners were asked if the screening should be implemented universally.

Results: 3059 infants have been screened (99% of infants of whom parents consented). In 33 infants screening was positive, of which 6 were not recognized and not referred. In 16 positive referred screenings, we detected 5 non-critical CHD, 5 infants with persistent pulmonary hypertension or wet lung, 1 meconium aspiration, 2 polycythaemias, 3 infants received sepsis therapy. In 9 infants

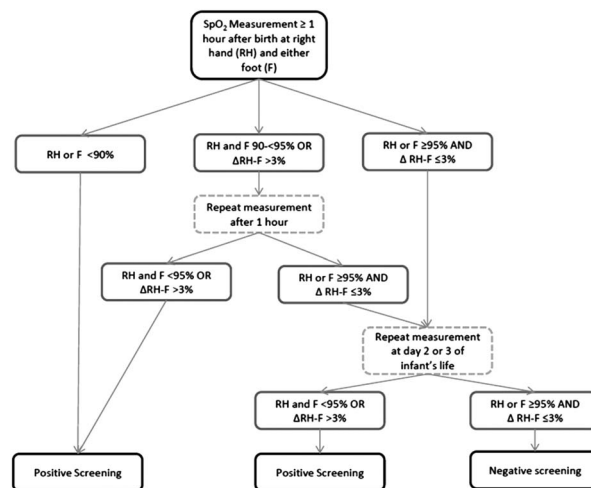


Figure 1.

measurements were normal in the hospital. No CCHD was detected and there were no false negative screenings. Median (IQR) pre and post ductal SpO₂ in the first hour of life was 99% (98-100) and 99% (97-100) respectively ($n = 382$). Of the responding mothers 69% would absolutely recommend the test to others (25% probably, 6% neutral, 1% probably not, 0% absolutely not). 82% of the screeners think that the screening should be universally implemented in the Netherlands (13% neutral, 5% disagree).

Conclusions: This is the first European pilot study assessing CCHD screening in a country with home birth and early discharge from hospital. In this setting CCHD screening is feasible and detects potential life-threatening pathology in newborns. Median pre and post ductal SpO₂ in the first hour of life is already 99%. Most mothers would recommend the test to others and the majority of screeners want the screening to be universally implemented in the Netherlands.

O10-5

High prevalence of low serum 25-hydroxyvitamin D levels and secondary hyperparathyroidism in Fontan patients: need for increased surveillance of vitamin D status

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Background: Suboptimal vitamin D status may negatively affect many organ functions. Limited data exist on the vitamin D status in Fontan patients. The aims of this study were to determine the prevalence of low serum 25-hydroxyvitamin D ($\geq 25[\text{OH}]\text{D}$) levels among Fontan patients, and to explore potential risk factors for vitamin D deficiency. **Methods:** A retrospective chart review was performed of Fontan patients who had been screened for vitamin D status. Vitamin D deficiency was defined as a $\geq 25(\text{OH})\text{D}$ level of < 20 ng/ml. The neutrophil-to-lymphocyte ratio (NLR), a marker of systemic inflammation, was calculated in all patients. Associations between laboratory measurements and patient characteristics were explored.

Results: Data were collected from 27 Fontan patients (55.6% male, mean age 8.1 ± 5.3 years). Protein-losing enteropathy (PLE) was diagnosed in 6 patients (22.2%). Mean $\geq 25(\text{OH})\text{D}$ level was 14.1 ± 10.4 ng/mL. Vitamin D deficiency was found in 19/27 patients (70.3%), and a severe vitamin D deficiency (< 10 ng/mL) was observed

in 12/27 patients (44.4%). s25(OH) D levels were not significantly lower in PLE patients. Patients with increased skin pigmentation had significantly lower s25(OH)D levels (7.9 ± 5.5 ng/mL vs 16.7 ± 5.5 ng/mL, $p = 0.04$). No further parameters were associated with low s25(OH)D. Hyperparathyroidism was present in 5/21 (23.8%) patients, and was more prevalent in PLE patients (PLE: 4/5 patients, 80.0%; vs non-PLE: 1/16 patients, 6.3%; $p < 0.001$). Interestingly, circulating parathyroid hormone correlated with parameters of systemic inflammation (NLR: $r = 0.484$, $p = 0.026$; relative lymphocyte count: $r = -0.635$, $p = 0.002$). Vitamin D supplementation significantly increased s25(OH)D levels ($p < 0.0001$), and was accompanied by a reduction in parathyroid hormone concentration ($p = 0.032$).

Conclusions: A high prevalence of vitamin D deficiency was found among Fontan patients, independent of age, time after Fontan procedure, ventricular morphology, and presence of PLE. A potential important link between parathyroid hormone and systemic inflammation is suggested. Further studies are needed to elucidate the clinical role and mechanisms underlying vitamin D deficiency in Fontan patients.

O10-6

Human Leucocyte Antigen, Infections and Systemic Inflammatory Biomarkers in Early Atherosclerosis in Children and Adolescents with Type 1 Diabetes

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Background: This prospective study focuses on factors associated with arterial damage in children with type 1 diabetes (T1D).

Materials and Methods: Eighty children and adolescents with T1D (mean age 15, range: 8–20 yrs; mean diabetes duration 7, range: 0.5 to 19 years) were investigated twice, approximately 2 years apart, for carotid artery intima-media thickness (cIMT) and compliance (CAC), flow-mediated dilatation (FMD) of the brachial artery, and plasma levels of matrix metalloproteinase (MMP)-8. HLA genotypes were determined in dried spots of peripheral blood by polymerase chain reaction followed by hybridization assay. The number of respiratory tract infections (RTI) during the past year was obtained by a questionnaire in 56 patients.

Results: cIMT progression (% change of cIMT from baseline) correlated inversely with the % changes of both CAC ($p = 0.04$, $r = -0.3$, $n = 62$) and FMD ($p = 0.03$, $r = -0.3$, $n = 67$). RTI frequency correlated significantly with cIMT progression irrespective of age, diabetes duration, BMI, and HbA1c ($p = 0.03$, $r = 0.3$, in multivariate analysis). When patients were divided in relation to DQ2/8 genotype and RTI, the association of DQ2/8 with cIMT progression remained significant in patients with over three infections/year ($p = 0.04$, $r = 0.3$). During follow-up, the group of DQ2/8 patients with CRP > 1 mg/l showed significantly higher levels of plasma MMP-8 than the non-DQ2/8 group.

Conclusions: Diabetes-risk genotype DQ2/8 and inflammation contribute to vascular changes in children and adolescents with T1D.

O10-7

Cardioprotective Effect of Metformin Against Doxorubicin Cardiotoxicity in Rats

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Introduction: The clinical use of doxorubicin, which is a strong antineoplastic agent, is limited due to its cardiotoxic side effects. Metformin is a drug with anti-hyperglycemic effects, and it has been shown to have a cardioprotective effect on left ventricular function in experimental animal models of myocardial ischemia. The present study investigated the cardioprotective effect of metformin in rats with doxorubicin cardiotoxicity.

Methods: Forty male, 10-week-old Wistar albino rats were randomly divided four groups. The control group rats were intraperitoneally administered saline solution twice a week, four doses in total. The doxorubicin group rats received (cumulative dose: 16 mg/kg) intraperitoneally. The metformin group rats received metformin with gavage. The doxorubicin + metformin group rats received doxorubicin (cumulative dose: 16 mg/kg) intraperitoneally and metformin orally with gavage.

Left ventricular functions were evaluated by using M-mode echocardiography one day after the last dose of doxorubicin. Heart tissue samples were histopathologically examined. Cardiomyocyte apoptosis was detected using *in situ* terminal deoxynucleotide transferase assay (TUNEL). Serum brain natriuretic peptide and C-type natriuretic peptide levels were measured. Catalase, superoxide dismutase, glutathione peroxidase, and tumor necrosis factor alpha levels were analyzed in the heart tissue.

Results: Our results showed that doxorubicin treatment caused significant deterioration in left ventricular functions by echocardiography, histological heart tissue damage, and increase in cardiomyocyte apoptosis. The while, doxorubicin + metformin group showed protection in left ventricular function, inhibition of cardiomyocyte apoptosis (Figure).

Conclusion: The present study provided evidence that metformin has cardioprotective effects against doxorubicin cardiotoxicity.

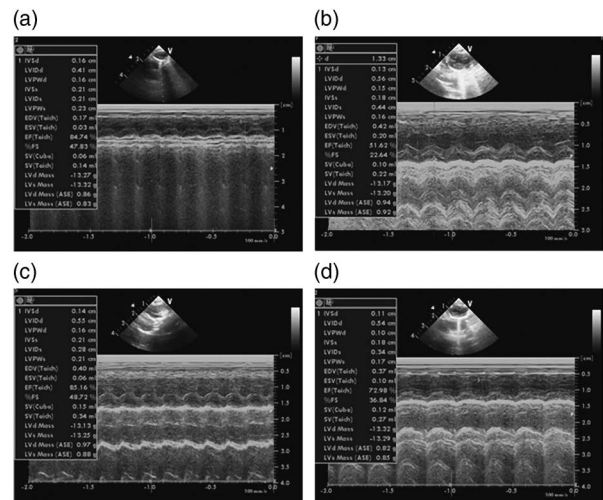


Figure. M-mode left ventricle echocardiography examination in experimental groups. (a; control group, b; doxorubicin group, c; metformin group, d; doxorubicin + metformin group).

O11-1

Acute Desaturation Following Balloon Angioplasty of Aortic Arch Obstruction after Norwood Surgery

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Introduction: Obstruction of the reconstructed aortic arch is a recognized complication after Norwood surgery, this can be

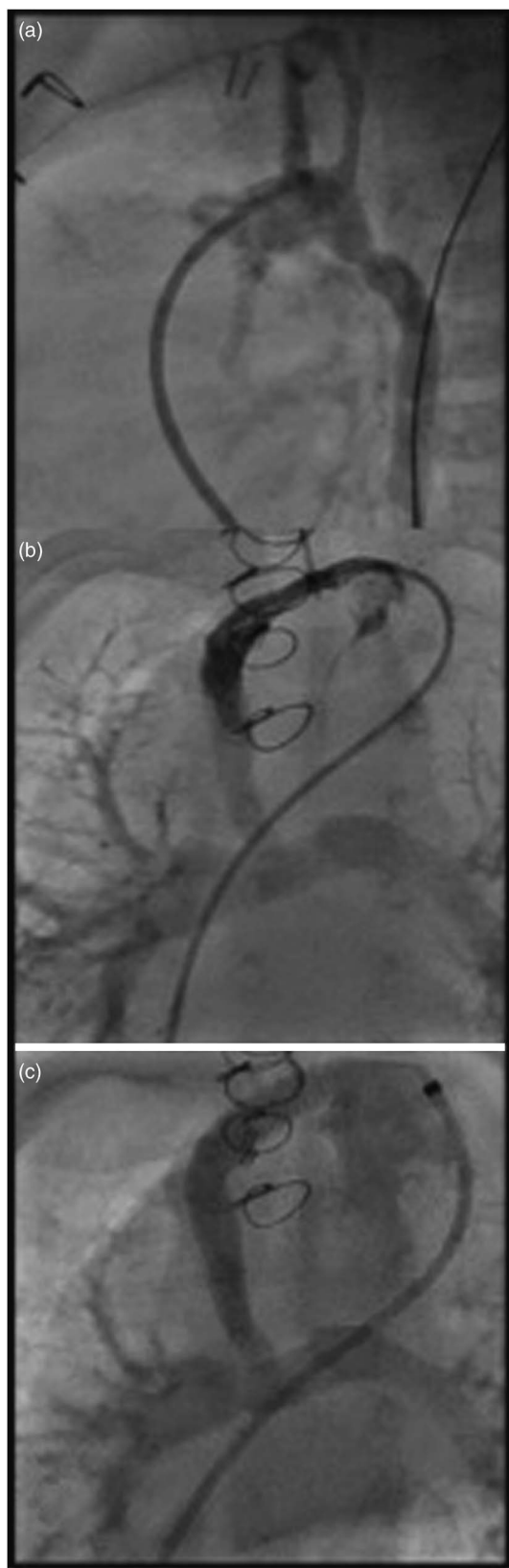


Figure 1. A, Aortic re-coarctation. B, Stenosed proximal Sano. C, Above conduit post-stenting.

addressed by balloon angioplasty prior to stage 2 surgery (cavopulmonary (CP) shunt.) However, the increased afterload created by the arch obstruction can act as an important driving pressure for pulmonary blood flow. Thus, relief of arch obstruction after Norwood may lead to acute desaturation. This report seeks to describe occurrence and management options for this phenomenon.

Methods: Retrospective, single center case-note review of all post-Norwood patients who underwent balloon angioplasty of aortic arch obstruction between 2008 and 2014. Success of angioplasty was considered a reduction of invasive peak gradient to <10 mmHg.

Results: Norwood surgery was performed in 232 neonates, with 195 surviving to pre-stage 2 catheter (84%). Balloon angioplasty to re-coarctation was performed in 52 (26%) patients, three interventions were unsuccessful, two underwent re-intervention. Of these, 15 (29%) suffered acute severe desaturation (drop of >10%). One (age < 3 months) underwent emergency Sano conduit revision, 1 underwent emergency CP shunt within 12 hours, 2 underwent CP shunt < 7 days, 2 desaturated >10% but were managed conservatively and underwent elective CP shunt. Nine patients (18%) underwent concomitant stenting +/- upsizing of the RV-PA conduit or BT shunt, see figure 1 for example angiography. In these patients, stage 2 surgery was deferred until the child achieved optimum age and weight, (median 54 days (31-102)). Two patients had transient complete heart block, one patient had VT requiring DC cardioversion.

Conclusion: Balloon angioplasty of significant aortic arch obstruction after Norwood 1 surgery can result in acute severe desaturation requiring early intervention. This complication must be anticipated. Concomitant stenting of the RV-PA conduit or BT shunt is an effective technique to address this problem. It is recommended that operators are prepared to stent the RV-PA conduit or BT shunt to avoid emergency conduit revision or high risk early cavopulmonary shunt.

O11-2

Embolization of Veno-venous Collaterals after the Fontan Operation is Associated with Decreased Survival

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Introduction: After Fontan operation, hemodynamically significant veno-venous collateral (VVC) vessels can lead to systemic arterial desaturation. Outcomes after embolization of VVC have not been determined. We sought to determine the frequency of and outcomes for patients undergoing VVC embolization after Fontan operation.

Methods: We retrospectively analyzed clinical and hemodynamic data of patients who underwent cardiac catheterization after the Fontan operation from 1995-2014 at Mayo Clinic. Clinical, imaging, and hemodynamic data from patients with VVC were compared based on intervention (embolization) versus non-intervention.

Results: 496 patients with prior Fontan operation were identified. 109 VVC were identified in 72 patients (37 males, mean age 26 ± 12 years). VVC most commonly originated from the innominate vein (43%), inferior vena cava (IVC)/hepatic vein (20%), and the superior vena cava (19%) and most commonly connected to the right upper pulmonary veins (31%), left upper pulmonary veins (23%), directly to the pulmonary venous atrium (18%), and the coronary sinus (17%). Embolization was performed in 31/72 patients (43%). Following embolization, no improvement was demonstrated in systemic oxygen saturation, hemoglobin concentration, and cardiac index declined. Overall, fifteen patients

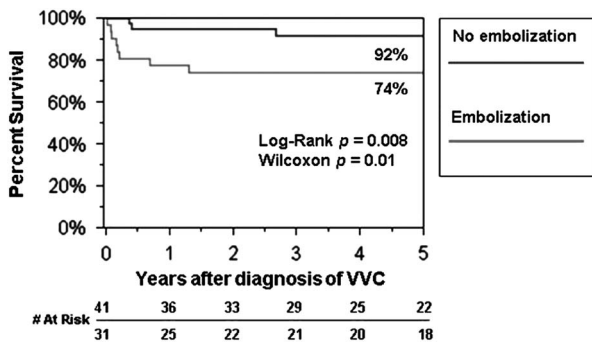


Figure.

(21%) died at a mean of 2.8 ± 4 years after embolization. 5-year survival of patients with VVC undergoing embolization was 74% compared to 92% in those patients who did not undergo embolization ($p < 0.01$) (Figure). In multivariate analysis, significant predictors of death were embolization (HR = 9.3 [95% CI, 2.8 – 42], $p = 0.0001$); atriopulmonary Fontan (HR = 4.2 [95% CI, 1.4 – 15], $p = 0.01$), and heterotaxy (HR = 3.7 [95% CI, 1.0 – 15], $p = 0.05$).

Conclusion: This is the first investigation to examine outcomes after embolization in patients with VVC after Fontan operation. We observed a significantly decreased 5-year survival in patients who had embolization. Additionally, after embolization no benefit was demonstrated in systemic arterial oxygen saturation and hemoglobin concentration and cardiac index was decreased. Embolization of VVC in patients after Fontan should be avoided especially in patients with atrio-pulmonary type Fontan and heterotaxy. These patients may benefit from the “natural” fenestration that VVC provide.

O11-3

Transcatheter pulmonary valve implantation (TPVI): 3D-roadmapping in grown-up congenital heart disease

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Introduction: Three dimensional rotational angiography (3DRA) is a useful technique for ruling out coronary compression in pre-testing for transcatheter pulmonary valve implantations (TPVI): One single contrast admission in the aortic root while high pressure ballooning the pulmonary artery (PA) shows the spatial relationship of the coronaries to the position to the planned stented pulmonary valve. Selective angiographies of single coronary arteries become unnecessary. The 3D information from the 3DRA additionally enables intra-procedural 3D guidance after integration and overlay of routinely performed pre-interventional MRI.

Methods and Results: Between September 2012 and December 2014, 33 patients were analysed after pre-testing and TPVI with 3DRA and 3D-guidance in the cardiac catheterization laboratory. 3DRA helped to rule out contraindications for TPVI like coronary compressions in 3 cases. In 30 cases, TPVI were successfully performed. 3D-guidance with MRI overlay was used in 20 patients. Radiation dose, consumption of contrast dye and fluoroscopy time were analysed. In the 3D-guided group median dose-area product (DAP) was 4741.8 (1010.9–12944) μGym^2 , contrast consumption 2.3 (1.0–3.8) ml/kg and fluoroscopy time 22 (7.8–126.5) minutes compared



Figure.

to the non-guided group with DAP of 4513.4 (1861–19820) μGym^2 , contrast consumption of 2.6 (1.4–7.1) ml/kg and fluoroscopy time of 33.6 (9.4–66.3) minutes [Median, (Min–Max)]. In follow-up, no complications like stent fracture or endocarditis occurred.

Conclusions: 3DRA while pre-testing safely displays coronary compression. MRI-overlay is possible by 3D–3D registration with the 3DRA, facilitates TPVI and has the potential to reduce fluoroscopy time, radiation and contrast dye.

O11-4

Single center experience on cutting balloon angioplasty in the treatment of branch pulmonary artery stenosis after surgery in small children

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Introduction: Cutting balloon angioplasty (CBA) is a promising technique for the treatment of highly challenging vascular stenosis especially in peripheral pulmonary artery stenosis. We will present our experience on CBA for the treatment of branch pulmonary stenosis (BPS) in childhood.

Methods: Seventeen children <5 years old, median age of 18 (4.5–54) months, median weight of 9 kg (5.5–16), with BPS after surgical repair of congenital heart diseases treated with CBA were prospectively analyzed. We used staged approach for dilation of stenotic vessel to avoid rupture. After CBA, further dilation was performed with optimal size low pressure balloon (LPB), and if it was not effective, dilated 1–2 mm < optimal size high-pressure balloon (HPB) was used. If result was insufficient, optimal size LPB was used again.

Results: Diagnosis of children was s/p Jatene in 12, s/p tetralogy of Fallot in 2, repaired truncus arteriosus in 2, s/p Glenn in 1. Twenty-eight vessels were dilated with CBA. Stenotic vessels were RPA in 4, LPA in 2, bilateral pulmonary artery in 11. Nine vessels underwent subsequent LPBA and 19 vessels underwent

HPBA. The diameter of the vessels increased from 3.2 ± 1.08 to 5.2 ± 1.4 mm ($p < 0.001$). The RV/LV pressure ratio decreased from 0.92 ± 0.14 to 0.59 ± 0.17 ($p = 0.001$). Vessel diameter increased by $> 50\%$ in 14 patients; procedural success rate was 82%. Increase in vessel diameter $< 50\%$ was observed in 2 patients, CBA was failed to dilation in 1 patient. Patients with supra-valvar stenosis after Jatene procedure were least responsive group (2/5). Success rate was significantly higher in patients who underwent additional HPBA as compared with additional LPBA (%95 vs % 66). The procedure and fluoroscopy times were 213 ± 56 and 58 ± 16 minutes, respectively. In one s/p Jatene patient, ascending aorta to the RPA fistula was developed and underwent to surgery due to significant residual shunt after device closure. No procedure related mortality was observed. At a median follow-up of 15 months, 2 patients who successfully treated with CBA underwent operation for supra-valvar pulmonary stenosis.

Conclusions: CBA is a feasible technique for treatment of BPS. CBA can be useful to delay the intervention up to appropriate age for pulmonary stent implantation.

**O11-5
Is ultrasound guided vascular access for cardiac catheterization safe and fast? Prospective and randomized study**

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Objectives: This prospective and randomized study was designed to assess the safety and efficacy of two methods of vascular access obtaining - anatomy/pulse palpation or ultrasound (USG) guided.

Table 1.

	Randomization (patients)	Conversion (patients.)	Mean time to conversion (s)	Procedure duration (min)	p	Switch to other site (patients)
Group V						
USG	43	1	500	$48,7 \pm 20,7$	0,65	5
Anatomy	45	5	934	$47,8 \pm 22,5$		11
Group A						
USG	28	1	180	$57,5 \pm 27,9$	0,22	5
Anatomy	16	3	470	$48,1 \pm 22,1$		3

Table 2. Results in the group V - Table 2.

Group V								
	BSA (m ²)	p	Age (months)	p	Time to complete (s)	p	Punctures (number)	p
USG	$0,61 \pm 0,41$	0,94	14 (0-199)	0,94	$145,5 \pm 123,0$	0,007	$1,2 \pm 0,0$	$< 0,001$
Anatomy	$0,58 \pm 0,37$		12(0,1-207)		$283,0 \pm 308,2$		$2,9 \pm 3,0$	

Table 3. Results in the group A - Table 3.

Group A								
	BSA (m ²)	p	Age (months)	p	Time to complete (s)	p	Punctures (number)	p
USG	$0,49 \pm 0,30$	0,44	14 (0-199)	0,74	$199,9 \pm 170,3$	0,11	$1,3 \pm 0,5$	$< 0,02$
Anatomy	$0,52 \pm 0,29$		9,5 (3-143)		$284,1 \pm 161,1$		$2,9 \pm 2,7$	

Table 4. Complications (*significant difference)

Group V						
	Bruise/Hematoma*	A-V fistula	Thrombosis	Transient ischemia	Bleeding	Vessel injury
USG	0	0	0	1	0	0
Anatomy	5	1	0	0	0	0
Group A						
USG	8	0	0	2	1	0
Anatomy	3	0	0	1	0	0

Methods: in patients qualified for cardiac catheterization venous (group V) and/or arterial (group A) access was chosen based on the type of the study and anatomy. Patients were randomized to cannulate the vessel using anatomy/pulse palpation or USG. Two experienced operators participated.

Results: in 105 patients we obtain 88 venous and 44 arterial accesses. In the group V: 9 (10%) patients have right internal jugular vein punctured, 79 (90%) either right (51patients) or left (28 patients) femoral vein. In group V ultrasound guided puncture was utilized in 47 (53%) patients. In the group A, either right (34 patients) or left (10 patients) femoral artery was punctured, USG was used in 30 (68%) - Table 1.

Conclusions:

1. USG guided vascular access is shorter and needs less punctures to complete.
2. In the study group, in both methods complications were benign, rare and transient.

**O11-6
Patent ductus arteriosus closure (PDA) by Amplatzer Duct Occluder II additional size (ADOIIAS) in premature infants less than 1500 grams**

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Objective: Treatment of PDA in micropremies less than 28 wks GA is a dilemma when medical treatment fails or is contraindicated. Conservative therapy and surgical ligation are other alternatives. Considering the complications of these treatments and the advent of ADOIIAS, we developed a program of PDA closure by interventional catheterism to close Hemodynamic Significant (HSPDA) in very preterm infants.

Methods: HSPDA was defined by high pulmonary blood flow and low organ blood flows and a tubular duct diameter > 1.5 mm/kg. Procedures were done in cath lab and babies anesthetized (ketamine-sufentanil-midazolam). A 4 F sheath was inserted via the femoral vein and the Torqvue catheter positioned into the descending thoracic aorta. The ADOIIAS occluder was delivered under lateral X ray and ultrasound (US) guidance, without contrast injection. It was first used on four babies > 2 kg.

Results: fifteen infants were included. The mean birth weight was 788 ± 162 g (550-1400) mean gest age 26.55 ± 1.55 wks. In this series three infants presented pulmonary hemorrhage, nine renal failure, six were under mechanical ventilation and three pulmonary hypertension + nitric oxide. Mean weight at catheterization was 984 ± 186 g (680-1400). Mean age at procedure was 19 ± 6 days (10-44). Mean Procedure duration was 25 ± 6 min (15-40) and mean X ray exposure 11 ± 5 min (7-18). Mean DA diameter was 3.27 ± 0.53 mm (2,3-4,2) and length 4.66 ± 0.93 (3-6). Fourteen ADOIIAS were released $4 \times 2(4)$, $4 \times 4(3)$, $5 \times 2(6)$, $5 \times 4(1)$ mean waist diameter 4.5 ± 0.5 mm. Two were repositioned and two switched to a larger size due to instability and periprothetic shunt

(DA diameter 3.5 mm and waist diameter 4 to 5 mm). Two transitional intraprothetic shunts were closed in the following days. Post procedure events: higher oxygen requirements, oliguria (4) and diuretics, Pulmonary Hypertension (3 Nitric Oxide), one post closure syndrome, blood transfusion (3) Three infants died. One due to an hemopericardium which was drained (680 g); One infant who was previously anuric, died ten days after the closure. One death, one month after the procedure, was due to a fulminant enterocolitis. **Conclusion:** Interventional catheterism can be considered as an alternative to other treatments to close HSPDA with the ADOIIAS in very premature infants when combining X-ray and US.

O11-7

Perimembranous Ventricular Septal Defect Device

Closure: Choosing between ADOI and ADOII

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Background: Transcatheter closure of perimembranous ventricular septal defects (pmVSDs) is a well-established procedure. Recently, Amplatzer duct occluders (ADO) I and II have been reported to close large series of pmVSDs successfully (off-label use). ADOs are economical compared with the standard Amplatzer VSD occluders; a major consideration in developing countries with low budget programs.

Aim of the work: We report closure of symptomatic, hemodynamically significant pmVSDs using the ADOI and ADOII devices. Although there are no set criteria for choosing between ADOI and ADOII, the former's price tag includes snare and long sheath. Thus, we aim to predetermine device usage based on transthoracic echocardiography (TTE) findings.

Methods: Between March 2013 and November 2014, 30 patients had transcatheter closure of pmVSDs using the ADO devices. The median age was 4.5 years (range: 1.1-54 yrs) and median weight was 15 kg (range: 6-87 kg). ADOII could not be used in VSDs larger than 6 mm and/or with a large aneurysm. The median VSD size as assessed by echocardiography was 6 mm (range: 3-12 mm), while by angiography it was 5 mm (range: 3-9.5 mm).

Results: The median fluoroscopy time (FT) was 8 minutes (range: 5-38 min). We inserted ADOI in 14 patients and ADOII in 16 patients (no significant difference between median age and weight in each group). VSD size was significantly larger and FT was longer in ADOI patients; the device type matched what was decided from TTE data in 85% of cases. Follow up ranged from 1-21 months (median 8 months). The mean LVEDD z-score of the patients was 1.1 before VSD closure, while it was 0.63, 0.35 and 0.23 at the 1-month, 3-month and last follow up, respectively. Complete closure rates immediately, at 24 hours and at last follow-up were 80%, 90% and 94% respectively. No patient developed heart block or any other complication.

Conclusion: ADOI and ADOII are equally safe and effective in pmVSD closure. ADOII use, although cheaper than ADOI, is limited to smaller VSDs. The choice between ADOI and ADOII can be decided by TTE prior to procedure which is convenient in low economic programs.

O12-1

Automatic computerized evaluation of resting ECG in preparticipation screening of young athletes: validation study

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Introduction: There is ongoing debate on inclusion of resting ECG as part of pre-participation examination of asymptomatic competitive athletes. The aim of this study was to test newly designed computer algorithm for detection of ECG abnormalities according to the Seattle criteria for pre-participation examination of adolescent and young adult athletes.

Methods: Seattle criteria were split into 16 binary items and final binary statement was added. Computerized standard 12-lead ECGs were obtained using a device compatible with AHA standards (BTL 08-LC, BTL Industries, UK) from consecutive examinations of athletes and from selected patients with cardiac disease of comparable age. Reference evaluation of all items in all records was obtained by agreement of two skilled physicians. Prototype of computer algorithm was developed as extension of marketed software (BTL CardioPoint 2.23) developed by Medical Technologies CZ, a.s. Items were classified (highest priority first) as abnormal, borderline or normal. Borderline ranges of measured parameters (e.g. PR interval, QRS duration) were extension of abnormal range depending on mean error of automatic measurement. Both abnormal and borderline findings were regarded to reflect pathology. If automatic algorithm failed to evaluate the item was classified unknown. Final statement was based on one or more items with the highest priority.

Results: Comparison of computer algorithm with reference was performed on 316 records (258 athletes, 58 patients). Sensitivity/specificity varied considerably in particular items between 22-100/56-100% with lowest values for pre-excitation/ST segment depression. Negative predictive value was high (98-100%). Unknown classifications occurred in 3/16 items: left atrial enlargement (11% of records), intra-ventricular conduction delay (2%) and pre-excitation (1%). Sensitivity/specificity/negative predictive value of the final statement was 100/23/100%.

Conclusions: The study demonstrates that automatic computerized evaluation may be suitable as the initial step of the pre-participation ECG screening process because it has a high negative predictive value and successfully eliminates part of normal records. Low sensitivity for some criteria (pre-excitation) and low specificity limits, however, its current use. (Supported by MH CZ – DRO, University Hospital Motol, Prague, Czech Republic 00064203 and TA2-1258 Technology Agency of the Czech Republic).

O12-2

RHOA-ROCK signalling is necessary for lateralization and differentiation of the sinoatrial node

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Background: Disturbances in the normal electrical activation pattern of the heart are an important cause of mortality and morbidity in the pediatric and adult population. Up to date, the mechanisms responsible for conduction diseases like sick sinus syndrome remain unknown. RHOA, involved in cellular migration, proliferation and transcriptional regulation, is expressed within the developing cardiac conduction system in chick and disruption in adult mice results in arrhythmias including bradycardia and atrial fibrillation. How this occurs is largely unknown.

Aim: To assess the role of the RHOA-ROCK signaling pathway in the formation and differentiation of the sinoatrial node (SAN) during embryonic development.

Methods: The role of RHOA-ROCK signalling was studied using chemical inhibition (Y-27632) during chicken heart development. The electrogram and atrial activation patterns were studied by ex ovo electrophysiological recordings. The developing SAN area was characterized by gene expression analysis using quantitative real time polymerase chain reaction (qPCR) for known pacemaker genes and with immunohistochemical stainings.

Results: Early in development, the entire myocardium of the inflow area (sinus venosus) of the heart has pacemaker potential. This area includes a 'transient left-sided' SAN as well as the definitive right-sided SAN. The pacemaker potential was confirmed by ex ovo electrophysiological measurements, expression of the cation channel HCN4, lack of NKX2.5 and gene expression of the transcription factors SHOX2 and TBX3. Later in development, this pacemaker potential is restricted to the right-sided SAN both in function and in gene expression. Disruption of RHOA-ROCK signalling results in an immature sinus venosus myocardium, maintaining the overall pacemaker potential with a significant lower heart rate. The sinus venosus myocardium has aberrant left-right patterning shown by down-regulation of PITX2C at the left side, up-regulation of NKX2.5 at the right side and global up-regulation of ISL1 expression. ROCK inhibition also results in abnormal pulmonary vein development. In vitro experiments confirm the effect of Y-27632 is exclusive for sinoatrial cardiomyocytes.

Conclusions: Abnormalities in the RHOA-ROCK pathway result in aberrant right/left patterning, abnormal differentiation of the sinus venosus progenitor cell population and abnormal differentiation of the right-sided SAN as definitive pacemaker of the heart.

O12-3

Genetics in postmortem pediatric sudden death

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The reason behind a sudden death of a young individual remains unknown in up to 50% of postmortem cases. Pathogenic mutations in genes encoding heart proteins are known to cause sudden cardiac death. Objective. The aim of our study was to ascertain whether genetic alterations could provide an explanation for sudden cardiac death in a juvenile cohort with noconclusive cause of death after comprehensive autopsy. Once this has been proven, analyse the familial screening and see the implication within each family.

Methods: Twenty-nine cases < 15 years showing no-conclusive cause of death after a complete autopsy were studied. Genetic analysis of 7 main genes associated with sudden cardiac death was performed using Sanger technology in low quality DNA cases, while in good quality cases the analysis of 55 genes associated with sudden cardiac death was performed using Next Generation Sequencing technology. With these results, mutations were screened through parents and first and second degree relatives and clinical decisions were taken accordingly.

Results: Thirty-five genetic variants were identified in 12 cases (41,37%). Ten genetic variants in genes encoding cardiac ion channels were identified in 8 cases (27,58%). We also identified

9 cases (31,03%) carrying 25 genetic variants in genes encoding structural cardiac proteins. Nine cases carried more than one genetic variation, 5 of them combining structural and non-structural genes.

In 80% of the cases familial screening could be performed. Carriers of these mutations were identified and clinical and therapeutic decisions were taken accordingly following international guidelines for sudden death prevention (data to be finished by march 2015).

Conclusions: Our study supports the inclusion of molecular autopsy in forensic routine protocols when no conclusive cause of death is identified. Around 40% of sudden cardiac death young cases carry a genetic variant that could provide an explanation for the cause of death. Because relatives could be at risk of sudden cardiac death, our data reinforce their need of clinical assessment and, if indicated, of genetic analysis.

O12-4

Prospective School-based Electrocardiographic Screening Program* in Children: The Widest Experience in Europe

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Introduction: This project aims to demonstrate the normal electrocardiographic (ECG) data and percentiles of children aged 6-18 via 12-channel ECG recordings and to determine cardiac conduction problems and incidence of congenital heart diseases.

Methods: The project "ECG screening of school-aged children" was started in January, 2013 following approval by the Public Hospitals Association of Çekmece-Istanbul, Ministry of Health, Republic of Turkey. Informed consents were received from families; 12-channel ECGs of children recorded school were transferred to the ECG storage system MUSE® at our hospital. The children with pathologies in the report were re-evaluated at our hospital; tests and treatments were performed.

Results: 120.164 children, whose ECGs were recorded over 2 years, were included in the study. 7209 children (6%) reported as probably pathologic were evaluated. Electrical and structural abnormalities were determined in 596 children (0.49%) whose necessary tests (ECHO in 2822, Holter in 505 and exercise testing in 304) were completed. The electrical abnormalities were: first degree atrio-ventricular block (AVB) in 47 (0.039%), second degree AVB in 12 (0.099%), complete AVB in 2 (0.0016%), WPW syndrome in 13 (0.0191%), borderline long QT + long QT syndrome in 56 (0.046%), early repolarization in 13 (0.011%), premature ventricular complexes (PVC) in 55 (0.045%) and premature atrial complex (PAC) in 43 (0.035%) patients. The structural pathologies were: atrial septal defects in 36 (0.029%), ventricular septal defects in 6 (0.0049%), aortic valve pathology in 50 (0.0412%), mitral valve pathology in 107 (0.089%), patent ductus arteriosus in 8 (0.0066%), hypertrophic cardiomyopathy in 8 (0.0066%), patent foramen ovale in 60 (0.05%) patients. 15 patients with congenital heart diseases were treated with surgical or trans-catheter procedures and dual chamber pacemakers were inserted in 2 patients. EPS and/or ablation were performed

in 18 patients with different arrhythmia substrates. Clinical follow-up was planned for the other patients on medical therapy.

Conclusion: The normal ECG data of healthy school-age children were demonstrated in this study, which includes the largest group of children in Turkey and Europe. As a result, we think that structural-electrical pathologies that may cause morbidity and sometimes sudden cardiac death can be detected earlier and treatment can be provided.

O12-5

Risk factors for sudden death and for heart failure-related death in childhood hypertrophic cardiomyopathy

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Background: To date limited data is available to predict the progression to end-stage heart failure with subsequent death (non-SCD), need for heart transplantation or sudden cardiac death (SCD) in children with hypertrophic cardiomyopathy (HCM). Current treatment strategies in children are predominantly based on the known risk factors for SCD in adults with HCM.

Objectives: The aim of study was to determine predictors of long-term outcome in children with HCM including known adult risk factors for SCD and previously proposed pediatric risk factors for death. Moreover, we distinguish both arrhythmic and heart failure end points to determine specific risk factors these two modes of unfavorable outcome.

Material and Methods: A total of 112 children (median age 14.1, IQR 7.8-16.6 years, 60% male) were followed up for the median of 6.5 years for the development of morbidity and mortality, including the pre-specified primary end points: the cardiovascular death, resuscitated cardiac arrest, appropriate implantable ICD discharge, a heart transplant and arrhythmic or heart failure-related secondary end points. Heart failure end point included heart failure-related death or heart transplant and arrhythmic end point included resuscitated cardiac arrest, appropriate ICD discharge or sudden cardiac death.

Results: Overall 23 (21%) patients reached the pre-defined composite primary end point. At 10 year follow-up the event-free survival rate was 76%. Thirteen patients (12%) reached the secondary arrhythmic end point and 10 patients (9%) reached the secondary heart failure end point. In multivariate model prior cardiac arrest ($r=0.658$), QTc dispersion ($r=0.262$) and NSVT ($r=0.217$) were independent predictors of the arrhythmic secondary end point while heart failure ($r=0.440$), left ventricular posterior wall thickness ($r=0.258$), left atrial size ($r=0.389$) and decreased early transmitral flow velocity E ($r=0.202$) were all independent predictors of the secondary heart failure end point.

Conclusion: There are differences in the risk factors for sudden death and for heart failure-related death in childhood hypertrophic cardiomyopathy. Only prior cardiac arrest, QTc dispersion and non-sustained ventricular tachycardia predicted arrhythmic outcome in patients aged <18 years. Left atrial size, left ventricular posterior wall thickness and decreased early transmitral flow velocity were strong independent predictors of heart-failure related events.

O12-6

Clinical Outcomes of Left Cardiac Sympathetic denervation: Single Centre experience

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Introduction: Left cardiac sympathetic denervation (LCSD) has been used for many years in treating refractory ventricular arrhythmias. We report our single-center experience in performing LCSD.

Methods: We have performed a prospective study of consecutive 18 patients, who underwent LCSD since Nov-2011 until April-2014.

Syncope, aborted cardiac arrest (ACA) and appropriate ICD discharge, were classified as cardiac events (CE).

Results: Median age at LCSD was 12.5 years (range 1.0-52.1), weight was 30.0 kg (range 7.8-100), 12 (67%) were male. Diagnoses included 7 (38,9,5%) patients with LQTS (including 4 (22,2%) with JLN), 7 (38,9%) with CPVT, 1 (5,6%) with Histiocytoid cardiomyopathy, 1 (5,6%) with Emery-Deifuss syndrome, and 2 (11,1%) with Idiopathic VF. Prior to LCSD, 9 (50%) had experienced ≥ 10 CE, 6 (33%) had 5-to-9 CE, and 3 (17%) had 1-to-4 CE, with an overall median of 10 (2-87) CE. All patient had tried maximal dose of beta-blockers (BB) before LCSD, 4 were on BB and flecainide, 1 on BB and mexilitine and 1 had tried BB, amiodarone, sotalol and lignocaine. 13 (72%) had an ICD before LCSD, all for secondary prevention. Video assisted thoracoscopic approach (VATS) could be performed in 17 (94%) patients. Only the smallest patient (7,8 kg) underwent a posterolateral thoracotomy approach as the subpleural ICD coil interfered with VATS ports incisions. Intraoperative complications included 1 VF episode that was promptly managed with esmolol and cardioversion.

After this incident we changed our protocol adding administration of local anaesthesia before applying diathermy with no further intraoperative arrhythmia. Long-term side effects included 1 patient with a transient Horner's syndrome. Over a median follow up of 29.2 months (range 1,7-42,5), 12 (67,7%) patients had no breakthrough of CE (BCE), 4 (22,2%) had 1-to-4 BCE, 1 (5,6%) had 5-to-9 BCE, and 1 (5,6%) had >10 BCE. Overall, there was a significant reduction of CE after

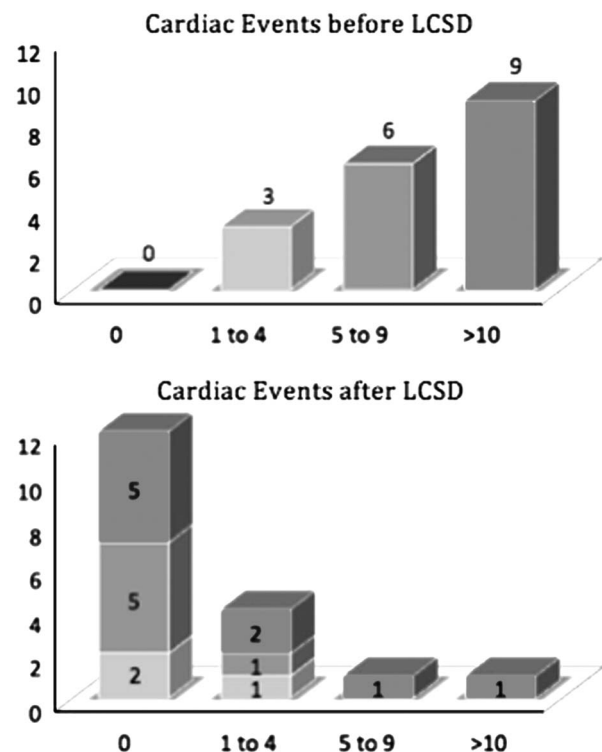


Figure 1.

LCSD (10.0 (2-87) to 0 (0-10); $p < 0.001$). Figure 1. The 1 child with histiocytoid cardiomyopathy underwent heart transplantation for persistent arrhythmias.

Conclusions: LCSD is a safe and effective technique with few complications in drug refractory ventricular arrhythmias due to channelopathies and other cardiomyopathies. VATS approach is a safe and feasible procedure but may be difficult in very small patients. A significant reduction of BCE has been seen in the majority of patients.

O12-7

An optimal approach for endocardial mapping of idiopathic ventricular arrhythmias in pediatric patients: 11 years experience

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Objective: To optimize the methods of endocardial mapping of idiopathic ventricular arrhythmias (VA) in pediatric pts.

Methods: In 2003-2013, 387 pts (165 - females) aged 6 to 17 yo which received invasive treatment of VA in one hospital were included into the study. ECG, 24-hour Holter monitoring and echocardiography were performed. The following criteria were used for endocardial mapping of VA substrate: presystolic activation time and pace mapping results. Two approaches for interpretation of those data were used. «Typical approach»: good prognosis for ablation – presystolic activation time is > 25 ms, spontaneous and artificial QRS morphology are identical. «Novel» approach was based on calculation of the probability of successful ablation using regression model, which included results of activation and pace mapping. Pts were divided in two groups depending on approach chosen: I (156 pts) – «Typical» approach, II – (231 pts) «Novel» approach. Efficacy, duration of procedure and effective doses were evaluated.

Results: In group I efficacy of procedure – 69.7%, in group II – 93.6% ($p < 0.05$). Using the logistic regression model the probability of successful ablation (P) was divided into three grades: “low” ($P < 0.75$), “medium” ($0.75 < P < 0.9$) and “high” ($P > 0.9$). During evaluation of presystolic activation time (T) “low” P was calculated for $T < 29$ ms, “medium” P was calculated if $29 \text{ ms} < T \leq 73 \text{ ms}$ and “high” P was determined for $T > 73 \text{ ms}$. Pace mapping results were divided in two groups: “similar” and “identical”. Only “medium” P was found in both cases. But P in “identical” pace mapping group was found in 2.6 times higher than in “similar” ($p < 0.05$).

There were no significant differences in effective doses and duration of procedure ($p > 0.05$).

Conclusions: An optimal approach for endocardial mapping of VA in pediatric pts should be based on evaluation of probability of successful ablation. Pace mapping may be useful when activation mapping results are not acceptable.

MP1-1

Heart-beating aortic arch repair: preliminary results in neonates and infants

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Introduction: The strategies for aortic arch repair (AAR) in neonates/infants have evolved along the time. Simultaneous cerebro-myocardial perfusion (heart-beating technique) is the most recent modification, although it is not widely performed worldwide. We present our preliminary results by using this strategy.

Methods: Since 2013 we included this technique as the standard approach for AAR: 18 patients (20 procedures; January 2013–December 2014). Median age 50 ± 27 days (range 3–270); median weight 3.7 ± 1.4 kg (range 2.1–7). Hypoplastic aortic arch (HAA) was associated with cor triatriatum ($n = 3$), ventricular septal defect (VSD) ($n = 3$; 1 interrupted aortic arch), transposition of great arteries ($n = 2$; 1 VSD), partial atrioventricular septal defect (AVSD) ($n = 1$), hypoplastic left heart syndrome ($n = 4$), double-outlet right ventricle ($n = 1$), severe aortic stenosis ($n = 2$) and as isolated defect ($n = 3$). Coronary perfusion (25°C) was maintained through a cardioplegia delivery system connected to the aortic cannula and heart-beating AAR was performed. When intracardiac repair was required, antegrade cardioplegia was delivered via the same catheter used for myocardial perfusion.

Results: 15 patients had intracardiac lesions: left atrium membrane resection ($n = 3$), VSD closure ($n = 3$), arterial switch operation ($n = 2$), partial AVSD repair ($n = 1$), Norwood-Sano procedure ($n = 3$), comprehensive-procedure following hybrid-approach ($n = 1$) and aortic commisurotomy ($n = 2$). Average cardiopulmonary bypass time 168 ± 73 minutes (range 93–292). Myocardial ischemia time median 30 ± 11 , (range 0–93); it was zero (5 patients). Selective cerebral perfusion median 35 ± 9 minutes (range 18–50). No cases of operative mortality were detected. No neurologic adverse events occurred. 30-day mortality was 5.5% ($n = 1$, following extra-corporeal-membrane-oxygenation). At median follow-up (11 ± 9 months; range 1–23), mortality was zero, 2 patients (11.7%) had reinterventions (cavopulmonary shunt + AAR due to arch stenosis following Norwood-procedure: 1 underwent heart transplantation 2 months later) and 3 (17.6%) underwent ballooning/stent due to aorta/pulmonary branches stenosis. All the patients ($n = 17$) remain with optimal functional class; 1 is awaiting for Ross-Konno (severe aortic valve stenosis).

Conclusions: Selective/independent cerebro-myocardial perfusion in aortic arch pathology is a safe/feasible technique, with low rates of adverse events. Heart-beating AAR should be recommended due to the reduction of the myocardial ischemia time, although comparative results with classical techniques need to be addressed.

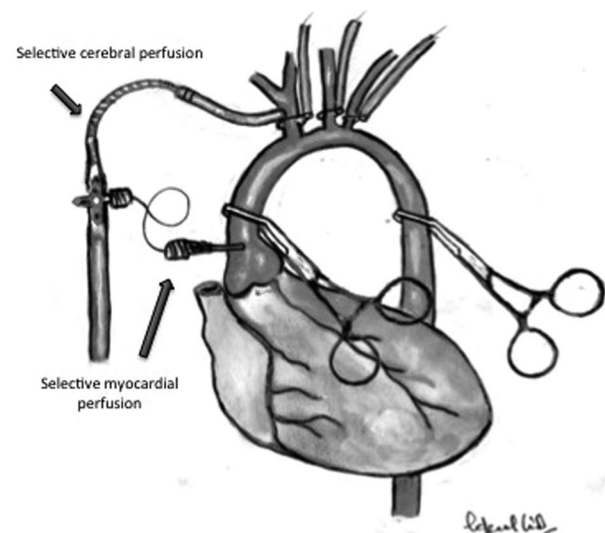


Figure.

Table.

30-day-mortality	Follow-up-mortality	Redo-incidence	Freedom-from-redo	Percutaneous-procedure-incidence	Freedom-from-percutaneous-procedure	Significant-echo-findings
5.5%-(n=1)	0%-(n=0)	11.7%-(n=2), Glenn-shunt	88.3%-(n=15)	17.6%-(n=3)	82.4%-(n=14)	-Slight stenosis right PA-(n=1) -Mild-to-moderate Ao valve stenosis-(n=1) -Severe Ao valve stenosis-(n=1)

MP1-2**Predictors of the incidence of acute kidney injury in neonates treated with peritoneal dialysis after surgery for congenital heart disease**

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Background: Acute kidney injury (AKI) is one of the most common complications of cardiopulmonary bypass surgery in children with congenital heart disease. The purpose of this study was to investigate the risk factors associated with peritoneal dialysis (PD) in a neonatal patient who developed acute renal failure (ARF) in the perioperative period after open heart surgery.

Methods: We performed a retrospective chart analysis of 259 neonates that underwent complex cardiac repair under cardiopulmonary bypass between January 2006 and September 2014. AKI was defined based on the Acute Dialysis Quality Initiative's modified for children RIFLE (pRIFLE) definitions for acute kidney risk or injury (AKI-RI) and based on the requirement of PD. **Results:** AKI occurred in 132 patients (51%), and 43 patients (17%) required renal replacement therapy within three days post-operatively. Based on a univariate analysis, the body weight, age, an Adjustment in Congenital Heart Surgery (RACHS-1) score of 4 or higher, the surgical procedure (palliative or collective), use of lower body circulatory arrest and the vasoactive-inotropic score (IVS) were associated with post-operative AKI.

In a regression analysis, the use of lower body circulatory arrest, a RACHS-1 score ≥ 4 and the preoperative serum creatinine (SCr) levels (odds ratio: 3.003, 0.271 and 2679.525; 95% confidence interval: 1.103 to 8.176, 0.087 to 0.848 and 96.110 to 74704.416; $p = 0.30$, 0.027 and 0.000) remained predictive of a subsequent need for PD. The patient age at the time of surgery and the required PD (odds ratio: 1.184 and 4.835; 95% confidence interval: 1.027 to 1.365 and 1.124 to 20.806; $p = 0.20$ and 0.034) were associated with mortality.

Conclusions: We conclude that the use of lower body circulatory arrest, a RACHS-1 score ≥ 4 and the preoperative SCr levels were associated with a high incidence of postoperative AKI and the use of PD in neonates who underwent cardiac surgery under cardiopulmonary bypass. In this study, the need to undergo PD and a younger age at the time of surgery were associated with mortality.

MP1-3**First multi-center experience with EXCOR pediatric in Poland – successful introduction of pediatric VAD therapy**

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Objective: The EXCOR pediatric VAD is an important treatment option in children with severe heart failure as bridging to heart transplantation or myocardial recovery. This retrospective study reviews the multi-center results in Poland since the introduction of EXCOR pediatric in December 2009.

Methods: A total of 23 patients were implanted (mean weight 19.7 kg, mean BSA 0.74 m², mean age 5.2 years) between December 2009 and December 2014. All patients were on inotropic support at time of implantation, nine patients required mechanical ventilation, two patients were on previous ECMO and one patient on ECC support. Leading diagnosis was idiopathic cardiomyopathy (8), CHD (5), myocarditis (6), LVNC (2), postcardiotomy syndrome (1) and restrictive cardiomyopathy (1). **Results:** Mean support time on device was 153 days (min. 3 d, max. 971 d), LVAD/BVAD ratio was 19 vs. 4 (83%, 17%). Overall survival was 74% (17 out of 23) with nine patients being successfully transplanted (39%) after a maximum support time of 433 days, four patients with device removal after recovery (17.5%) and four patients still on system (17.5%). Overall 15 pumps had been changed (1.6 events per patient year). Signs of local infection were seen in 5% (0.10 EPPY) and the rate of severe neurological events was 9% (0.21 EPPY). Major bleeding occurred in 14% (0.31 EPPY).

Conclusion: The EXCOR pediatric VAD has been successfully introduced in Poland with excellent results comparable to other experienced centers throughout US and Europe. Despite a relevant proportion of complex congenital heart disease patients and overcoming a potential learning curve the new device technology was applied with remarkable outcome and low complication rate. Long-term support of almost 3 years on VAD reflects the overall trend of extended waiting time until HTX.

MP1-4**Pattern of somatic growth and neurodevelopmental outcome in children with congenital heart disease undergoing cardiovascular surgery during first year of life**

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Background: Children undergoing cardiovascular (CVS) surgery for congenital heart disease (CHD) are at risk for growth retardation of multifactorial etiology including failure to thrive and chronic heart failure. Standard growth charts have not yet been established for this population.

Objective: To determine pattern of growth for children with CHD and to relate it to cognitive outcome based on long-term follow up data until six years of age.

Methods: We prospectively assessed growth indices for body weight (BW), body length (BL) and head circumference (HC) of 167 children (106 male) with CHD undergoing CVS within the first year of life at time of surgery, at one, four and six years of life.

Growth curves were compared with normative growth charts. Children with a genetic comorbidity were excluded. Growth was expressed as z-scores.

Results: Median gestational age at birth was 39.4 weeks (34.0–42.0) and median age at bypass surgery was 1.97 months (0.07–10.7), 111 children (66.5%) had a cyanotic CHD, including 18 with univentricular CHD. There was a large interindividual variability in growth pattern, but all growth parameters were below expected percentiles at the time of surgery, which was followed by a catchup growth of BW and BL, but not of HC until 4 years of age. Median HC remained at –0.674 standard deviations below the expected median (25th percentile instead of 50th percentile) for both girls and boys. HC at six years of age significantly correlated with IQ at six years of age ($r = 0.24, p = 0.01$) whereas BW and BL did not. D-transposition of great arteries was associated with better growth until the age of six years (BW, $p = 0.003$, BL, $p < 0.001$, HC, $p = 0.15$), whereas univentricular CHD was associated with poorer growth in BL and HC ($p = 0.05$, resp. $p = 0.03$). Other diagnoses of CHD, male sex, lower gestational age and higher surgical risk category were not associated with poorer growth.

Conclusions: Children with CHD undergoing CVS within the first year of life show a catchup growth of BW and BL, whereas HC remains more than half a standard deviation below normal, correlating also with intellectual performance at that age.

MP1-5

New enhanced left ventricular training in corrected transposition of the great arteries by increasing after- and preload

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Purpose: Patients with corrected transposition of the great arteries (ccTGA) beyond the newborn age need a left ventricular (LV) training to undergo a double switch operation (DS) (atrial plus arterial switch operation). A new approach with pulmonary arterial banding (PAB) and atrioseptectomy to increase pre- and afterload is evaluated in regards of effectivity, numbers of re-operations necessary and the outcome of DS.

Method: We report on six consecutive patients with ccTGA to be trained for DS using this enhanced LV training (eLVT). Five patients had conventional PAB before, but did not reach a sufficient gradient across the banding.

Results: Five of six patients reached systemic pressure in the LV and underwent successful DS at in mean 1.2 years after eLVT was established. The postoperative period was short and uneventful in all patients with a total ventilation time of 24 hours, stopover on ICU for 3 days and dismissal from the clinic after 11 days (median). Over the follow up period of 1.5 years (7.2 patient' years) an unrestricted cardiac function, reduction of medication and marked regression of tricuspid valve regurgitations was observed in all patients. The six patient is on eLVT for 2 years and has not reached systemic LV pressure for DS.

Conclusion: With the eLVT the number of re-PAB operations can be reduced and DS performed effectively. The course of the eLVT and the results of this patient group may justify the additional atrioseptectomy. If it has advantages in regards of LV remodelling in patients older than 12 years is not yet proven, but a reconfiguration with reduction of tricuspid valve regurgitation was seen. Long term follow-up is still needed and results from other centres are essential to underline the benefit of this procedure.

MP1-6

Early Hybrid Approach and Enteral Feeding Algorithm Prevents Necrotizing enterocolitis in Neonates with hypoplastic left heart syndrome

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Introduction: The reported incidence of necrotizing enterocolitis (NEC) in neonates with complex congenital heart defect with ductus dependent (DD) systemic circulation ranges between 6.8% and 13% despite surgical treatment; its overall mortality is between 25% to 97%. Gastrointestinal complications after hybrid palliation for neonates with DD systemic circulation is not defined yet, but seems to be comparable to Norwood Procedure.

Methods: We reviewed the incidence of gastrointestinal complication in a series of 43 consecutive neonates with DD systemic circulation underwent early hybrid palliation associated with a standardized feeding protocol. Median age and birth weight at time of surgery were respectively 1 day (range 1 to 10 days) and 3,07 kg (range 1.5 to 4,5 kg.). Median ICU length of stay was 7 days (1- 70 days) and median in- hospital length of stay was 16 days (6 - 70 days). Median of mechanical ventilation was 3 days. Hospital mortality was 16% (7/42). All patients in the post-operative period were subjected to fast weaning from ventilatory support and received treatment with vasodilatory therapy.

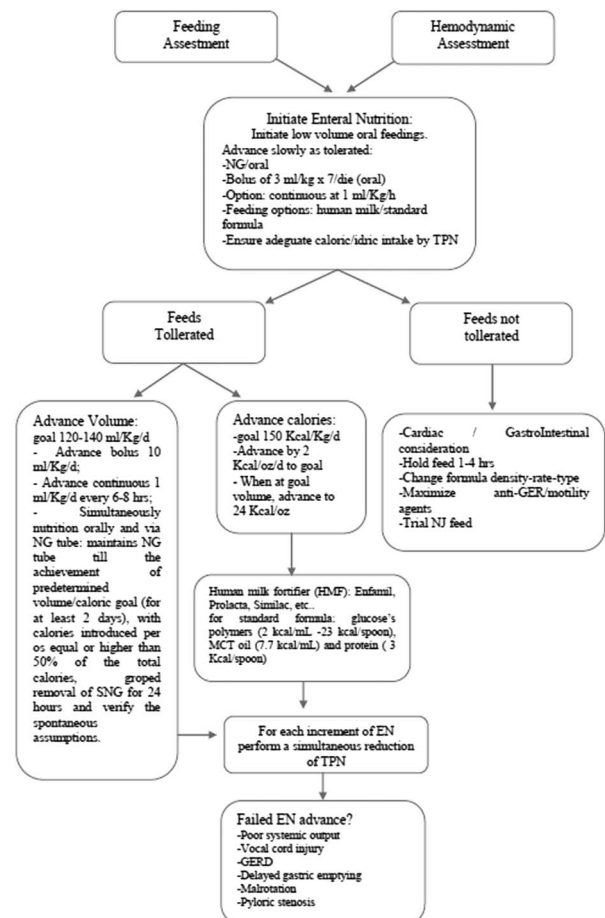


Figure.

Feeding was started six hour after estubation according to a dedicated feeding protocol. In the majority of patients enteral feedings were initiated in the first 48 hrs post procedure and increased to full volume (120 mL/kg/day) within 5 days. According with Slicker and colleagues experience, nutritional intake was increased slowly as tolerated with nasogastric (NG - continuous at 1 ml/kg/h) or oral (bolus of 3 ml/kg X 7/die). We used human milk or standard formula, ensuring adequate caloric and water intake by total parenteral nutrition (TPN). The final volume of 120 - 140 ml/kg/die can be reached by advance bolus 10 ml/kg/die or increasing continuous infusion of 1 ml/kg/die every 6 hrs

Results: None of our patients after treatment experienced any grade of NEC or major gastrointestinal adverse events.
Conclusions: Our experience indicates that combination of "early hybrid approach", fast weaning from ventilatory support, vasodilator therapy and dedicated feeding protocol adherence could reduce the incidence of gastrointestinal complication in this group of neonates.

MP1-7

Acute Kidney Injury post Cardiac Surgery and the Development of Chronic Kidney Disease in Congenital Heart Disease Survivors

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Introduction: Among CHD survivors, there is a distinctive history of cardiac surgery (CS) associated acute kidney injury (AKI) at rates up to 30-50%. The magnitude of the link between CS associated AKI and the development of chronic kidney disease (CKD) is unknown. The aim of our study was to determine the association of CS AKI with CKD in CHD survivors.

Methods: Using Danish regional population-based registries, this cohort study aimed to include all CHD patients born from 1990-2010 with first time CS between 2005 and 2010 before the age of 15 years. Utilizing in- and out-patient lab data (LABKA), we identified subjects fulfilling any KDIGO stages of AKI. A unique personal identifier enabled unambiguous data linkage and virtually complete follow up. We computed cumulative incidences of CKD stages 3-5 for patients with and without post-surgery AKI using creatinine measurements during follow-up. Individuals with

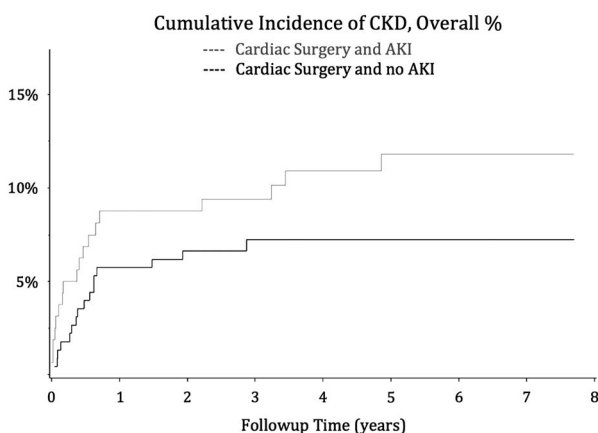


Figure.

an eGFR < 60 prior to the date of surgery were excluded. Using Cox regression we computed corresponding hazard ratios, adjusting for sex, age at first surgery, calendar period of surgery, and CHD severity.

Results: Out of 387 CHD survivors undergoing CS, 160 (41%) experienced AKI. Infants with CS in the first year of life constitute the majority (143/160, 89%) of subjects with AKI. Median follow up for all subjects was 4.66 years. The cumulative incidence of CKD at 5 years following surgery for subjects with and without post CS AKI was 12% (95% CI:7%-18%) and 7% (95% CI:4%-11%), respectively (Figure). The corresponding hazard ratio was 1.8 (95% CI: 0.8-3.7).

Conclusion: These pilot data indicate that CHD survivors with AKI following CS have an increased risk for CKD. More analysis is forthcoming to elucidate the relationship of post CS AKI and CKD; an important clinical relationship given the potential that AKI is a modifiable exposure and the longitudinal health burden associated with CKD.

MP1-8

Aortic valve repair in rheumatic disease and left ventricular hypertrophy: what's new in developing countries?

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Objective: Left ventricular hypertrophy frequently accompanies the progression of aortic valve disease in children and young adults.

Unobstructed left ventricular outflow tract with a competent aortic valve should help ameliorate both concentric and eccentric left ventricular hypertrophy that most often accompanies the pre-existing aortic valve pathology.

Methods: 147 patients, median age 14 years, underwent aortic valve repair.

Aortic stenosis was the lesion in 62 patients and aortic insufficiency or mixed lesion in 85 patients.

The aortic valve was inspected for stenosis, calcification, perforation, annular dilatation, leaflet prolapse, and deficient or retracted leaflet tissue. Operative findings included commissural fusion, cusp thickening, calcification in cusps, cusp perforation, cusp prolapse, rolling of the cusps. A variety of reparative procedures were performed, like commissurotomy, cuspal thinning, subcommissural annuloplasty, commissural plication, closure of the perforation and decalcification of cusps.

Patients were prospectively followed with clinically and echocardiographic assessment for a median of 3 years.

Preoperative and all postoperative echocardiograms were reviewed to analyze the hypertrophy grade of left ventricle. Decrease of left ventricular hypertrophy occurred after the first year, with continued gradual decline over follow-up period.

Results: No hospital death occurs. No early mortality. 7 patients required reoperations due to the progression of rheumatic disease. There were no thromboembolic complications, hemolysis or infective endocarditis in the operative survivors.

Aortic valve repair in young adults result in a decreased left ventricle mass index at the most recent follow-up. Regression of left ventricle hypertrophy in children following the repair procedure occurred regardless of the timing of the operation. Children and young adults demonstrated a similar response in left ventricle hypertrophy regression within the first year post-operation followed by a steady decline throughout the course of the follow-up period.

Conclusion: The late survival after aortic valve repair was excellent and valve-related complications were minimal.

The repair procedure results in significant left ventricular hypertrophy regression during follow-up period. Indices of left ventricular dilatation and hypertrophy regress after repair when the operation precedes important deterioration in preoperative ventricular function.

MP1-9

Lowered oral anticoagulation in young pregnant women carrier of mechanical aortic valve prostheses in developing countries

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Objective: Newer-generation bileaflet mechanical aortic valve used for valve replacement shows success in allowing young pregnant women to be safely and effectively managed with low doses of anticoagulant therapy, in developing countries.

The aim of the study was to demonstrate tolerance of an inconsistent INR level, without increase in thromboembolic events, in pregnant girls underwent aortic valve replacement with mechanical prostheses.

Methods: A population of 324 young pregnant women with mechanical aortic valve prostheses were consulted.

Between 6–12 weeks of pregnancy, warfarin has been substituted with unfractionated heparin 17.500–20.000 units every 12 hours. During the second and third trimester patients received lower-dose warfarin (INR 1.5–2.0). Low risk patients received 81 mg aspirin daily. High risk patients received 325 mg aspirin daily. Risk factors were: heart rhythm problems, left ventricular dysfunction (EF less than 30%), previous neurological events (stroke or transient ischemic attack), hypercoagulability status.

Two weeks before delivery warfarin and aspirin were discontinued and switched again to heparin. 4–6 hours after delivery standard anticoagulation therapy was resumed.

Results: Patients had significantly lower bleeding event rates.

There were no significant differences in terms of total neurological events between low-dose and standard-dose warfarin use.

The occurrence of thromboembolic events (transient ischemic attack without neurologic sequelae) in two cases might be explained by several factors. INR values were less than 1.5 when the event occurred. Patients had known risk factors: atrial fibrillation and high fibrinogen concentrations.

Conclusion: Our data demonstrate that low-dose INR does not increase the risk of thromboembolic events compared with conventional dose INR, in pregnant women carrier of mechanical aortic valve prostheses.

Lower dose anticoagulation therapy, combined with low-dose aspirin, resulted in a reduction of 60% of the incidence of adverse bleeding events without significant increases in total neurological events when used in conjunction with new design mechanical aortic valve.

MP1-10

Second stage following initial hybrid palliation for Hypoplastic left heart syndrome: Arterial or venous shunt?

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Objective: Hybrid palliation for Hypoplastic Left Heart Syndrome (HLHS) has been developed as an alternative to neonatal Norwood surgery. At the second stage, a source of pulmonary blood flow has to be established; typically in the form of superior cavopulmonary connection. Concerns have been raised

considering the significant challenge in the reconstruction of branch pulmonary arteries and the less development of the pulmonary arteries at the time of Fontan completion. In this study we compare 2 groups of patients; those who received an arterial modified Blalock Taussig shunt (2 stage Norwood I) or a venous superior cavopulmonary shunt (comprehensive Norwood I&II).

Methods: We retrospectively reviewed patients who received second stage palliation following initial hybrid. Patients were stratified according to the source of pulmonary blood supply into arterial shunt (n = 17 patients) or venous shunt (n = 26 patients). We compared results considering mortality, morbidity and findings at the pre-Fontan MRI.

Results: Age and weight at operation were lower in the arterial group (104.9 ± 57.6 Vs 184 ± 108.3 P0.001, 4 ± 1.6 Vs 5.5 ± 1.7 P0.001 respectively). All recorded surgical times were shorter in the arterial group. Considering post-operative stay, only mechanical ventilation times were in favour of the venous group (9.9 ± 5.5 Vs 7.1 ± 5.3 P 0.043). There was no difference in mortality ($2/17$ Vs $5/26$ P0.685) or incidence of complications. In the immediate post-operative period, there was tendency toward higher need for intervention on branch pulmonary arteries in the venous group while there was a higher tendency for interstage branch pulmonary artery intervention in the arterial group, however neither did reach significance. The arterial group has shown better development of branch pulmonary arteries with a higher lower lobe index (158.3 ± 39.4 Vs 114.9 ± 43.96 P0.037). There was no difference in the EF or indexed EDV (57.00 ± 8.29 Vs 59.80 ± 10.93 P0.541 and 92.7 ± 19.3 Vs 75.3 ± 25.2 p0.112 respectively).

Conclusion: Both arterial and venous shunt are viable options with mortality and morbidity results comparable to literature. The two stage Norwood I seems to be non inferior to the comprehensive/combined Norwood I&II with better development of branch pulmonary arteries at the time of Fontan completion.

MP1-11

Use of transcatheter pulmonary valve (Melody valve) for surgical mitral valve replacement in infants and children: the Italian experience

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Introduction: Infants and children with irreparable mitral valve (MV) disease have limited options for prosthetic valve replacement. Based on a decade-long experience with the externally stented bovine jugular vein graft (Melody valve) in the right ventricular outflow tract (shown to restore pulmonary valve competence and relieve obstruction) we tested the use of the Melody valve as a surgical implant in the mitral position in patients with severe mitral disease, where other prostheses were neither small enough to be implanted nor expandable as the child grows.

Methods: The medical records of patients who had undergone Melody valve implantation in the mitral position between March and November 2014 in our Institution were reviewed.

Results: Six patients had undergone surgical Melody valve implantation in the mitral position. Selected patients were between 11 months and 6 year-old, and had an average body weight of 12 kg (range 6.1–25). All patients underwent at least one previous surgical attempt to repair the valve, and still had severe stenosis and/or regurgitation. Surgically, the native MV was approached via right atriotomy and removed. Once sutured to the

mitral annulus, the Melody valve was expanded through a balloon catheter to achieve the best diameter according to the age and the patient's BSA. The balloon size used for Melody valve expansion during surgery was minimum 12, maximum 20 mm. One patient needed ECMO assistance after surgical MV repair and a Melody valve was successfully implanted few days later as possible bailout. However she did not recover from multiorgan failure and died in spite of the well-functioning Melody prosthesis. At discharge all survivals had good valvular function. At 10-month follow-up one patient had undergone catheter-based balloon expansion of the valve, while the other 4 patients were in good hemodynamic condition with mild or less mitral stenosis and/or regurgitation.

Conclusions: The short-term results of this innovative procedure are very encouraging. The paradigm of expandable MV prosthesis opens up the opportunity to carry out MV replacement in more children and at an earlier time point, and has potential to revolutionize care for infants and children with complex MV disease.

MP1-12

Implementation of a policy of delayed intervention in post-operative cardiac surgery patients with chylothorax: a prospective audit

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Introduction: Chylothorax is an uncommon complication of paediatric heart surgery. Standard management includes dietary modification, TPN, octreotide and surgical intervention. There is very little evidence to support the use of these treatments; the literature consists only of case reports and case series. A retrospective audit of outcomes in our unit using standard treatments demonstrated no significant difference between treated and untreated patients. Our department implemented a policy of delayed intervention (7 days) and we have subsequently audited the impact of this delay.

Methods: Data was reviewed from a three-year period (2011-14) from patients aged <18 who were tested for suspected chyle in pleural effusion post-surgery and survived until discharge. Positive or borderline patients were managed conservatively for 7 days prior to any interventions. Key outcomes were duration of drainage and length of hospital stay. The data was analysed against the previous cohort.

Results: Introducing a period of conservative management did not impact LOS in any group. In definite chylothoraces, delayed intervention appears to prolong the DOD; however, drainage resolved with without intervention in 59% and these patients had significantly shorter LOS compared to those who were treated. Treated patients drained for a median of 20 days after commencing treatment.

Conclusions: Since delaying treatment does not appear to impact LOS it is the authors' view that conservative management is

probably still in the patients' best interests. Since the DOD was similar in all groups the authors' strongly suspect that the chyle remains a bystander phenomenon and that drainage is caused by other processes. Identifying and treating those processes is likely to have more impact on DOD.

MP1-13

Is the 6 minute walk test or platelet count helpful in determining prognosis of patients with Eisenmenger Syndrome and Down syndrome?

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Introduction: 6MWD (6 minute walk test distance) is the standard test in the clinic for patients with Pulmonary hypertension. Doubt has been cast on its usefulness. We wished to try to determine whether this or blood tests were good measures of prognosis in our patients with Eisenmenger syndrome (ES) and Down syndrome (DS).

Methods: Patients with ES and DS had serial 6 MWD, oxygen saturations at rest and on exercise and full echocardiographic assessment from our pulmonary hypertension clinic in the last 3 years. RV function was indicated by TAPSE and lateral wall S'. Platelet count was corrected by haematocrit for the presence of erythrocytosis.

Results: Of 154 ES patients, 89 had DS (42 male) with mean resting saturations of 81%. WHO classification was a poor indicator of 6MWD (WHO I = 279 m, WHO II = 279 m, WHO III = 256 m, WHO IV = 215 m) and did not correlate well (coefficient 0.15). WHO class and 6MWD did not correlate overall with TAPSE or S' but within class III this became significant (correlation = 0.46, p = 0.02). Saturation at start and end exercise did not correlate with 6MWD, survival or RV function. 79 DS patients were commenced on therapy (phosphodiesterase inhibitor or endothelin receptor antagonist) but the change in 6MWD was not significant. TAPSE and S' correlated well with each other, but only predicted 6MWD and survival in WHO class III. 55% of DS patients had a low platelet count <150⁹/L (range 14-148) with a strong positive correlation between platelet count and rest oxygen saturations, even when corrected for Haematocrit. The lower counts were also strongly predictive of a poorer prognosis.

Conclusions: We did not find that the 6MWD or RV function was a good indicator of prognosis in DS unless within WHO group III. This suggests that until the condition is more severe, our measures of assessment are not sensitive enough to detect significant change. Platelet count is a predictor of poor prognosis, especially in DS patients, but only partly due to bone marrow hypoxia. Further measures are needed to allow better determination of prognosis in DS patients with Eisenmenger syndrome.

Table.

	Outcomes (median ((Interquartile range)) (days)					
	Duration of drainage (DOD)			Length of stay (LOS)		
	2010	2014	p value	2010	2014	p value
Positive	n=34 10 (5.75-20)	n=44 21 (9-28)	0.011	n=34 19 (13-38.5)	n=44 33 (17.5-43.75)	0.161
Borderline (all criteria)	n=5 12.0 (9.5-18)	n=2 12.5 (8-12.5)	0.693	n=5 46 (15-47.5)	n=2 21.5 (8-21.5)	0.241
Borderline (1-2 criteria)	n=20 9.5 (6.5-16.5)	n=7 20 (5-49)	0.318	n=20 17 (10.5-34.5)	n=7 28 (9-51)	0.455
Negative	n=18 14.0 (7-17.5)	n=6 11.5 (8.5-44.25)	0.546	n=18 19.5 (10.75-32.75)	n=6 33 (14.5-60)	0.442

MP1-14

Neuropsychological and intellectual assessment of 45 adolescents (GUCH patients) after congenital heart defect surgery with/without cardiopulmonary bypass.

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CHD are associated with risk for cognitive development. The cognitive impairment risk in consequence of extracorporeal circulation is still largely unknown.

Objective: neuropsychological and intellectual assessment of GUCH pts after surgical procedures for CHD in childhood with/without the use of cardiopulmonary bypass (CPB).

Material: 45 patients aged $x 17.4 \pm 19.2$ years, 35 after surgery with CPB (10 pts post-VSD; 10 post-ASD II; 2 post-ASD sin.ven. + PAPVR; 1 post- ASD II,VSD,PDA; 4 post-AVS; 2 post-PVS; 2 post-VSD, ASDII; 1 post-ASD II, TVR; 1 post-AVR, AVS; 1 post-AVR; 1 Cor triatriatum) and 10pts without CPB (all post-CoAo).

Methods: Wechsler/Raven Intelligence Scales, clinical trials assessing memory, attention, praxis, abstract thinking and visuospatial functions, WCST.

Results: Only 5 pts scored normal on all the tests (3 post-CPB pts = 8.5%, 2 post-VSD, 1 post-ASD sin.ven. + PAPVR; 2 non-CPB pts = 20%, 2 post-CoAo). Post-CPB pts obtained 75.5% normal scores, while non-CPB group - 81.3%. The non-CPB group scored significantly lower in visuospatial functions (70% subnormal scores vs. 42% in CPB pts) and verbal abstract reasoning (30% subnormal scores vs. 5.7% in EC pts). Frontal impairment was the most common in both groups (70% in non-CPB, 60% CPB pts), formal verbal fluency generating the worst results (62% CPB/50% non-CPB pts below normal), WCST (20% non-CPB/14% CPB pts below normal), and auditory verbal learning (20% non-CPB/14% CPB pts below normal). In the CPB group, intellectual development disruption was more common (28%), with intellectual disability in 2 pts, and disharmony typical for organic dysfunction in 8 pts (1 pt in non-CPB group). Medium IQ score was slightly higher in post-CPB group (94 vs. 102 IQ points).

Conclusions: 1. Regardless of the CPB use, executive function impairments are the most common neurodevelopmental abnormalities among GUCH patients, being more frequent and severe in post-CPB pts. 2. General intellectual development in post-CPB pts is more commonly disturbed, but associated with higher total scores than in non-CPB pts. 3. Visuospatial and verbal memory dysfunction is significantly more common in non-CPB group, suggesting a possible association with defect type (all CoAo pts). The study was implemented using funds allocated by the National Center of Science.

MP1-15

Physical Activity in Patients with Congenital Heart Disease. Evaluation of Subjective and Objective Data on Activity and Exercise Tolerance

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Introduction: Data concerning physical activity in patients with CHD is scarce. Comparative evaluation of self-reported physical activity and cardiopulmonary exercise tolerance measurements at our patient population was performed.

Methods: In 2014 199 patients, median age 26 (8-69) years, 102 female (51%), were examined by cardiopulmonary exercise testing (CPET) during their routine check-up at our outpatient department. 173 patients reached physical capacity, sufficient for analysis. Predicted VO₂max was calculated accordingly to Cooper (1984, 2000). A questionnaire on physical activity was filled out by the patients. Results were compared with 2 studies on healthy German children (KiGGS n = 17,000) and adults (DEGS1 n = 8152).

Results: 95 patients (52.2%) answered to be active in sports. 32% of the active and 31% of the non-active patients showed interest in a CHD - sports group, supervised by a physician. Patients with CHD were found to be less active in sports than healthy subjects and this gap was larger in male patients. The largest differences were seen in women 18-29 years (30% active

with CHD vs. 74% active healthy women) and men 30-39 years (40% active with CHD vs. 72% active healthy men). In median patients stated to be physically active 3 hours per week. Time spent with physical activity correlated with VO₂max ($r = 0.316$ p: 0.01).

Reasons stated for absence of activity in sports: not interested (31%), lack of time (12%), requirements too high (28%). Of all school children 9% stated that their parents did not approve of physical activity. The teacher was in no case given as a reason not to participate in school sports.

Self-reported fitness correlated to VO₂max: very good: 91%, good: 84%, moderate: 78%, not very good: 66%, not good at all: 74%.

Conclusions: Physical activity in patients with CHD should be encouraged more actively since lack of time and interest are main reasons. CHD- sport groups should be established to provide adequate supervision and level of physical requirements to achieve a higher percentage of physical activity in the CHD population.

MP1-16

Regional differences in WT-1 and Tcf21 expression during ventricular development: implications for myocardial compaction

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Background: Morphological and functional differences of the right and left ventricle are apparent in the adult human heart. A differential contribution of cardiac fibroblasts and smooth muscle cells (populations of epicardium-derived cells) to each ventricle may account for part of the morphological-functional disparity. Wilms Tumor-1 (WT-1) is expressed in a.o. epicardial cells and its derivatives during cardiac development. Epicardial contributions are disrupted in mice deficient of the receptor for platelet derived growth factor alpha (PDGFR alpha).

Aim: To study left-to-right differences in relation to epicardial derivatives and the development of compact ventricular myocardium, in wildtype and in PDGFR alpha knockout embryos.

Methods: Wildtype and Wt1CreERT2/+ reporter mice were used to study WT-1 expressing cells, and Tcf21lacZ/+ reporter mice, general and epicardial specific PDGFR α null mice to study the formation of the cardiac fibroblast population.

Results: After covering the heart, epicardial cells were first observed at the inner curvature, the right ventricle postero-lateral and left ventricle apical wall. Later, WT-1+ cells were present in the walls of both ventricles, but more pronounced in the left ventricle. Within the right ventricle, WT-1+ and Tcf21-lacZ+ cell distribution was more pronounced in the anterior or inlet part. A gradual increase in myocardial wall thickness was observed early in the left ventricle and at later stages in the right ventricle. PDGFR α null mouse models show a deficient epicardium and diminished number of WT-1/Tcf21lacZ+ cells, most pronounced in the right ventricle. The normal increase in myocardial thickening was severely impaired in PDGFR α null mice, most pronounced in the right ventricular inlet.

Conclusions: During normal heart development, spatio-temporal differences in contribution of WT-1 and Tcf21lacZ positive cells to right versus left ventricular myocardium occur parallel to myocardial thickening. PDGFR α null mice show no myocardial compaction, most outspoken in the right ventricle. Our findings may relate to lateralized differences in ventricular (patho)morphology in humans.

MP1-17

A Rare Cardiac Pathology: The Levoatriocardinal Vein

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Background: Levoatriocardinal vein (LACV) is a rare cardiac pathology that represents a connection between pulmonary venous and cardinal systems. LACV is commonly associated with left-sided obstructive conditions but can be seen with normal intracardiac anatomy and pulmonary venous return. The aim of this study is to discuss the morphological and clinical characteristics, diagnostic methods and outcomes of cases with LACV.

Patients and Methods: An eleven patients with LACV between 2010–2014 were examined retrospectively. The presence of LACV was confirmed with echocardiography, 128-Slice Dual Source CT Angiography and catheter angiography in all patients. The primary obstructive lesion, associated with cardiac defects and the integrity of interatrial septum, was determined for each of the patients with left-sided obstructions.

Results: The overall mean age was 79 ± 1.83 days (range 1 day–390 days), mean weight was 4.4 ± 0.4 kilograms (range, 2–8 kg). Seven the patients were female. The age at presentation was under one year in 81.8% of the patients. Nine patients had left-sided obstructions and two patients had normal intracardiac anatomy and pulmonary venous returns. All of the nine patients with left-sided obstructions were referred to our hospital because of tachypnea, respiratory distress and murmurs. In all of the patients with left-sided obstructions, LACV was demonstrated initially with echocardiographic evaluation. Atrial septum was restrictive or intact in the patients with left-sided obstructions. LACV originated directly from the left atrium in all of the patients.

Conclusions: LACV is an extremely rare cardiac pathology, existing almost exclusively in patients with left-sided obstructive lesions. It is thought to form an alternative route for left atrial blood flow when the interatrial septum is intact or restrictive. However, the presence of an interatrial or alternative shunt with LACV may postpone clinical presentation. LACV must be kept in mind for patients with left-sided obstructions. It may also be seen in patients with normal intracardiac anatomy and pulmonary venous return.

MP1-18

Reactive Hyperemia Index and Detection of Endothelial Dysfunction after Acute Leukemia Treatment in Children

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Introduction: Endothelial dysfunction (ED) is thought to be an important factor in the development of atherosclerosis, hypertension, and heart failure. Reactive Hyperemic Index (RHI) is considered as an indicator of endothelial function. This plethysmographic method is based on noninvasive assessment of endothelium-dependent changes in vascular tone (PAT) in patient fingertips. Highly efficient treatment protocols of acute lymphoblastic leukemia (ALL) have enlarged the number of leukemia survivors in children over the last decades. ALL survivors are among those at increased risk of cardiovascular complications. Early identification of impaired vascular health may allow for interventions to improve these outcomes. This study was to evaluate vascular health using peripheral artery tonometry and specific biochemical markers in pediatric ALL survivors and compare results with healthy controls (HC).

Materials and methods: Following approval by the institutional review board 45 eligible participants were enrolled in the study 26 ALL patients (14.6 ± 3.4 yrs) matched with 19 HC (19.9 ± 4.5 yrs). Endo-PAT recorder was used for the determination of RHI as well as specific biochemical markers of endothelial function were assessed (hsCRP, ADMA, E-selectin, VCAM). RHI was evaluated in ALL children and further compared with HC.

Results: Significantly lower RHI were revealed in ALL patients in comparison with HC (1.57 ± 0.52 and 2.03 ± 0.55 ; $p \leq 0.01$). In addition, E-selectin ($p < 0.01$), asymmetric dimethylarginine ($p < 0.01$) and high sensitive CRP ($p < 0.001$), but not vascular cells adhesive molecule-1 values, were also significantly increased in the ALL subjects compared with the control group.

Conclusions: Significantly decreased RHI and elevated plasma levels of specific biochemical parameters seems to be related to ED in children after acute leukemia treatment. This combined method assessment might be a useful tool for detection of ED and stratification of cardiovascular risk in pediatric patients after acute leukemia treatment.

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MP1-19

Examining the World Heart Federation criteria for echocardiographic diagnosis of rheumatic heart disease in patients with isolated rheumatic arthritis or chorea

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Background: According to World Health Organization estimates, a minimum of 15.6 million cases with rheumatic heart disease exist with 233.000 deaths attributable to it annually; this makes it of utmost importance to early apply secondary prophylaxis in mild asymptomatic patients in order to adequately hinder the progression of a still lethal disease.

Methods: Over a period of one year, children presenting to Cairo University Hospitals with isolated acute rheumatic arthritis or chorea (all with no clinical evidence of carditis) had echocardiographic screening, then they had been categorized according to the World Heart Federation criteria for echocardiographic diagnosis of rheumatic heart disease.

Results: 54 patients with acute rheumatic fever were included (mean age 8.92 ± 1.7). 48 patients (88.8%) had arthritis; 14 of them (29.1%) fulfilled criteria for definite rheumatic heart disease (8 patients in the subcategory of pathological mitral regurgitation and at least two morphological features of rheumatic affection of the mitral valve, and 7 patients with borderline disease of both aortic and mitral valves), 11 of them (22.9%) fulfilled criteria for

borderline rheumatic heart disease (7 with pathologic mitral regurgitation, and 4 with pathologic aortic regurgitation). 6 patients (11.1%) had chorea; 4 of them (66.6%) fulfilled criteria for definite rheumatic heart disease (2 patients in the subcategory of pathological mitral regurgitation and at least two morphological features of rheumatic affection of the mitral valve, and 2 patients with borderline disease of both aortic and mitral valves), 1 of them (16.6%) fulfilled criteria for borderline rheumatic heart disease (pathologic aortic regurgitation).

Conclusion: The category of patients which is classified according to the recent World Heart Federation criteria as borderline rheumatic heart disease is frequently encountered during the primary attack of rheumatic fever especially in cases with isolated aortic valve affection; and while the current guidelines made no solid recommendation for secondary prophylaxis in this category of patients; it seems prudent to recommend it for children in high-prevalence area.

MP1-20

Genetic testing of child and adolescent relatives in a family screening program for Hypertrophic Cardiomyopathy

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Introduction: Hypertrophic cardiomyopathy (HCM) is most often autosomal dominantly inherited with incomplete penetrance and variable expressivity. The main purpose of family screening is to identify relatives who have the same disease as the proband. The 2014 ESC Guidelines on diagnosis and management of HCM recommend the screening of child relatives beginning at 10 years of age.

However, the prognostic value of identifying sarcomere gene mutations in children without phenotypic manifestations of HCM remains unclear.

Aims: We retrospectively studied the outcome of clinical screening and predictive genetic testing of child probands and relatives (<18 years of age) from families with HCM and assessed the age-related penetrance of HCM during the follow-up in these young relatives.

Methods and results: Twenty patients from 10 families were included in a family screening program for HCM (2004–2013). Patients were offered clinical examination, standard resting 12-lead ECG, ambulatory ECG monitoring, transthoracic echocardiogram and genetic testing for the 10 most common sarcomeric protein gene mutations. The diagnosis of HCM was made when left ventricular wall thickness was > 2 standard deviations over the predicted mean for body surface. “At-risk child relatives” were defined as those positive for the mutation, either they had a positive or a negative phenotype at enrollment. Two probands and 18 first-degree relatives were included (80% male; median age = 9.5 years). Fourteen child relatives were mutation carriers (70%; median age = 7.9 years), and 5 were noncarriers (25%; median age = 11.5 years). One child had unknown genetic status.

Twelve (86%) of the 14 mutation carriers were diagnosed with HCM at first evaluation.

After 6 ± 3.5 years follow-up, the two mutation carriers without phenotypic expression developed HCM at 10 and 18 years of age (14% penetrance rate). Both had mutations in the MYBPC3 gene.

Conclusions: The penetrance of HCM in phenotype-negative child relatives at risk of developing HCM was 14% after 6 years of follow-up. This underlines the relevance of the long term follow-up of mutation carriers irrespective of the presence of a positive phenotype.

MP2-1

What are the causes of death in patients with congenital heart defects? An analysis of data from the German National Register for Congenital Heart Defects

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Introduction: Thanks to progress in paediatric cardiology, cardiology, cardiac surgery and intensive care, the mortality rates of patients with congenital heart disease (CHD) was reduced significantly in the past decades. However, the mortality rates in CHD patients are still higher than in the general population. The aim of further improvements calls for a detailed analysis of the causes of death. So far, such an analysis has not been available in Germany.

Methods: The German National Register for Congenital Heart Defects (NRCHD) was scanned systematically for deceased patients. Deceased patients with a confirmed diagnosis of CHD were included. Data relating to the cardiac diagnosis, symptoms, operations, interventions, concomitant diseases and causes of death were used for analysis. Two age groups were formed (<18 years; ≥ 18 years).

Results: 133 deceased patients were included in the analysis (median age at the time of death: 20.6 ± 19.4 years; 42.1% female). Of those, 12.8% had mild, 28.6% had medium and 58.6% had complex CHD. In 117 patients, the CHD was the direct cause of death (88%), 67 (57%) of those died perioperatively. 16 patients (12%) died from non-cardiac causes. 75 patients (56.4%) died before their 18th year of life, 58 patients (43.6%) afterwards. The two age groups did not differ with respect to sex distribution ($p=0.36$), complexity of CHD ($p=0.36$), NYHA status ($p=0.27$), presence of cyanosis ($p=0.29$) or cause of death (died from the CHD: $p=0.06$). There was also no difference regarding the frequency of perioperative death ($p=0.56$). Regarding concomitant diseases, pulmonary diseases ($p=0.02$) and renal diseases ($p=0.008$) were found significantly more often in patients over 18 years.

Conclusions: The analysis of causes of death in patients with CHD enrolled in the NRCHD revealed that the deceased patients frequently had complex CHD and that the majority of them died as a consequence of their CHD. The fact that approximately two thirds of named deaths occurred perioperatively highlights the need to investigate strategies to optimise both the time of re-operation and operation methods including catheter based alternatives.

MP2-2

NOTCH 1 variants in children with aortic coarctation

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Objectives: To investigate the contribution of the genetic component in the COA development by analyzing the family histories and searching mutations in NOTCH1.

Methods: We collected 68 unrelated children with COA, diagnosed by echocardiography, cardiac catheterization, and or surgical observation. Echocardiographic data were obtained for the relatives when available. The genetic study included 51 patients with CAO. We applied a strategy of targeted mutation screening for 10 out of 34 exons of the NOTCH1 gene by direct sequencing. Control DNA was obtained from 200 healthy donors.

Results: The age of patients at the time of the study was 11.2 ± 1.2 . The sex distribution was 47 males and 21 females. The mean pressure gradient in the ascending aorta was 45.7 ± 2.6 mm Hg. COA was combined with BAV in 55.9% cases, with hypoplasia of the aortic arch 32.4% or descending aorta 13.2%; 2.9% patients had complete interruption of the aortic arch and 2.9% had subaortic stenosis. Mitral valve pathology was observed in 13.2% cases. Combinations of COA with other forms of CHD were identified in 26.5% of the cases (12 PDA, 9 VSD, 6 ASD, 1 TGA). Echocardiography data was available for 82.4% mothers and 30.9% fathers. CHD were noted in 13 parents (7 BAV, 1 VSD, 4 ASD, 1 PVS).

29 variants of the NOTCH1 gene were identified. Four of them led to amino acid exchange, but only R1279H was observed in patients 7/51 and controls 4/200 ($p=0.00026$). Exon substitutions were more representative for children with COA. For example, at least one variant in exons 12 (g.30469C/T, g.30445C/T), 23 (g.39006 G/A), 34 (g.48696 G/A, g.48901 G/A, g. 48930 G/A) was identified in 25.5% of patients and 4.0% of the controls. Intron substitutions, on the contrary, frequently observed in the controls: variant intron 29 (g.43831 T/C) in the control and study groups was identified in 75.5% and 21.6% ($p=6 \times 10^{-13}$), intron 12 (g.30591C/T) - in 70.0% and 15.7% ($p=1 \times 10^{-12}$).

Conclusions: Statistical analysis showed, that the weighted combination of substitutions gene NOTCH1 will concern this observation as favorable in 82.5% of cases in healthy donors and in 96% of patients will indicate a risk of COA.

MP2-3

Time trends of operative mortality in children with congenital heart defects (CHD), 1994-2009

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Objectives: New diagnostic tools and improved treatment options have led to increased survival of children with CHD. In the present nation-wide study including all children with CHD born in Norway 1994-2009, we have assessed time trends in operative mortality.

Methods: Information on children with CHD was ascertained from national health registers (The Medical Birth Registry of Norway and the Norwegian Cause of Death Registry) and databases (Oslo University Hospital's clinical database for congenital heart defects and the Cardiovascular Disease in Norway (CVDNOR)). All children were followed until 31 December 2012. Operative mortality was defined as all deaths that occurred during the hospitalization in which cardiac surgery was performed, regardless of length of stay or deaths occurring after discharge from the hospital within 30 days of the procedure. Time trends in operative mortality among children with cardiac surgery during the first two years of life were analyzed using Joinpoint regression program and are presented as the expected annual percent changes with 95% confidence interval (CI), describing trends by periods, using the best-fit model.

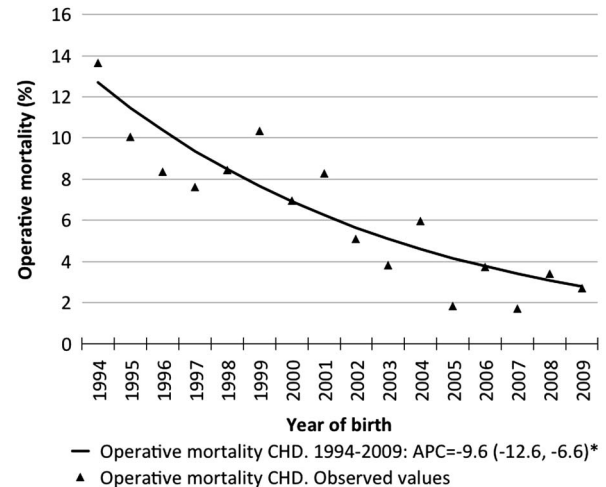


Figure.

Results: Among the 943 871 live births in Norway 1994-2009, CHD were identified in 11 272 (1.2%) children, of whom 2559 (22.7%) underwent cardiac surgery before two years of age. Comparing the first (1994-1998) and last five-year periods (2005-2009), the median time to first surgical procedure was significantly reduced from 93 days to 53 days ($p < 0.001$). The complexity of surgery described by median Aristotle score increased from the first five-year period (7.4 ± 2.5) to the last (7.8 ± 2.9) ($p < 0.001$). Among children with cardiac surgery before two years of age, only 162 (6.3%) died due to causes related to the surgical procedure. The operative mortality was significantly reduced with an annual percent change (APC) of -9.6 (95% CI: -12.6 ; -6.6) (figure).
Conclusions: Despite earlier and more complex surgery the operative mortality in children born 1994-2009 with congenital heart defects was reduced in Norway.

MP2-4

Decreasing mortality in children with severe congenital heart defects (CHD), 1994-2009

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Objectives: Despite improved diagnostic tools and new therapeutic options, congenital heart defects (CHD) are still an important cause of death in young age. In this nationwide cohort study we describe time trends in two-year mortality among all children with severe CHD born in Norway 1994-2009.

Methods: Information on children with CHD was ascertained from national health registers (The Medical Birth Registry of Norway and the Norwegian Cause of Death Registry) and databases (Oslo University Hospital's clinical database for congenital heart defects and the Cardiovascular Disease in Norway (CVDNOR)) by use of specific diagnostic codes. All children were followed until 31 December 2012. All individuals with CHD codes were assigned to only one diagnostic group by a hierarchical system and classified as having severe (heterotaxy, conotruncal defects, atrioventricular septal defects, anomalous pulmonary venous return, left ventricular outflow tract obstruction, right ventricular outflow tract obstruction (except pulmonary valve stenosis) and other complex defects) or non-severe CHD (septal defects, isolated

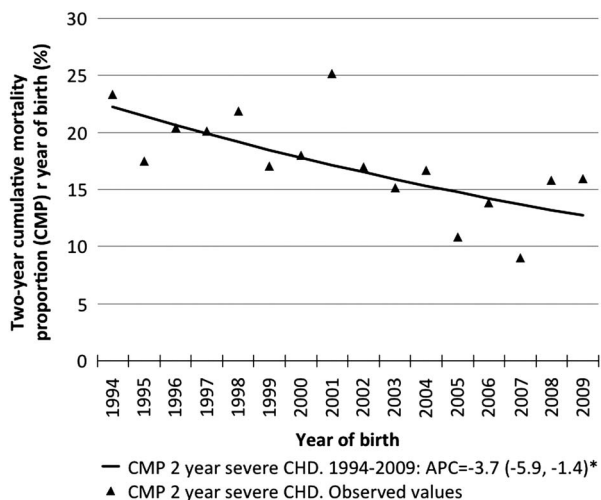


Figure.

patent ductus arteriosus, other specified CHD, pulmonary valve stenosis and unspecified CHD). Cumulative mortality proportion (CMP) was defined as the proportion of deaths regardless of cause in a specified cohort during a given period. Time trends were analyzed using Joinpoint Regression Program and are presented as the expected annual percent changes with 95% confidence interval (CI), describing trends by periods, using the best-fit model.

Results: Among the 943 871 live births in Norway 1994–2009, CHD were identified in 11 272 (1.2%) children. The proportion classified as having severe CHD was 23.7% ($n = 2\ 673$). The two-year cumulative mortality proportion ($CMP_{2\text{year}}$) during the entire study period was 17.4% ($n = 466$) in children with severe CHD and 0.3% ($n = 3\ 044$) in children without CHD. The $CMP_{2\text{year}}$ per year of birth in children with severe CHD decreased significantly during the study period with an annual percent change (APC) of -3.7% (95% CI: $-5.9, -1.4$) (figure).

Conclusions: In Norway, the two-year mortality among children with severe CHD has decreased during the period 1994–2009, but is still high compared to the general population. Further investigations are needed to clarify the factors of importance for death in children with severe CHD.

MP2-5

The efficacy and safeness of steroid combination therapy for high risk cases of Kawasaki disease

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Background: There are three scoring systems for the prediction of non-response to IVIG in Japan. Steroid combination therapy for initial treatment is recommended for high risk cases in the new guideline from the Japanese society of pediatric cardiology and cardiac surgery in 2012.

Objective: To evaluate efficacy of steroid combination therapy for high risk cases which have high scores in more than two prediction scoring systems.

Subjects and Methods: From January 2009 to 2014, a total of 54 hospitalized patients with high scores in more than two prediction scoring systems were enrolled. The patients were separated

into two groups. Group S ($n = 21$): who were administered steroid + IVIG, Group N ($n = 33$): who were administered only IVIG. Clinical data, clinical course, and circumstances of additional therapies were compared.

Results: In the patients who had high scores in two prediction scoring systems, there were fewer patients who received additional therapies in group S compared to that in group N (13% 1/8 vs. 38% 8/21). On the other hand, in patients who had high scores in all prediction scoring systems, those who received additional therapies had not decreased in group S compared to that in group N (50% 6/12 vs. 46% 6/13). However, there was no patient who had residual enlargement of the coronary artery over 2 months in group S (15% 3/20 in group N). In the patients who had high scores in all prediction scoring systems, there were more patients who had hypothermia and bradycardia as adverse effect in group S compared to that in group N (35.6 degrees C, 70 bpm vs. 36.0 degrees C, 100 bpm). However, non-administration of aspirin for 2 days while initial therapy was initiated prevented them (36.4 degrees C, 87 bpm).

Conclusions: Steroid combination therapy is beneficial for high risk cases of Kawasaki disease. In super-high risk cases those who have high scores in all prediction scoring systems, steroid combination therapy does not avoid the additional therapy and it reduces residual enlargement of coronary artery. Non-administration of aspirin while initial therapy avoids adverse effects.

MP2-6

Indirect assessment of hemodynamic parameters in congenital heart disease: comparison with data obtained by thermodilution

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Objectives: There has been concern about using predicted oxygen consumption (pVO_2) to calculate pulmonary and systemic blood flow (Q_p and Q_s , Fick method) and therefore, pulmonary vascular resistance (PVR) in patients with congenital systemic-to-pulmonary shunts. Actually, this has been considered as a major source of error. We assessed pVO_2 by five different mathematical models, and compared results to VO_2 calculated by the reverse Fick principle (cVO_2) using thermodilution technique. Subsequently, we used pVO_2 (five methods) to calculate Q_p and PVR, and compared data to those obtained by thermodilution procedure.

Methods: Thirteen patients aged one to 25 years with previous systemic-to-pulmonary shunts associated with pulmonary hypertension were subjected to right cardiac catheterization six months after surgical repair of lesions, for monitoring pulmonary hemodynamics. Indirect parameter estimation was carried out using the Fick principle, with pVO_2 according to the mathematical equations proposed by Wessel HU et al. (1969), LaFarge CG and Miettinen OS (1970), Lindahl SGE (1989), Bergstra A et al. (1995) and Lundell BPW et al. (1996). Thermodilution was used for direct parameter obtainment and assessment of cVO_2 .

Results: Indirect parameter estimation: closeness to data obtained by thermodilution.

Conclusions: The coefficients on the Table show that there is a lack of precision in the obtainment of VO_2 and Q_p using indirect methods. However, the final impact on PVR calculation seems to be not so relevant. Thus, although direct measurements are always preferable, indirect assessment of PVR may be of help if it is the only option available. Replicate PVR estimates, using different pVO_2 values should be obtained in this case.

Table.

Parameter		Wessel HU et al.	LaFarge CG & Miettinen OS	Lindahl SGE	Bergstra A et al.	Lundell BPW et al.
VO2	R ²	0.855	0.852	0.836	0.778	0.851
	p	<0.0001	<0.0001	<0.0001	<0.0001	<0.0001
Qp	R ²	0.459	0.473	0.504	0.372	0.500
	p	0.0110	0.0094	0.0066	0.0269	0.0069
PVR	R ²	0.905	0.913	0.906	0.911	0.917
	p	<0.0001	<0.0001	<0.0001	<0.0001	<0.0001

MP2-7**Accelerated rhabdomyoma regression under Everolimus therapy of the newborn infant**

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Introduction: Tuberous sclerosis complex (TSC) is characterized by the growth of benign tumors in multiple organs, caused by the disinhibition of the mammalian target of rapamycin (mTOR) protein. Recent reports on Everolimus, an mTOR inhibitor, are encouraging with size reduction of renal angiomyolipomas and subependymal giant cell astrocytomas, and rhabdomyomas (RHM). We herein report the efficacy of everolimus on large RHM in neonates compared to historic controls from our center.

Methods: Cases recently treated with Everolimus were reviewed and compared to historic controls. For comparison, the largest dimension of the largest RHM from each patient was measured and reported as a percentage compared to the earliest echocardiography study. Babies who underwent the first echocardiography imaging within 1 month of life were eligible.

Results: Studies from 4 Everolimus treated cases were compared to 10 controls. Treatment was started at 6.5 days (range 2–20) with an Everolimus initial posology of 0.1 mg po die, leading to a therapeutic serum level (5–15 ng/mL). Therapy was maintained for a median of 73 days (range 34–138) without significant side effects. Compared to historic controls, Everolimus -treated patients (Figure) had a RHM size regression rate 11.8 times faster than historic controls (linear regression slope -0.0285 vs. -0.0024 ; $p < 0.001$). Following medical therapy, 2 cases were operated for congenital heart disease, without requirement of RHM resection, 1 had a near-obstructive sub-aortic tumor shrink to non-consequential size. In addition, there was no rebound in size following Everolimus discontinuation.

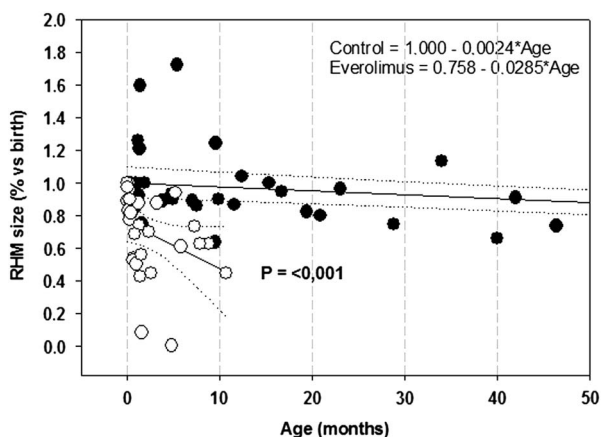


Figure.

The latter had RHM disappeared, but is still on medication to maintain efficacy on significantly reduced cerebral tumors. **Conclusion:** According to this early clinical experience, Everolimus is safe and efficacious for accelerated RHM size reduction in the neonate. Since long term effects on the newborn remain unknown this approach should be used with caution only in selective cases.

MP2-8**Influence of risk factors of cardiovascular diseases upon essential hypertension formation and target organs lesion in adolescents**

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Aim: To study the frequency of risk factors (RF) of cardiovascular diseases (CVD) in adolescents with essential arterial hypertension (EAH) at different stages of its formation and to assess their influence upon the target organs lesions.

Methods and materials: 299 adolescents with EAH at the age from 12 to 18 years old (average age 14.9 ± 2.0) were examined: 215 (71,9%) youths and 84 (28,1%) girls. According to 24-hour blood pressure monitoring adolescents with EAH were divided into three study groups: first group – 98 adolescents with "white coat hypertension" (WCH), second group – 108 patients with liable AH (LAH), third group – 93 adolescents with stable AH (st AH). RF study included the assessment of frequency of passive and active smoking, low physical activity, family history of CVD and overweight. Brain MRI was performed using MRI scanner "Magnetom-OPEN". Left ventricle (LV) function and central hemodynamics were assessed according to Echo data.

Results: Frequency of active and passive smoking was 15,7% and 36,5%, correspondingly; overweight 22,4%, family history of CVD – 51,2%, out of which family history of idiopathic hypertension (IH) was disclosed in 94,1% of cases. Covariance analysis showed that adolescents with family history of IH in relation to patients without given RF have the possibility of st AH formation higher in 5,5 times than that of WCH ($p = 0,0048$) formation and in 3,5 times - LAH ($p = 0,012$) formation, taking into account age and sex. The analogous data were got in relation to passive smoking and overweight. Family history of IH and active smoking influenced on LV hypertrophy by means of pulse pressure increase, and overweight – by increase of insulin resistance HOMA index. Overweight influenced indirectly upon hypertensive encephalopathy by means of increase of time index of systolic blood pressure at night.

Conclusion: Smoking, overweight and family history of IH favour increase of stable form of disease and target organs lesions formation in adolescents with essential AH. Preventive measures in the given adolescents allow stopping the progression of the disease and favour involution of disease symptoms.

Key words: essential arterial hypertension, adolescents, risk factors, cardiovascular diseases.

MP2-9**Dilatation of pulmonary artery correlates with severity of Marfan phenotype in childhood**

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Table 1. Comparison of prevalence and age of manifestation (Age) of dilatation of SV, mitral valve prolapse (MVP), systemic manifestation (SysMan), ectopia lentis (EL), FBN1 mutation between MFS with and without dilatation of PA.

	Patients with dilatation of PA (n = 9, 6.5 ± 5.5 y)		Patients without dilatation of PA (n = 105, 10.0 ± 5.9)		P	
	Prevalence (%)	Age (y)	Prevalence (%)	Age (y)	Prevalence	Age
SV	100	5.9 ± 4.0	69	10.0 ± 5.8	0.102	0.023
MVP	87.5	7.0 ± 5.0	53.3	10.3 ± 5.7	0.049	0.144
SysMan	62.5	7.1 ± 5.9	50.5	13.0 ± 4.3	0.717	0.092
EL	37.5	6.1 ± 4.0	18.1	7.80 ± 5.4	0.185	0.551
FBN1	45	–	69	–	0.435	

Introduction: Marfan syndrome (MFS) is an inherited connective tissue disorder with multifaceted phenotype especially in childhood. Due to risk of dilatation of sinus of valsalvae (SV) and aortic dissection frequent echocardiographic follow-up is indispensable for MFS patients. Another symptom of MFS is dilatation of pulmonary artery (PA). Measurement of diameter of PA is easy executable using echocardiography and MRI. This study aims to demonstrate the correlation of dilatation of PA with other organ manifestations in MFS in childhood to evaluate its usefulness for estimation of severity of Marfan phenotype.

Methods: We investigated 114 patients (11.1 ± 5.5 years, 64 male) with confirmed MFS. We subjected all patients to a standardized diagnostic program including echocardiography, MRI and clinical examination according to revised Ghent Criteria. We evaluated dilatation of PA according to Zilberman et al. Finally we analyzed correlation of dilatation of PA with other organ manifestations.

Results: Pediatric patients with dilatation of PA (8%) developed dilatation of SV and systemic manifestation earlier than patients without dilatation ($p < 0.05$). Mitral valve prolapse occurred more often in this patient group (Table 1).

Conclusions: We conclude that dilatation of PA correlates with age of onset of dilatation of SV which is most important for morbidity and mortality and systemic manifestation which represents a major part of the MFS phenotyp. Thus measurement of pulmonary artery in pediatric patients with MFS is essential and relevant for assessment of potential morbidity and mortality.

MP2-10

Angiotensin II type 1-receptor blocker therapy in pediatric patients with Marfan syndrome - Update after six years of experience with particular regard to tolerability

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Introduction: Marfan syndrome MFS is an inherited connective tissue disorder in which aortic root dilatation remains the significant indicator for morbidity and mortality. After recent publication of the pediatric multicenter study and publication of our monocentric data in 2013 concerning the effectiveness and tolerability of therapy with angiotensin II type 1-receptor blocker

therapy (ARB) vs beta blocker (BB) we would like to present an actual update after six years of experience.

Methods: We identified 114 pediatric patients with confirmed MFS. Indication for medical prophylaxis was found in 54 patients. All patients were subjected to standardized diagnostic program including echocardiography, MRI and clinical examination according to revised Ghent Criteria. We examined the effectiveness of medical therapy with ARB (n = 34) on the growth of the sinus valsalvae (SV) with comparison of z-scores of SV before treatment and during follow up and compared these data to patients treated with BB (n = 20) only.

Results: Treatment by ARB and BB leads to significant reduction of SV dilatation ($p < 0.05$). The deviation of SV enlargement from normal as expressed by the rate of change in z-scores was significantly reduced by a mean difference of -0.57 ± 0.65 z-scores ($p < 0.05$) under ARB therapy and by a mean difference of -0.42 ± 0.55 z-scores ($p < 0.05$) under BB therapy. The prophylactic effect of ARB and BB on aortic root dilatation is similar in both groups ($p > 0.05$). Therapy with BB was discontinued in 20% (4/20) due to intolerance. Therapy was not discontinued in the ARB group (0/34).

Conclusions: The prophylactic effect of ARB and BB in pediatric patients with MFS is similar but tolerability of ARB is clearly superior. Furthermore the inhibition of TGF- β signaling by ARB which is supposed to contribute to the pathogenesis of MFS has to be considered. Clinical study results to confirm this hypothesis are still missing.

MP2-11

Dynamic evaluation of moderate aortic stenosis in the young with treadmill exercise challenge

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Objectives: The optimal time of intervention for moderate aortic stenosis (modAS) is unknown. We sought to investigate the discriminant value of biomarker levels (NT-proBNP and hs-TnT) following exercise challenge in children with asymptomatic modAS. The aim of this study is to determine optimal timing for biomarker measurement after peak exercise and to determine the discriminant value of biomarker response to exercise challenge in a pediatric population with modAS.

Methods: Three groups (N = 8-10 each) were compared: (1) Moderate aortic stenosis (modAS) with mean LV-Ao gradient of 20 to 39 mmHg, (2) moderate aortic regurgitation without stenosis (A-Reg) and (3) control group (CTL) matched for age and gender. Subjects were excluded in the case of coexisting conditions potentially influencing biomarker levels. Blood samples were obtained at rest and at 20, 40 and 60 minutes after peak exercise. Echocardiography with 2D strain and functional data was obtained at rest and 2-3 minutes after exercise testing.

Results: Exercise performance was equivalent in all groups. Exercise testing yielded ST segment depression in 1 modAS subject. Holter monitoring showed rapid non-sustained ventricular tachycardia in another modAS subject. No difference was noted between groups for biomarkers levels at baseline. NT-proBNP increase in response to exercise was significant only in modAS, (57.8 ± 38.9 ng/L at rest vs 62.7 ± 41.8 ng/L at 40-min; $p = 0.02$). Despite relatively higher NT-proBNP in A-Reg at rest, levels did not vary after exercise (91.1 ± 62.9 ng/L vs 91.2 ± 57.73 at 60-min; $p = 0.98$) (figure 1). Hs-TnT did not yield a significant difference in any group or between groups. Strain was altered on

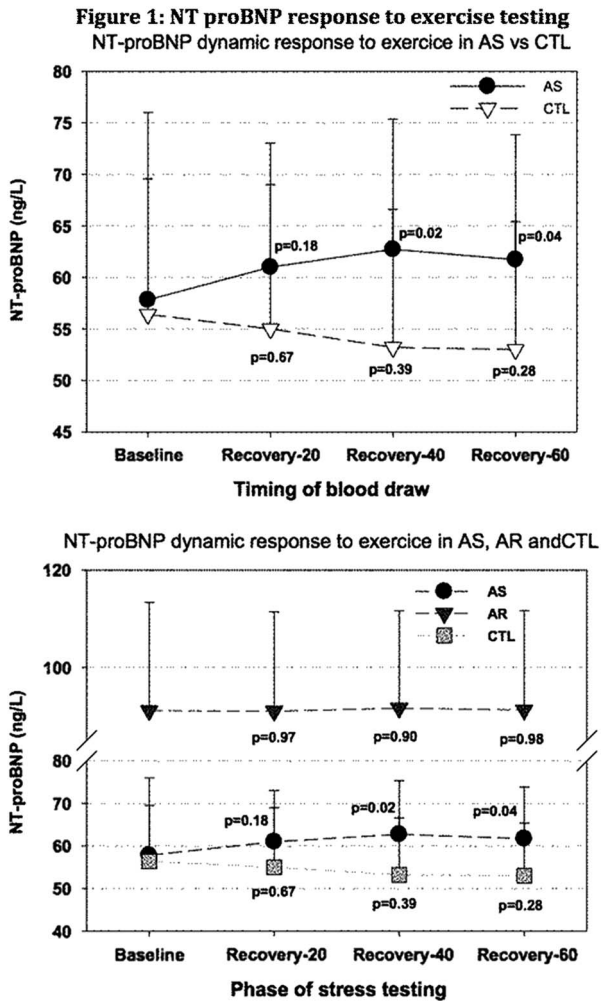


Figure 1.

global longitudinal assessment in modAS and modAR compared to CTL at rest ($-19.1 \pm 2.9\%$ and $-19.8 \pm 2.3\%$ vs $-22.3 \pm 1.3\%$, respectively; $p=0.013$) and following treadmill challenge ($-19.2 \pm 3.0\%$ and $-19.0 \pm 2.5\%$ vs $-22.6 \pm 2.0\%$; $p=0.008$). No significant differences were observed within groups after exercise challenge.

Conclusion: NT-proBNP following exercise challenge is a discriminant biomarker of modAS from A-Reg and CTL. This difference becomes significant from 40 min after peak exercise onward. 2D-Strain shows significantly reduced GLS in modAS and A-reg compared to controls at rest and after exercise challenge. This novel way to differentiate patients with moderate AS from controls offers a promising avenue for future stratification of moderate AS patients and more optimal interventional timing.

MP2-12
Malnutrition and the effect on length of hospital stay post cardiac surgery

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Introduction: Recent work considering the effect of malnutrition on length of stay, has shown improved nutrition status to within

Table. Weight for age at time of surgery, risk adjustment for congenital heart surgery and total length of stay in children < 60 months of age

	WAZ>0 (N=64)	WAZ>0 Surgery	LOS - total WAZ>0	RACHS WAZ>0	WAZ<-2 (N=132)	WAZ<-2 Surgery	LOS - Total WAZ<-2	RACHS WAZ<-2
Mean ±SD	1.39±0.49	4.76±2.33	21.44±19.8	3.01±1.63	-4.16±1.65	3.32±1.98	30.23±42.6	1.80±0.80
95%CI	1.26;1.51	4.18;5.34	16.48;26.4	2.6;3.42	-4.44;-3.87	2.97;3.65	22.8;37.5	1.6;1.9

*LOS = length of stay; WAZ = weight-for-age; RACHS = risk adjustment for congenital heart surgery.

-1 to +1 z score can significantly reduce the median hospital stay ($p < 0.005$) by up to 10 days.

Methods: Using Heartsuite database which included children < 60 months of age undergoing cardiac surgery during the time period 2012 – 2013 (n = 196) in which the following variables were included diagnosis, weight and age at time of PICU admission.

Results: In children who had WAZ (Weight for age) score < -2 [N = 132, 30.23; ± 42.69] at the time of surgery compared to those with a WAZ > 0 [N = 64 SEM 21.44; ± 19.85] had a significantly longer length of hospital stay of 8.79 days ($p = 0.0056$). Interestingly those of normal WAZ at the time of surgery had a significantly greater RACHS score ($p < 0.0001$) 3.01 ± 1.63 vs. 1.8 ± 0.08 .

Conclusion: Children with CHD are at significant risk of growth faltering which negatively impacts on post-operative course which was evident in this cohort of children. Interestingly RACHS score was significantly higher in children with a normal WAZ suggesting that the complexity of the surgery did not impact on length of stay. It could also be that dietetic resources were targeted towards those with a more severe CHD lesion perhaps at the expense of infants with traditionally less severe anomalies. In order to improve nutrition service provision to this vulnerable population group, our aim is to develop a nutrition care pathway for children with CHD targeting growth and adopting red flag where targets are not achieved with the aim of reducing length of stay.

MP2-13
Sildenafil in children with single ventricle physiology and elevated pulmonary artery pressure

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Introduction: the total cavopulmonary connection (TCPC) is the only surgical palliation in children with complex congenital heart defects and single ventricle physiology. The prerequisite for the passive pulmonary blood flow are low mean pulmonary artery pressure (MPAP) and pulmonary vascular resistance and preserved ventricular function. High risk patients prior to TCPC are those with MPAP > 15 mmHg. The use of selective pulmonary vasodilator drugs, such as phosphodiesterase-5-inhibitor Sildenafil, may lower the MPAP to acceptable value and make it possible to complete TCPC. The purpose of this retrospective study was to evaluate the effect of Sildenafil in patients with single ventricle physiology and elevated MPAP pre- and post TCPC.

Methods: from January 2011 to February 2014, 30 patients underwent TCPC. Eleven children with no morphological factors for elevated MPAP were treated with Sildenafil prior to TCPC.

All had previous palliations in early infancy, 7 with pulmonary artery banding (PAB) and 4 with Blalock-Taussig shunt. All patients had hemodynamic study prior to TCPC. The indication for starting Sildenafil was MPAP > 15 mmHg measured invasively after Glenn procedure in ICU (4 patients) or during preoperative catheterization (7 patients). The treatment was maintained for 12 ± 10 months (3–34 months) at oral doses of 2–3 mg/kg/day. In 8 patients with persistently elevated MPAP or unfavorable hemodynamic data after TCPC, Sildenafil was continued. They had follow-up catheterization.

Results: patients with higher preoperative MPAP are those after PAB, elevated end-diastolic pressure and ventricular dysfunction. The Sildenafil therapy allowed TCPC completion in all. In the follow up postoperative catheterization, a significant decrease in MPAP (18 ± 1 mmHg vs. 13 ± 2 mmHg, $p = 0.023$) was found. An improvement in the ejection fraction ($58 \pm 3\%$ vs. 67 ± 4 , $p = 0.002$) and ventricular end-diastolic pressure (VEDP) (13 ± 3 mmHg vs. 10 ± 2 mmHg, $p = 0.048$) was observed. Postoperative hemodynamic data in 3 patients allowed Sildenafil discontinuation. Satisfactory results were achieved after a minimum of 6 months Sildenafil treatment course duration. Minor adverse effects were observed in one patient, abolished after lowering the dose.

Conclusions: our preliminary result demonstrates that Sildenafil treatment improves hemodynamic parameters in high risk TCPC patients. A randomized trial can identify safety, effectiveness and long-term effect.

MP2-14

Relationship between high-risk electrocardiogram, mortality and genotype in a geographical cohort with childhood hypertrophic cardiomyopathy

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Introduction: An ECG risk score has been described, scoring amplitudes and morphological abnormalities, which predicts a very high relative risk of sudden death if the point score is >5. The genotype-phenotype relationships, and correlation with sudden death, have been studied in a geographical cohort of patients diagnosed with HCM <19 yrs of age in the West Götaland Region, Sweden.

Methods: Diagnosis was established with echocardiography, and ECG risk score was quantified from a 12-lead ECG according to Eur.Heart J 2010;31:439. DNA was analysed at international laboratories.

Results: 69 patients with childhood presentation had ECG and echocardiographic data, 64% male. 46 (67%) had familial disease, and 12 Noonan-spectrum dysmorphology. Based on ECG-risk score 29 patients were high-risk (>5 points), and 40 low-risk (<6 points). The risk score correlated with death (correlation coefficient 0.57, $p < 0.0001$); there were 8 sudden deaths in the high-risk group and none in the low risk group ($p = 0.003$). There were also two heart failure deaths in the high-risk ECG group. Familial disease was more common in the low-risk (85%) than in the high-risk group (37%). 57 of the patients had genotyping performed, 23 in the high risk group, and 34 in the group with low risk ECG. In the former a very high proportion of pathogenic mutations were found, 86%, versus 66.7% in the latter. However, both groups had significantly higher proportion of causative mutations found than 83 patients with adult disease-presentation from the same geographical area, where only 45% of patients had

clearly pathogenic mutations ($p = 0.0019$), and 21% had variants of unknown significance. The spectrum of mutations varied strikingly between groups, MYH7-mutations were prominent in the high-risk group (26.1%) but not in low-risk paediatric group (16.7%), or adult group (12.1%) with corresponding figures for MYBPC3 being 26.1%, 33.3% and 26.5% respectively. Noonan-spectrum dysmorphology appeared in both ECG groups, and were associated with RAF1 or PTNP11 mutations. Further genes affected in the high-risk group were ACTC, MYL and LAMP2.

Conclusions: The simple inexpensive ECG risk score effectively pinpoints high-risk individuals who require urgent full risk assessment, and where the yield of genetic testing is particularly high.

MP2-15

Surgical repair of Total Anomalous Pulmonary Venous Connexion in neonates and infants: Immediate results and long-term follow-up.

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Introduction: The Total Anomalous Pulmonary Venous Connexion (TAPVC) represents 1 to 3% of congenital heart diseases. An early surgical repair is mandatory due to the severity of the hemodynamic impact. Objectives were to analyse a large population of patients operated for TAPVC with a special attention given to the post-operative mortality, risks factors, morbidity and long-term follow up.

Methods: a cohort of 92 neonates and infants referred in a single center between 2000 and 2014 were retrospectively reviewed. Median age at surgery was 9.5 days (range 1–143), 65.2% were neonates, 63% of patients were mechanically ventilated before surgery. TAPVC was supracardiac in 38%, intracardiac in 15.2%, infracardiac in 37% and mixed in 9.8%. Preoperative obstruction was found in 58.7% of cases, significantly associated with infracardiac connexion (OR = 5.74, 95% CL: 2.07–15.97, $p < 0.001$). Patients with atrioventricular septal defect, heterotaxy with single ventricle, cor triatriatum or partial anomalous pulmonary venous connexion associated were excluded.

Results: Post-operative mortality was 4.3% (95% CL: 0–8.5). The median follow-up was 6.7 years (range: 0.3–13.4 years). The overall actuarial survival rate of operated patients was 86.5% (95% CL: 8.7–94.3) at one year, remaining stable beyond. Reoperations for pulmonary vein stenosis were required in 13 patients, leading to death in 23.1% (95% CL: 0.0–46.1). The first reoperation was performed after a median delay of 3.6 months (range: 0.4–127.1) from the surgical repair. In multivariate analysis, combination of heart defects associated with TAPVC ($p < 0.001$), prematurity ($p < 0.001$) and a persisting postoperative pulmonary hypertension ($p = 0.04$) were found as risk factors for death. A postoperative portal venous thrombosis occurred in 4 patients after infracardiac TAPVC repair ($n = 34$). All surviving patients were in NYHA class I after 5 years of follow up.

Conclusion: Surgical repair of TAPVC gives excellent immediate and long-term results. Mortality was limited to the first postoperative year. The main long term complication was the pulmonary vein obstruction that led to a high-risk reoperation. The occurrence of portal venous thrombosis after infracardiac

TAPVC repair reached more than 10% of patients, justifying a systematic research.

MP2-16

Comprehensive Magnetic Resonance Assessment Long-term after AtrioVentricular Septal Defect Correction

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Introduction: Atrioventricular septal defect (AVSD) contains a spectrum of anomalies with a common atrioventricular (AV) junction. Several studies report excellent long term survival with a substantial incidence of re-operation, because of left atrioventricular valve (LAVV) regurgitation. However, little data is available on cardiac function long after AVSD correction and no magnetic resonance data is available.

Methods: 34 corrected AVSD (22 partial and 12 intermediate or complete) patients were prospectively enrolled from a surgical database and 30 age related controls were included for comparison. Patients and controls underwent a comprehensive MRI examination to assess chamber volume, systolic (ejection fraction, septal to lateral delay) and diastolic (E/A ratio, E' velocity, velocity propagation (Vprop)) function and direct quantification of trans-valvular flow. Patients additionally underwent an exercise test with peak oxygen consumption (VO₂) and pro-BNP was measured.

Results: All patients were in NYHA class I or II. Nine patients used cardiac medication (beta-blocker, diuretics and/or ACE-inhibitors) and 3 had a history of atrial fibrillation. Two patients underwent LAVV replacement and 3 patients LAVV repair. Mean regurgitation fraction was 14 ± 8%. Differences between patients and volunteers are reported in the Table) and show a reduced ejection fraction, larger left atrial volume, reduced E' velocity of the lateral wall and reduced Vprop in corrected AVSD patients compared to controls. Maximum heart rate was 91 ± 13% of expected, maximum watt was 95 ± 23% of expected and peak-VO₂ was 80 ± 21% of expected. Pro-BNP ranged between 17 and 341. No relation to regurgitation fraction or parameters of systolic nor diastolic function were observed.

Conclusions: Despite the fact that corrected AVSD patients were largely asymptomatic, subtle impaired systolic (EF%) and diastolic (E' velocity of lateral wall, velocity propagation) function was observed. Although future studies are required to detect the predictive value of these changes, it stresses that careful follow-up is warranted.

Table.

	Controls	Patients	P
Age (years)	25 ± 14	27 ± 13	0.492
Gender (% male)	32	47	0.180
Body surface area (BSA)(cm ² /m ²)	1.7 ± 0.3	1.7 ± 0.3	0.311
Ejection fraction (%)	61 ± 4	55 ± 5	< 0.001
Left ventricular end diastolic volume/BSA	87 ± 13	93 ± 17	0.100
Left atrial volume/BSA	35 ± 13	48 ± 22	0.007
Regurgitation fraction (mL)	-	14 ± 8	
E/A ratio	2.5 ± 0.8	2.3 ± 1.1	0.352
Septal to lateral delay (ms)	14 ± 13	16 ± 20	0.666
E' velocity septum (cm/s)	8.6 ± 2.3	7.6 ± 3.0	0.210
E' velocity lateral wall (cm/s)	13.0 ± 3.7	9.3 ± 3.3	< 0.001
E' velocity average (cm/s)	10.8 ± 2.7	8.5 ± 3.0	0.002
Vprop (cm/s)	0.85 ± 0.20	0.70 ± 0.26	0.034

MP2-17

Anti-inflammatory effect of Obestatin in Experimental Rat Autoimmune myocarditis model

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Background and aim: Obestatin is a popular endogeneous peptide, known to have autoimmune regulatory effect on energy

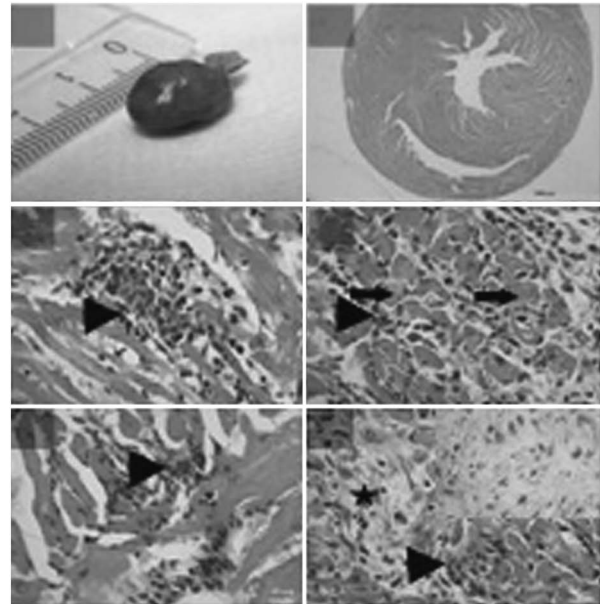


Figure 1

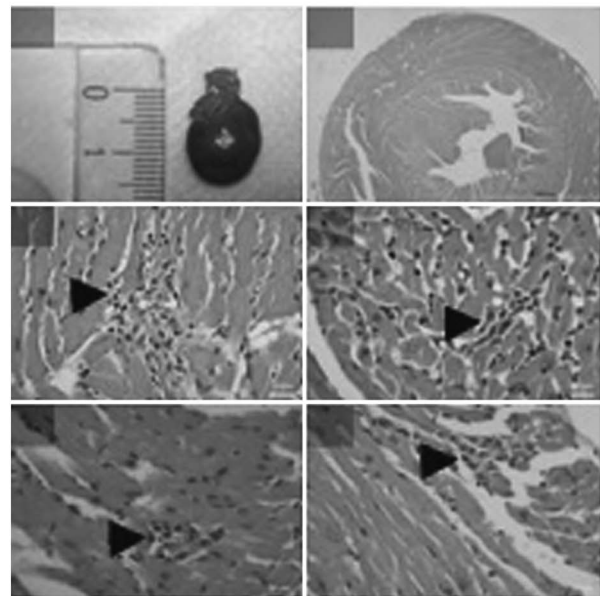


Figure 2

Figures 1 and 2.

metabolism and gastrointestinal system. The studies regarding the anti-inflammatory effect of obestatin are limited in number. In this study we aimed to show anti-inflammatory effect of obestatin in experimental rat autoimmune myocarditis model.

Material and Methods: Experimental autoimmune myocarditis was induced in Lewis rats by immunization subcutaneously twice at a 7-day interval with porcine cardiac myosin in an equal volume of complete Freund's adjuvant supplemented with *Mycobacterium tuberculosis*.

Intraperitoneal pretreatment with obestatin (50 µg/kg) was started before the induction of myocarditis, and continued for 3 weeks. The severity of myocarditis was evidenced by clinical, echocardiographic and histological findings. In addition by-products of neutrophil activation, lipid peroxidation, inflammatory and anti-inflammatory cytokines were measured in serum.

Results: Obestatin significantly ameliorated clinical and histopathological severity of autoimmune myocarditis. Therapeutic effect of obestatin in the myocarditis was associated with reduced lipid peroxidation, suppression of PMNL infiltration and enhancement of glutathione synthesis [malondialdehyde (MDA), myeloperoxidase (MPO), glutathione peroxidase (GSH) activities], inhibition of serum inflammatory (IL-1β, IL 6, TNFα), and activation of anti-inflammatory cytokines (TGFβ, IL-10). Histopathologically; there was obvious cavity dilatation and a increase in wall thickness in left ventricle, widespread lymphocytic and histocytic infiltrate (arrow) is associated with myocyte damage. Severe infiltration of relatively large mononuclear cells (solid arrow) was observed myocardium. Large areas of necrosis and myofiber degeneration were present (Figure 1) in myocarditis. These histopathological changes were all shown to be decreased in obestatin treated rats. Minimal focal inflammatory lesion is shown macroscopically in Figure 2. There was mild dilatation and a in wall minimal thickness in left ventricle. Myocardial inflammatory infiltration (arrow) was also seen but decreased in comparison to myocarditis group (figure 2).

Conclusion: This study demonstrated novel anti-inflammatory effect of obestatin in an experimental model of autoimmune myocarditis. Consequently, obestatin administration may represent a promising therapeutic approach for myocarditis and so for the dilated cardiomyopathy in the future.

MP2-18

The Use Oxygen Uptake Efficiency Slope (OUES) at different Levels of Submaximal Exercise in Children

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Introduction: The oxygen uptake efficiency slope (OUES) was first described by Baba et al in 1997 as parameter of submaximal exercise. It is unknown if there is a lower limit of exercise level the patient needs to reach to calculate OUES. We investigated whether OUES at different levels of exercise during cardiopulmonary exercise test (CPET) could still be a predictor of maximal oxygen uptake (VO₂ max) and if so, whether OUES values at different levels of exercise differed between each other.

Methods: 60 children presenting with innocent chest pain or palpitations who performed an uneventful maximal exercise test were enrolled in the study. The patient group was compared with their predicted values in order to evaluate the normality of the group. OUES was determined at 50%, 75%,

90% and 100% of the test duration. OUES/BSA was calculated for value normalisation as stated in literature. OUES50/BSA, OUES75/BSA, OUES 90/BSA and OUES100/BSA were correlated with VO₂max, VO₂max/kg, and VE/VCO₂. Secondly, the OUES50, 75 and 90 were compared to OUES at 100% of exercise.

Results: In 1 patient, OUES could not be calculated. Mean age was 11,5 ± 2,3 years, weight 43,6 ± 12,4 kg, length 151,2 ± 13,6 cm. OUES50/BSA, OUES75/BSA, OUES90/BSA and OUES 100/BSA was 1269 ± 235, 1403 ± 253, 1463 ± 262 and 1493 ± 265 respectively. VO₂max was 1668,5 ± 518 ml/min VO₂max/kg is 41,3 ± 9,7 ml/kg/min. Correlation between OUES and VO₂ max was strongest for OUES100/BSA (Pearson 0,732, P < 0,001), followed by OUES90/BSA (0,709, P < 0,001), OUES75/BSA (0,665, P < 0,001) and OUES50/BSA (0,614, P < 0,001). OUES50/BSA, OUES75/BSA and OUES90/BSA were different from OUES100/BSA. There was no correlation between OUES values and VE/VCO₂.

Conclusion: Even when children have only performed 50% of maximal exercise, the OUES/BSA value at this level of exercise has strong positive correlation with VO₂max/kg. The correlation however becomes stronger if the child can perform a nearly maximal exercise test. There is significant difference between the OUES values at different levels of submaximal exercise. This finding indicates that the reference values for OUES50, OUES75, OUES90 and OUES100 are different.

MP2-19

Variability of different Z-score calculations of the aortic sinus of Valsalva in Marfan syndrome and in healthy children

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Introduction: Aortic root aneurysm is one of the key diagnostic criteria of Marfan syndrome, and its rate of dilatation determines clinical course and management. 2D-echocardiographic dimensions of the sinus of Valsalva (SoV) are standardised for age, gender and body size, and are expressed as Z-scores, with values ≥ 2 suggestive of aortic root dilatation. The aim of this study was to assess the potential variation of different published Z-score algorithms used in clinical practice.

Methods: 23 patients with fibrillin-1 mutation-positive Marfan syndrome (according to the revised Ghent nosology) (48% female, 11.1 ± 4.1 years) and 23 healthy children (56% female, 11.5 ± 3.7 years) were assessed retrospectively. Echocardiographic measurements were performed using both inner edge and leading edge conventions at end-systole and end-diastole. Z-scores were calculated using published methods of Warren et al. (Halifax), Pettersen et al. (Detroit), Daubeney et al. (Wessex), Gautier et al. (Paris), and the recently published Z-score equations based on the largest cohort of children and adults (849 subjects) by Campens et al.

Results: The mean diameter of SoV was significantly larger in the Marfan cohort than in healthy children (30 ± 6.3 mm vs 23 ± 3.4 mm, p < 0.01) with mean Z-scores varying from 1.6 ± 1.50 (Paris) to 2.77 ± 2.09 (Wessex) in the Marfan group compared to -0.95 ± 1.01 (Paris) to -0.11 ± 0.85 (Halifax) in the control group. There was substantial variation in the proportion of patients fulfilling criteria for aortic root dilatation (Z ≥ 2) according to different calculations: 43% (Detroit and Paris), 57% (Halifax), 61% (Wessex), and 65% and 74% (Campens et al.). Z-scores in the control group were all less than 2, except for one value of 2.06 from the Wessex dataset.

Conclusions: The variability of Z-score results derived from the dimension of the sinus of Valsalva by different Z-score equations has important implications for the diagnosis of Marfan syndrome in clinical practice. Z-score data in children with Marfan syndrome should therefore be interpreted with caution, and further, prospective studies are required to establish the accuracy of different Z-score calculation algorithms.

MP2-20

Impact of Preoperative Coronary Anatomy Assessment with Echocardiography on Morbidity after Arterial Switch Operation of Transposition of the Great Arteries

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Background: In transposition of the great arteries (TGA), certain coronary patterns have been associated with major adverse events early after the arterial switch operation (ASO). We sought to determine the impact of preoperative echo diagnosis on the intra- and postoperative morbidity.

Method: All patients (n = 182) with TGA who underwent ASO during June 2001 to December 2013 were reviewed. Data on presumed CA preoperatively was obtained from the pediatric cardiologist's echo report. Intraoperative CA was categorized according to Yacoub's classification (type A to E). Major postoperative morbidity included one or more of the following: delayed sternum closure (DSC), prolonged (>72 hours) mechanical ventilation, reintubation, peritoneal dialysis (PD), ECMO, reoperation and readmission within 30 days after surgery.

Result: Mean age at ASO was 11.5 (1-614) days, and mean weight was 3.6 (1.9-8.4) kg. There was no postoperative death. Intraoperatively, 132 patients (73%) were found to have type A, 22 patients had type B or C or intramural (B-C-IM; 12%), and 28 patients had type D or E (D-E; 15%). Patients with type B-C-IM had increased risk for DSC (7/22 vs 14/132 in type A and 5/28 in type D-E; p = 0.04), PD (4/22 vs 8/132 and 1/28; p = 0.05), prolonged ventilation (7/22 vs 13/132 vs 4/28; p = 0.02), and ECMO (2/22 vs 0/131 and 1/28; p = 0.01). Preoperative echo assessment of CA was available in 176 patients. Within the B-C-IM group, preoperative echo raised suspicion of type A in 12 patients (i.e., wrong diagnosis, WD; 55%), whereas non-A type was suspected in 10 patients (i.e. accurate diagnosis, AD; 45%). With the exception of reoperation, which was seen only in the WD subgroup (4/12 vs 0/10 in the AD subgroup; p = 0.04), the intraoperative (cardiopulmonary bypass time and cross-clamp time) and postoperative morbidity indices were comparable in both WD and AD subgroups (p > 0.1).

Conclusion: Although there is a significant risk for early postoperative morbidity in TGA patients with single, interarterial and intramural CA, there seems to be relatively little influence of preoperative echo assessment of coronary anatomy on this morbidity burden. Further large prospective studies are warranted.

MP2-21

Use of lung planar scintigraphy in pulmonary perfusion analysis in patients with HLHS after Fontan operation and stent implantation

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Introduction: Fontan operation results in low pulsatile pulmonary blood inflow that affects blood distribution in lungs. Moreover, left pulmonary artery (LPA) stenosis is a common complication in patients with hypoplastic left heart syndrome (HLHS).

Aim: Assessment of lung perfusion in HLHS patients after Fontan operation using planar lung perfusion scintigraphy.

Material and Methods: The study was performed in 56 patients (37 boys and 19 girls) aged from 4 years 3 months to 18 years 7 months (mean 8 years); 39 pts. (70%) had stent implantation due to LPA stenosis prior to Fontan operation.

All patients underwent planar lung perfusion scintigraphy (AP and PA projection) after peripheral injection of ^{99m}Tc - MAA (macroaggregated albumin) with activity calculated by Webster's formula. Lung perfusion studies were performed with Hawkeye hybrid gamma camera; studies were processed on Xeleris workstation. Percentage share of every lobe in a global lung perfusion has been calculated after averaging of counts in anterior and posterior projection - geometric mean. The presence of pulmonary arteriovenous fistulas (PAVF) was assessed by radionuclide activity in kidneys and brain.

Results: Mean left and right lung contribution to global lung perfusion were 47% (upper lobe 19%; lower lobe 28%) and 53% (upper lobe 13%; middle lobe 23%, lower lobe 17%) respectively. Normal left lung perfusion was observed in 30 patients (53,6%), normal right lung perfusion in 21 cases (37,5%).

Severe lung hypoperfusion defined as percentage ≤ 30% of global lung perfusion was noted in 9 patients for the left and in 10 patients for the right lung. The most common finding was hypoperfusion of middle and lower lobes of right lung - 16 patients (28,6%).

Postoperative diaphragmatic paralysis resulting in decrease of the size of one lung was noted in 7 patients (12%).

Radionuclide activity in kidneys and brain suggesting PAVF was noted in 4 cases (7%).

Conclusions:

1. Patients with HLHS have pulmonary perfusion abnormalities due to non-physiological pulmonary flow, pulmonary artery narrowing, or postoperative diaphragmatic paralysis.
2. Planar lung perfusion scintigraphy seems to be reliable in postoperative evaluation of patients after the Fontan procedure.

MP3-1

Catheter-based interventions for modified Blalock-Taussig shunt obstruction: a 20 years experience

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Introduction: Thrombotic occlusion of a modified Blalock-Taussig shunt is rare, leading to life-threatening hypoxemia. Rescue percutaneous interventions may allow recanalization of the systemic-to-pulmonary shunt but data on large patients' scales are lacking. We aimed to describe safety and effectiveness of catheter-based interventions to restore modified Blalock-Taussig shunt patency.

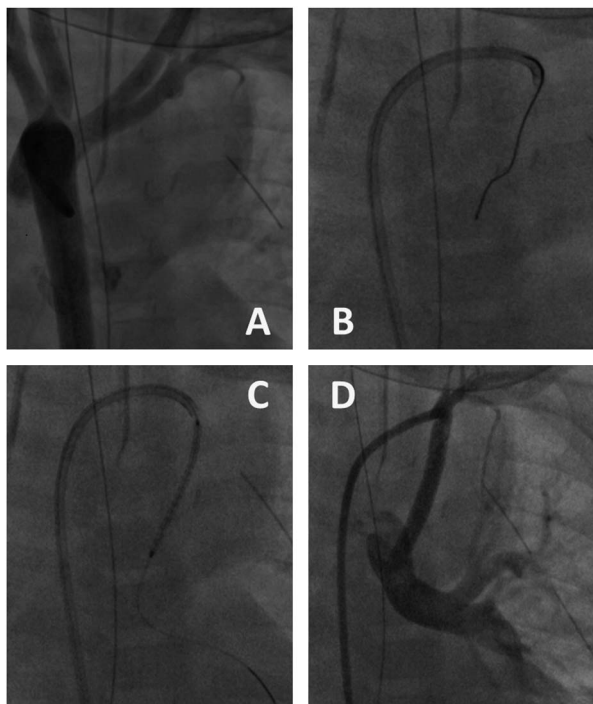


Figure.

Methods: All patients who attempted transcatheter intervention for thrombotic occlusion of a modified Blalock-Taussig shunt at our Institution from 1994 to 2014 were reviewed. Characteristics, management and outcomes of the 28 identified patients were analyzed.

Results: Thirty-three procedures were performed at a median age of 0,6 years old (range: 0,03 to 32,1 years) and a median weight of 5,8 kg (range: 2,2 to 82 kg), with a m/f sex ratio of 1.15. Percutaneous intervention consisted in 33 balloon angioplasty (100%) and 14 stent implantations (42.4%). Thrombolytic agents were also used in 6.1% cases. No peri-procedural death occurred but complications were observed in 5 patients (15.2%), including one catheter-induced transient complete atrioventricular block, one cardiac tamponade and one massive thrombo-embolic stroke. Early procedural success was obtained in 28 patients (84.8%) and remained long-lasting in 26 patients (78.8%). A young age and a low body-weight at the time of the procedure were significantly associated with procedural failure ($p=0.0364$ and $p=0.0247$ respectively).

Conclusions: Although technically challenging and carrying potential major complications, transcatheter intervention can be considered as an efficient rescue strategy to restore patency in case of thrombotic obstruction of a modified Blalock-Taussig shunt. However considering our experience and previous studies, the use of thrombolytic agents could also be considered as a first-line treatment when no anatomical cause is found to the Blalock-Taussig shunt's obstruction.

MP3-2

Percutaneous PDA Closure In Preterms Less Than 2 Kg

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Introduction: Preterms and low birth weights are the group that PDA is most common and intervention is most risky. Main aim of our study is to emphasize the effectiveness and safety of percutaneous PDA closure in the infants less than 2 kg.

Material Method: Between the dates July 1997 to October 2014 in our center 382 PDA closures were done. 18 patients less than 2 kg were included in this study. Demographic and angiographic data of the patients were reported.

Results: All the patients were symptomatic and PDA was decided to be contributor of this medical state. The median patient age 32days. The median weight of patients was 1603 gr (910-2000 gr). Mean PDA diameter was 3.2 ± 1.3 mm. Morphology of PDA: type A in 7patients, type C in 9patients, type E in 1, type B in 1 patient. Types of the devices used were: Cook coil in 2patients, ADO I in 2, ADO II in 3, ADO II-AS in 11 patients. There were no major complications reported. Left pulmonary arterial stenosis was detected in 4 patients which were all resolved in 6 months duration.

Conclusion: Preterm complications like chronic lung disease, necrotising enterocolitis etc. increase the mortality and morbidity. In order to decrease the complications early intervention is required but surgery could be too risky. Because of the risks of surgery in the recent years interventional catheterization procedures are more commonly used.

Up to our knowledge it is the only study that discuss safety and effectiveness of percutaneous PDA closure in the infants less than 2 kg. Patient population less than 2 kg are preterms and most have additional health problems that the surgery could be dangerous but catheterisation can be used safely.

MP3-3

Fever following interventional or surgical pulmonary valve replacement - SIRS like humeral and cellular immune response in the context of treating RVOT dysfunction in congenital heart defects

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Objective: Pulmonary valve implantation (PVI) led to a substantial increase in life expectancy for many patients with congenital heart disease. Transcatheter pulmonary valve implantation (TPVI) has evolved to an alternative when suitable, reached worldwide clinical acceptance and routine procedure status. By the help of TPVI, dysfunction of right ventricular outflow tract (RVOT) could be successfully treated in more than 3000 patients worldwide so far. Currently used pulmonary valve conduits possess xenograft materials (porcine or bovine). Procedure-induced and/or xenograft-tissue-induced febrile systemic inflammatory response syndrome (SIRS) could provoke a subsequent dysfunction of the engrafted valve. Hence, in this study, the immune response is analyzed in detail before and after TPVI and surgical pulmonary valve replacement (SPVI).

Methods: At defined time points before and after RVOT intervention (preliminary stent implantation ($n=20$); TPVI ($n=20$) or SPVI ($n=10$)), clinical data and blood samples of adults and infants (median age: 17 years) are compared regarding changes in general-clinical (heart and breathing frequencies, temp.), unspecific para-clinical (CBC, LDC, CrP) and specific para-clinical parameters (interleukin (IL) 12p70/1beta/2/6/8/10, TNFa, PCT, leukocyte analysis by flow cytometry).

Results: After ethics board approval 20 percutaneous and 10 surgical treated patients were included in the study from August 2013 to November 2014. First results of analysis of immune response reveal an increase of IL 6 and IL 8 concentrations within the first 72 h after the procedure (pre-stenting and both types of pulmonary valve implantation). Granulocytes count, especially neutrophils, raises 24 h after percutaneous or surgical valve implantation. Monocyte count does not change after each treatment option. Lymphocytes counts decrease within the first 24 h after both treatment options. This is explained by T cell diminishment.

Conclusion: The analysis of specific and unspecific parameters of humeral and cellular immunity leads to the characterization of the febril immune response after TPVI or SPVI. The number of included patients limits statistical relevancy by now, but trends of specific cellular populations changes are pointed out. Further investigations will be realized to understand this phenomenon in detail and to what extend this is provoked by xenograft material or intervention itself.

MP3-4

New Therapeutic Strategies for Patients with Atrial Septal Defect and Severe Pulmonary Arterial Hypertension (PAH): Combination of PAH-Specific Medical Therapy and Catheter Intervention

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Background: Therapeutic strategy for atrial septal defect (ASD) patients with severe pulmonary artery hypertension (PAH) still remains controversial. Recent advances in medical therapy for PAH and catheter intervention may provide new therapeutic approaches in these patients. Purpose of this study was to assess the efficacy of combination therapy (transcatheter atrial septal defect (ASD) closure and pulmonary arterial hypertension (PAH)-specific drugs therapy) in ASD patients with PAH.

Methods: Thirty-seven consecutive ASD patients with PAH who underwent transcatheter closure were enrolled (median age, 65 years). PAH was defined as a mean pulmonary artery pressure (PAP) ≥ 25 mm Hg at cardiac catheterization. Systolic PAP estimated by echocardiography was evaluated at baseline and follow-up (median, 25 months).

Results: As shown in Figure, systolic PAP improved significantly at follow-up examination (median, 57 to 35 mm Hg; $p < 0.001$)

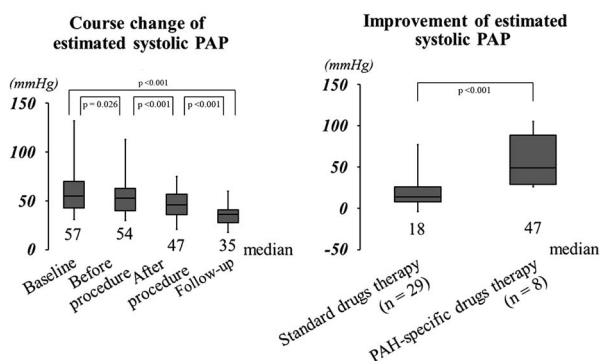


Figure.

compared with baseline examination. Reduction of systolic PAP was significantly greater in patients with PAH-specific drugs therapy ($n=8$) than in those without drugs therapy ($n=29$) (median, 47 vs. 18 mm Hg; $p < 0.001$).

Conclusions: Even in patients severe PAH, combination of transcatheter closure and disease-targeted therapy can contribute the hemodynamic improvement and expand the therapeutic possibilities in ASD patients with PAH.

MP3-5

A word of caution: Diabolic behavior of AndraStents® - Inflation of balloon during implantation leads to “diabolo”-misconfiguration

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Objective: Implantation of transcatheter valves requires almost every time pre-stenting of the right ventricular outflow tract (RVOT) with large caliber stents. Widely used are cheatham platinum (CP) stents, in some instances ePTFE covered CP stents. Alternatively other large caliber stents like the AndraStent® can be used. We report potentially fatal complications during the implantation of AndraStents® in five cases from two different institutions.

Method: Retrospective analysis of cases.

Results: In five cases a misconfiguration of the AndraStent® occurred forming a “diabolo”. During inflation of the balloon the stent expanded at both ends but the middle part remained unexpanded. In several cases higher inflation pressures lead to a burst of the balloon. Different measures where taken to rescue the stent. In one case the stent was expanded with a high-pressure balloon, in another the stent was retrieved by surgery and in one case during attempted interventional retrieval of the embolized stent eventually a perforation with a fatal bleeding occurred.

Conclusion: Misconfiguration of AndraStents® during implantation can lead to potentially fatal complications. Several rescue strategies including stent placement or re-dilatation with high pressure balloons or surgery should be available.

MP3-6

Stent Implantation in Aortic Coarctation in Turner Syndrome: a Safety Analysis

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Introduction: Stent implantation has proven to be a safe and effective way for the treatment of coarctation in adolescents and adults. Turner Syndrome is a genetic disorder marked by a partial or complete absence of one X-chromosome. CoA occurs in approximately 17% of Turner patients. Also, the aortic wall composition is abnormal in Turner Syndrome (TS). Changes in vascular smooth muscle cells, elastin and collagen fiber and cystic medial wall necrosis, as found in

Table.

Patient number	Baseline characteristics					Before stent implantation				
	Age	previous surgery	Age at surgery	Age at stenting	covered stent (1=yes)	CoA size (mm)	diaphragm diameter	PG (mm Hg)	CoA size (mm)	PG (mm Hg)
1	51	1	27	46	1	11	†	34	16	4
2	41	0	*	32	1	12	†	25	17	†
3	31	1	0	21	1	12	†	35	17	†
4	61	0	*	60	1	9	22	15	19	0
5	24	1	0	11	0	6	11	†	10	0
6	24	1	0	9	0	6	12	†	13	0
7	10	0	*	7	1	5	13	†	†	0
8	26	0	*	16	0	8	14	†	†	0
9	53	0	*	47	1	11	†	45	†	9
10	19	1	0	12	1	8	†	30	12	3
11	17	0	*	13	0	†	†	10	†	0
12	43	1	0	37	0	6	13	29	14	0
13	50	0	*	35	0	†	†	19	†	†
14	11	1	0	7	†	†	†	†	†	†
15	26	1	1	11	†	†	†	24	†	20
16	6	1	0	0	†	2	6	12	4	11
17	16	0	*	11	1	11	14	19	12	0
18					†	7	14	6	10,5	4
Mean	29,2		*0	22,0		8,1	13,2	23,3	13,1	*0
SD	16,7		**0 to 27	17,4		3,0	4,1	11,2	4,2	**0-20

†:unknown * Median **Range

†:unknown
* Median
**Range

Marfan syndrome, make patients with TS vulnerable to aortic dissection. Therefore concerns exist about coarctation stenting in TS.

Methods: In this multi-center retrospective study, all patients with Turner syndrome and a coarctation of the aorta, treated with percutaneous stent implantation, were included. The adverse events at short and long-term follow-up and qualitative parameters concerning the stent implantation were assessed.

Results: A total of 17 patients with 18 stenting procedures from 8 centers were included. Age at intervention (mean ± sd) was 22 (±17,4) years. The coarctation diameter pre-intervention was 8,1 (±3) mm and increased to 13,1 (±4,2). The peak gradient decreased (median (range)) from 24 (6-45) to 0 (0-20) mmHg. Baseline and intervention characteristics are included in table 1. At short term (<30days after the intervention) dissection of the descending aorta occurred in 2 patients (11%), of which 1 was fatal (5,5%). Both of these patients underwent covered stent implantation. The mean follow-up was approximately 8 years. Two additional patients died during follow-up due to non-related cardiac causes. One patient died awaiting aortic valve replacement showing a large thrombus located near the mitral valve at autopsy, the other patient died due to progressive heart failure.

Conclusions: These results suggest that, despite the use of covered stents, coarctation stenting in TS carries a higher risk than in non TS individuals. A prospective study is warranted, including

evaluation with pre and post procedural CT or MR imaging to better define specific risk factors.

MP3-7

The Association of Silent Cerebral Emboli Following Percutaneous Atrial Septal Defect Closure in Pediatric Patients: A Diffusion Weighted Magnetic Resonance Imaging Study

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Objectives: The aim of this prospective study was to investigate the incidence of silent cerebrovascular embolic events associated with percutaneous closure of atrial septal defect (ASD) in pediatric patients.

Material and Methods: 23 consecutive pediatric patients (12 male and 11 female) admitted for transcatheter closure of ASD with the mean age of 10.4 ± 3.8 years (range 4-17 years) were recruited for the study. The patients were scanned with a 1.5 Tesla clinical magnetic resonance imaging (MRI) system (Magnetom, Aera, Siemens Healthcare, Erlangen, Germany). Patients underwent first cranial MRI on the day before the procedure and a control MRI was acquired within 24 hours following the heart catheterization. MR examinations included turbo spin echo fluid-attenuated inversion recovery sequence and diffusion weighted imaging technique done with single shot echo planar spin echo sequence. The transcatheter closure of ASD procedures were performed by 3 expert interventional cardiologists. Amplatzer Septal Occluder (ASO) device was implemented for the closure of the defect. No iodinated contrast medium was administered in course of procedure.

Results: Preprocedural MRI of 2 patients revealed nonspecific hyperintense white matter lesions on FLAIR images with increased diffusion, which were considered to be older ischemic lesions associated with previously occurred paradoxical embolism. None of the patients had diffusion restricted cerebral lesions resembling micro embolic infarctions.

Conclusion: The current study suggests that percutaneous closure of the ASD when performed by experienced hands without administration of iodinated contrast medium, may be free of cerebral micro embolization in pediatric patients. However,

Table.

Characteristics	Values
Age (years)	10.4 ± 3.8 (4-16)
Gender (male)	12 (52.2%)
Weight (kg)	40.0 ± 14.4 (14-60)
ASD diameter (mm) (TTE)	16.0 ± 5.5 (9-26)
ASD diameter (mm) (TEE, Stop-flow)	16.6 ± 4.9 (9-25)
Device diameter (mm)	16.8 ± 5.1 (9-25)
Qp/Qs	2.2 ± 0.5 (1.5-3.2)
PVR (Wood/U/m2)	1.67 ± 0.38 (0.9-2.4)
Duration of the procedure (min)	44.9 ± 15.8 (14.0-85.0)
Fluoroscopy time (min)	9.7 ± 4.5 (3.0-18.0)

due to our relatively small patient group, further studies with larger patient groups and wider age range are needed for further investigation of the incidence of heart catheterization and its risk factors.

MP3-8

Safety of device closure of secundum atrial septal defects in children – does the weight at time of procedure matter?

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Background: Transcatheter closure of secundum atrial septal defect (ASD) is nowadays the main treatment option in many centers worldwide. In symptomatic infants or in those with certain comorbidities, there is a need for early intervention. Limited data are available about the procedural safety in infants below 15 kg of weight.

Methods: Retrospective review of all patients referred to our center for ASD closure by device between January 1998 and November 2014. Major complications included death, cardiac or respiratory arrest, stroke, device embolization and erosion, need for emergency surgery or recatheterisation due to other procedure-related complications, significant pleural/pericardial effusion requiring intervention, persistent arrhythmia or intraprocedural arrhythmia requiring cardioversion/resuscitation, postprocedural significant valvular insufficiency or pulmonary vein obstruction, need for transfusion due to significant bleeding, and permanent vein thrombosis. Minor complications included transient arrhythmia, significant access site hematoma, transient vein thrombosis, transient limb paresthesia, and development of postprocedural infection occurring within 48 hours. Data were retrieved from the hospital's database and from the Swedish Registry for Congenital Heart Disease (Swedcon).

Results: In total, 244 cases with age < 18 years were included. Data are summarized in Table 1. In the group < 15 kg (n = 104), there were 5 major (need of transfusion, surgical removal of device, arrhythmia requiring conversion in 2 patients, and pulmonary hypertension crisis) and 2 minor complications (infection and transient vein thrombosis). In the group > 15 kg, there were 6 major (one death related to device erosion in a 17-year female with 26 mm ASD and absent retroaortic rim, surgical removal of device, arrhythmia requiring conversion in 2 patients, and need of transfusion) and 4 minor complications (transient arrhythmia).

Conclusion: Transcatheter ASD closure appears to be safe and highly successful even in small infants.

Table 1.

	< 15 kg	> 15 kg
Number of patients	104	140
Age (years)	2.0 (0.3-5.2)	7.95 (2.1-17.6)
Sex (male/female)	38/66	58/82
Body weight (kg)	10.9 (3.6-14.8)	29.4 (15.0-87.5)
Total complications (n/%)	7/6.7	10/7.2
Major complications (n/%)	5/4.8	6/4.3
Minor complications (n/%)	2/1.9	4/2.9
Closure success rate with device (%)	99.0%	98.6

Data presented as mean (range).

MP3-9

Closure of coronary artery fistula in childhood: treatment indications and long-term follow-up

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Introduction: Coronary artery fistula (CAF) is a rare congenital anomalous connection between the coronary arteries (CA) and a cardiac chamber or great vessel bypassing the myocardial capillary network. Treatment options of symptomatic CAF consist of transcatheter or surgical closure.

Methods: Retrospective analysis of all patients with CAF in our outpatient clinics database diagnosed between 1986 - 2014 concerning significance, treatment approaches and follow-up after closure, if applicable.

Results: In a cohort of more than 25000 patients, 194 (<0.01%) were diagnosed to have CAF and were enrolled in the analysis. Median age at first diagnosis was 6 months (0d-18 y). In 79 patients (40.7%) CAF were isolated, whereas in 115 patients (59.3%) CAF were found in association with congenital heart disease (CHD). Origin of CAF was from the LCA in 77.2% with a left to right shunt in 86.5%. Treatment indication was given in 10 patients (5.2%): volume overload of the right sided cardiac chambers (n = 6, 60%), heart insufficiency (n = 2, 20%), aneurysmatic dilation of the CA with risk of ischemia (n = 2, 20%). Six patients (60%) were treated by catheter interventional approach (3 Coils, 2 Amplatzer Vascular Plugs, 1 Amplatzer Duct Occluder). Follow-up (median 7 years (2-12 years) revealed one major complication with dislocation of the occluder and thrombosis of CA one week after implantation. Persistent dilated CAs were seen in 3 patients (control angiographies in 2 and echocardiography in 1 patient) and normal CA dimensions in 2 patients (echocardiography). In these five patients, no thrombosis of CA occurred and CAF remained closed. Surgical closure of CAF was performed in 4 patients (median follow-up 2 years (0-7 y), during correction of CHD (n = 2), and, in the era before transcatheter closure of CAF, in patients with isolated CAF (before year 2000, n = 2).

Conclusions: In a pre-selected cohort of pediatric cardiology patients CAF is a very rare finding, although our numbers are probably underestimated. Intervention in childhood is rarely needed, nevertheless it is known, that small fistulas may become relevant in adulthood. Myocardial ischemia is rarely found in childhood CAF. Transcatheter closure techniques are considered the treatment of choice, especially in isolated CAF, and promote good long-term results.

MP3-10

Stent-implantation into the intraatrial septum after hybrid procedure in hypoplastic left heart syndrome with self-expanding PDA-stents

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Introduction: After bilateral pulmonary artery banding (PAB) and stent-implantation in the patent ductus arteriosus (PDA) as an alternative to the classical Norwood I surgery in neonates with hypoplastic left heart syndrome (HLHS), a restriction of the intraatrial septum occurs in approx. 50% of the cases. This can be treated interventional, the septostomy is usually performed during the comprehensive stage II.

Method: Since April 2013 we have implanted the self-expandable Sinus-superflex-DS stent, Optimed, Esslingen, in 12 patients (age median 45 days, range 5-114 days) with HLHS if the invasive gradient was higher than 5 mmHg in mean. In two cases the Sinus-superflex-DS stent 7 × 12 mm, in seven the 8 × 15 mm and in three patients the 8 × 12 mm stent was inserted through a 4 Fr sheath placed in the femoral vein.

Results: All PDA-Stents were successfully implanted in the IAS, no stent dislocation was observed. The mean gradient dropped from 15 mmHg median (range 6-20) to 2 mmHg median (range 1-4).

Discussion: The self-expanding sinus-Superflex-DS stent in open-cell design is very flexible and adjusts to the duct morphology. The pull-back mechanism allows a very precise positioning without jumping effects. All these characteristics are favourable for implanting this stent into the duct, but also into the IAS.

MP3-11

Premature Ventricular Contractions in Healthy Children: does the burden of ectopy matter?

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Background: Premature ventricular contractions (PVCs) are a common finding in healthy children. Whereas a burden of ventricular ectopy higher than 20% in adults with normal hearts was found to increase the risk of cardiovascular events and LV dysfunction, there is little knowledge about the consequences of frequent PVCs diagnosed in a healthy child.

Methods: We reviewed 24 h-Holters performed at our institution (2008-2012), and identified patients with structurally normal hearts and a burden of PVCs $\geq 10\%$ of total beats on any Holter during follow-up. Demographic data were collected and transthoracic echocardiography (TTE), ECG and 24 h-Holters at each evaluation were reviewed.

Results: A total of 47 patients (22 female; mean age 8.2 ± 6.5 y) had $19.2 \pm 10.3\%$ PVCs recorded on initial 24 h-Holter. Left bundle branch morphology was dominant in 33/45 (73%). No evidence of severe cardiomyopathy was found on initial TTE with mean shortening fraction (SF) Z-score of 0.1 ± 2.0 . Yet, 7 patients (14.9%) had decreased SF (Z-score: -2.4 to -4). On initial ECG, mean PVC coupling interval was 431.9 ± 110.5 ms, and mean PVC width was 118.6 ± 27.1 msec; runs of non-sustained or sustained ventricular tachycardia (VT) were present in 3 (6.4%) patients. On 24 h-Holter, PVCs were monomorphic in 44/47 (93.6%), ventricular bigeminy was recorded in $26.9 \pm 29.2\%$ of the total PVC time in 37/47 (78.7%) patients. SF Z-score was worse in cases with shortest coupling interval and with higher 24 h PVC proportion (figure 1). A similar trend was noted between the percentage of ventricular bigeminy and SF Z-score. During follow-up (3.9 ± 2.3 yrs) no death occurred, and a significant decrease in PVC burden was observed (from $19.2 \pm 10.3\%$ to $7.7 \pm 11.1\%$; $p < 0.001$) coupled with a trend towards improved systolic function (SF Z-score from 0.11 ± 2.04 to 0.69 ± 1.9 ; $p = 0.11$). Three patients with sustained runs of VT underwent successful PVC ablation. On final Holter, 19/47 (40%) patients had $< 1\%$ PVCs.

Conclusions: PVCs seem to be a benign finding in children with structurally normal hearts, with no reported death or severe cardiomyopathy in this small series. Yet, mild LV systolic dysfunction is observed in some patients, correlating with a higher burden of ectopy.

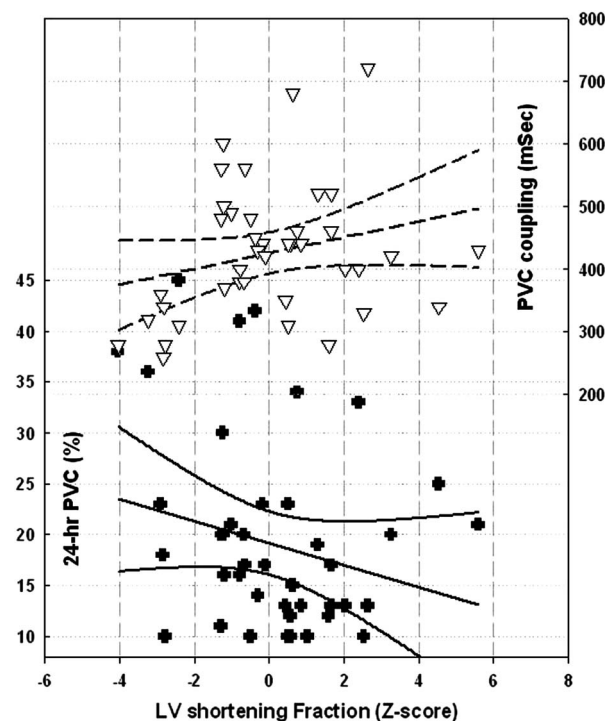


Figure 1.

MP3-12

Successful treatment with Flecainide in a patient with Andersen Tawil Syndrome Type I (ATS I) and incessant ventricular arrhythmias

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Introduction: We report a case of 15 year old girl who presented unexpectedly during anaesthesia for velopharyngoplasty with numerous polymorphic ventricular extrasystoles (VES) and short runs of VT.

Methods: ECG on admission showed sinus rhythm (SR) with regular time intervals (QTc 380) and prominent U wave. 24 hours ECG showed SR with very frequent VES (45% of all QRS complexes) 3129 bigemini, 3000 couplets, 973 triplets, 1365 short runs of VTs (2.3-12.7 sec, frequency 125-150 /min) which increased during periods of rest.

Exercise test revealed decreased VES during maximal exercise (2.9 Watt/kg) and early recovery.

Tilt test showed, during position change from supine to standing, heart rate increased and the amount of VES decreased. VES increased again in supine position.

Echocardiography and cardiac MRT showed a structural and functional normal heart.

Clinical examination revealed mild dysmorphic features known in patients with Andersen-Tawil-Syndrome (ATS) like short stature, micrognathia, hypertelorism, broad nasal bridge.

Genetic analysis revealed a heterozygote mutation in the KCNJ2 Gene (Exon 2 c.652C > T, p.Arg21⁸Tp).

Family history included unexpected death of grandfather at the age of 36 years and two maternal cousins in their early thirties.

Results: 24 hours ECG under Propranolol 3,4 mg/kg showed slight increase in the duration of VTs otherwise no significant change.

24 hours ECG under Flecainide 3,0 mg/kg showed significant reduction of arrhythmias: 12% polymorphic VES of all QRS

complexes, 1260 bigemini, 516 couplets, 12 triplets, 16 short runs of VTs (2.7–5.7 sec, frequency 111–140 /min) which increased during periods of rest.

24 hours ECG under Flecainide 6,5 mg/kgKG showed only 450 VES of two morphologies mainly during periods of rest. No VTs, triplets or couplets.

24 hours ECG after three months treatment with Flecainide 6,5 mg/kg showed only 40 VES of two morphologies. No VTs, triplets or couplets.

Conclusion: ATS I is a disorder of ventricular depolarisation caused by mutation in the KCNJ2 gene. It is characterised by a high burden of difficult to treat ventricular ectopics. In our patient treatment with Flecainide 6,5 mg/kg abolished all VT runs.

MP3-13

ICD-treatment in Swedish children and adolescents with long QT syndrome – a survey of the Swedish ICD- and Pacemaker Registry

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Background: In the congenital long QT syndrome (LQTS), mutations cause changes in the ion channels of the myocytes leading to severe arrhythmias like Torsades de Pointes. The symptoms vary from dizziness and syncope to aborted cardiac arrest (ACA) and sudden cardiac death (SCD). For patients with a high risk for ACA/SCD, implantable cardioverter defibrillator (ICD) is an option for treatment. This survey aimed to describe the pediatric LQTS-population with implanted ICDs in Sweden.

Methods: Patient data was extracted from the Swedish ICD- and Pacemaker Registry. The inclusion criteria were LQTS and implanted ICD, and 173 patients met the criteria. Informed consent was obtained from 138 patients. Medical records were reviewed and data concerning medical treatment with beta-blockers, indication for ICD and DNA analysis were collected. We compared the pediatric and the adult LQTS population.

Results: The number of ICD-implantations in patients diagnosed with LQTS in Sweden has increased exponentially since 1992. In the pediatric LQTS population, 22 patients received ICD in the age of 0–19 years. Three of these patients were found to have catecholaminergic polymorphic ventricular tachycardia (CPVT) when genetic testing was performed later. Boys were overrepresented until age 20 when it reversed. Based on medical records, the three most common indications for ICD-implantation were syncope, 55% vs 48% (age ≤ 19 vs > 19); ACA, 14% vs 31%; and primary prophylactic treatment, 32% vs 10%. At the time for symptoms indicating ICD implantation, 73% of children/adolescents were prescribed betablockers compared to 48% of the adults. DNA analysis was performed in 82%. A mutation was found in 70%, the three most common being LQT2, 38%; LQT1, 29%; and LQT3, 15%.

Conclusions: ICD-implantations in LQTS-patients are increasing, which may be due to increased awareness of the diagnosis and the choice of primary prophylactic treatment. Primary prophylactic ICD seems to be more common in children/adolescents than in adults. In contrast to current recommendations, relatively few adult patients are on betablockers before receiving ICD. Most of the LQTS patients in the Swedish ICD registry have been tested genetically and this survey also indicates that comparatively many LQT1-patients receive ICDs.

MP3-14

Clinical profile and follow-up data of patients included in the European Registry for ICD and CRT devices in Pediatrics and Adults with Congenital Heart Disease (Euripides)

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Introduction: The European Registry for ICD and CRT Devices in Pediatrics and Adults with Congenital Heart Disease (Euripides) has been existing since 2007 thanks to initiative of the AEPC, ESC and the German Competence Network Congenital Heart Disease. **Methods:** Demographic and medical data including indication and implantation details as well as obligatory yearly follow-up of treatment efficacy, complications and therapy termination were prospectively entered into the registry since 2007 using a pseudo-anonymized web-based data entry.

Results: A total of 242 ICD, 36 CRT and 6 CRT-D patients from 16 centres of 11 European countries have been included (Germany 77, Poland 66, Czech Republic 29, Finland 27, Hungary 26, Switzerland 18, Netherlands 12, others 29 pts.). At least one yearly follow-up was entered in 70% of all eligible patients. A median (IQR) follow-up period was 2.8 years.

Conclusions: Data entered allow for detailed analysis of therapy indications, efficacy and complications. Major problems are still slow data volume growth and a low number of obligatory annual follow-up reports.

MP3-15

Familial WPW and hypertrophic cardiomyopathy caused by PRKAG2 mutations: cardiac MRI and electrophysiology findings

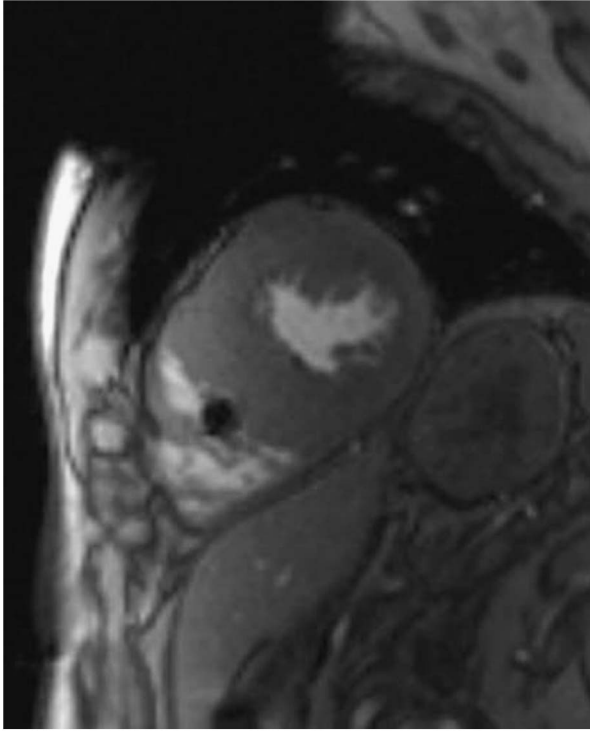
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Introduction: The rare combination of familial WPW and hypertrophic cardiomyopathy (HCM) is caused by dominantly inherited PRKAG2 gene mutations. In contrast to HCM-causing sarcomere disorders, the phenotype is caused by accumulating intracellular glycogen. Multiple accessory pathways and AV conduction problems are common electrical manifestations.

Methods: Cardiac MRI was performed in two families with PRKAG2 gene carriers. Segmental analysis of left and right ventricular hypertrophy, function, and late gadolinium enhancement (LGE) was performed. The electrophysiological findings were assessed.

Results: Of 8 patients, 7 had PRKAG2 mutations, R302Q or R344P. Six patients underwent cardiac MRI. Two symptomatic R302Q patients with palpitations and decreased exercise capacity had significantly elevated left ventricular (LV) mass in a symmetric pattern (picture 1). Midventricular myocardium was most frequently hypertrophied. Four (three R302Q: age 26, 24, and 19 years; one R344P, age 17 years) had normal LV mass but



Picture 1.

asymmetrical hypertrophy of mid-inferolateral or mid-inferior segments. Only the two symptomatic mutation carriers had left ventricular LGE, with 11.2% and 21.9% enhancement of total LV muscle volume.

Three patients had pacemakers, one for bradycardia due to sinus node disease at 15 years of age and two for AV block at 32 and 25 years. Two patients had WPW. They had para-His accessory pathways (APs) with benign antegrade conduction properties and in each of them left sided APs were treated with RF-ablation: a left anterolateral AP in both and a left posteroseptal AP in the other one with multiple APs. The latter had a very fast (290 bpm) SVT utilizing the two left sided APs. In addition, one patient had only retrogradely conducting left-sided AP.

Conclusions: Asymptomatic PRKAG2 patients show eccentric distribution of LVH involving mid-inferolateral parts. In symptomatic patients LVH showed symmetric pattern involving the whole left ventricle myocardium, but the thickest in septum. Cardiac MRI is useful in diagnostics of rare metabolic cardiomyopathies like PRKAG2. These patients need life-long follow-up, not only for HCM, but also for various electrophysiological abnormalities.

MP3-16

2:1 AV block due to extremely prolonged QT interval in newborns: variable genetic background

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Background: Three newborns presented with bradycardia and 2:1 AV block caused by markedly prolonged QT interval. Their parents were healthy with normal ECG's.

Methods and Results: Patient 1 was born with 2:1 AV block due to prolonged QTc-interval of 640 ms. Intermittent bradycardia had been observed since the 20th gestation week. Propranolol was started and 1:1 AV conduction was restored. At the age of 4 months, she had seizures and hysarrythmia on EEG. Her neurological development was slightly delayed. At 3 years she had syncope during exertion and deep bradycardia during sleep. On echocardiography and MRI, non-compaction cardiomyopathy with LVEF of 20% was diagnosed. She underwent left cardiac denervation and epicardial DDD pacemaker implantation, and showed episodes of torsades de pointes (TdP) during recovery. Cardiac transplantation was considered, but the patient developed cardiac ischemia with hemodynamic compromise and did not respond to resuscitation.

Exome sequencing revealed F142I de novo -mutation in CALM1 (calmodulin1) gene disrupting calcium signaling in cardiac cells. The combination of calmodulin defect with non-compaction cardiomyopathy is a new finding.

Patient 2 had 2:1 AV conduction in utero. Postnatally, QTc was 650 ms with T wave alternans and intermittent 2:1 conduction. There was no syndactyly. Echocardiography showed non-compaction cardiomyopathy. An epicardial DDD-pacemaker was implanted, and propranolol therapy 4 mg/kg/day was instituted. At the age of 6 weeks she died suddenly. Non-compaction cardiomyopathy was confirmed in pathologic specimens of the heart. LQT8 (Timothy syndrome) was diagnosed with de novo G406R mutation in CACNAC1 gene.

Patient 3 had intermittent AV block in utero. After birth 2:1 AV conduction and QTc of 530 ms were observed. Repetitive TdP bursts were controlled with esmolol infusion and later with propranolol. VVI-pacing was instituted at the age of 2 weeks and upgraded to DDD-pacing at 4 years. She is doing well. A homozygous LQT2 gene defect L552S (Finnish founder mutation) was diagnosed.

Conclusions: Newborns with 2:1 AV conduction and extremely prolonged QT have poor prognosis. Two of our three patients died; both had also non-compaction cardiomyopathy. One death was probably arrhythmogenic, the other one due to ischemia. Genetic testing may provide exact diagnosis.

MP3-17

Assessment of Systolic Left Ventricular Function with Speckle-Tracking Echocardiography in Adult Patients with Repaired Aortic Coarctation

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Introduction: After successful repair of coarctation of the aorta (CoA), systemic hypertension and development of left ventricular (LV) dysfunction are of major concern at long-term follow-up. Early detection is important to avoid further deterioration. Using speckle-tracking echocardiography (STE), we evaluated LV deformation in adult patients after CoA repair and studied the relationships with conventional diagnostic parameters.

Methods: In this cross-sectional study, we prospectively recruited CoA patients after repair and healthy controls. All subjects underwent echocardiography and electrocardiography. Additionally, NT-proBNP levels of the CoA patients were determined on the same day. With STE, we analyzed LV global longitudinal strain (GLS) at the apical four-, two- and three-chamber view, apical and basal rotation at the parasternal short-axis views, and assessed LV twist.

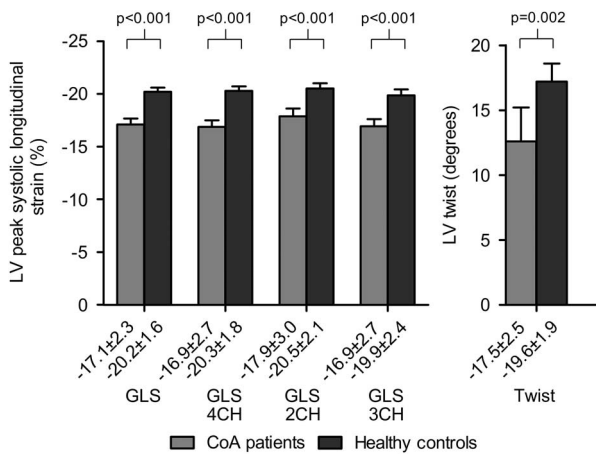


Figure. Left ventricular peak systolic global longitudinal strain and twist. GLS = global longitudinal strain composed of all three apical views; GLS 4CH = GLS measured at four-chamber view; GLS 2CH = GLS measured at two-chamber view; GLS 3CH = GLS measured at three-chamber view.

Results: We included 154 subjects: 77 with repaired CoA (44% female, age 33.6 ± 12.7 years, age at repair 2.5 [IQR:0.1-11.1] years) and 77 healthy controls (44% female, age 34.0 ± 10.6 years). LV GLS of all three apical views and LV twist were significantly lower in patients than controls (Figure). LV GLS of the patients was correlated with systolic and diastolic blood pressure ($r = 0.28$, $P = 0.021$; $r = 0.34$, $P = 0.005$) and QRS duration ($r = 0.35$, $P = 0.004$). NT-proBNP levels were only correlated with LV GLS measured at the apical four-chamber view ($r = 0.27$, $P = 0.023$). Visually assessed systolic LV function was normal in 65 (84%) patients, mildly impaired in 11 (14%), and severely impaired in 1 (1%). When comparing only patients with normal LV function with controls, LV GLS and twist remained significantly lower in the patients ($-17.3 \pm 2.3\%$ vs. $-20.2 \pm 1.6\%$, $P < 0.001$; $12.7 \pm 6.6^\circ$ vs. $17.2 \pm 6.1^\circ$, $P = 0.003$, respectively).

Conclusions: Adults with repaired CoA have reduced LV GLS and twist. Although the majority of patients seem to have a normal systolic LV function when assessed visually, LV GLS and twist are still reduced which could indicate early LV dysfunction.

MP3-18

Cardiac output during exercise in young adults operated for ventricular septal defect in early childhood

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Introduction: Recent studies are challenging the general perception of the long-term outcome after surgical closure of ventricular septal defect (VSD) in childhood. It has been demonstrated that surgical VSD-closure is associated with markedly reduced functional capacity and disruption of the right ventricular force-frequency relationship during exercise in adulthood. In order to describe the late cardiac morbidity, we performed a non-invasive assessment of cardiac output during exercise in adults operated for VSD in early childhood.

Methods: We conducted a prospective study at a tertiary referral center. Patients ($n = 20$) and healthy age- and gender matched controls ($n = 20$) underwent continuous supine bicycle ergometry during real-time phase-contrast magnetic resonance imaging

(MRI) using a Philips 1.5 Tesla Intera scanner. The cycling workload was automatically incremented by 25 watt every 75 seconds during the test session, until reaching submaximal heart rate of 75-80% of maximum (220 bpm - age). The ascending aorta and pulmonary trunk were imaged in a transversal view and real-time flow in each of the vessels was recorded at each exercise level. The cardiac output was determined by blinded, post hoc segmentation using dedicated software. The International Physical Activity Questionnaire and the SF-36 were applied for Health-Related Quality-of-Life assessment.

Results (Preliminary): In the VSD-group the mean age at surgery was 2.1 (SD ± 1.4) years and the age at the time of examination was 22.4 (SD ± 2.2) years in the VSD-group vs. 23.1 (SD ± 2.1) in the control group.

The VSD-group had a lower mean cardiac index (CI) during the test session: 5.19 (SD ± 1.70 L/min/m²) versus 5.65 (SD ± 1.78 L/min/m²) in controls, $p < 0.05$. Cardiac index was related to heart rate during exercise and the VSD-group had lower mean cardiac indices during the entire session although statistically insignificant at predefined heart rates.

Conclusions: Young adults with a surgically closed ventricular septal defect had a slightly reduced cardiac output during supine exercise compared with healthy age- and gender matched controls. Our findings may importantly contribute to the previously demonstrated reduction in peak exercise capacity.

MP3-19

Increasing body mass index is associated with the incidence of supraventricular tachycardia during follow-up of adults with complex congenital heart disease

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Introduction: Obesity and weight gain increase the risk of supraventricular tachycardia (SVT) in the general population. In adult congenital heart disease (ACHD), SVT is an important cause late of morbidity, hospital admissions and may predict mortality. Although SVT is a significant complication for patients with ACHD, the role of obesity as a potentially modifiable arrhythmic risk factor has yet to be explored.

Methods: We retrospectively studied a random sample of complex ACHD patients followed at our institution, all with an initial clinic visit at 17-24 years of age. Patients were categorized by BMI at initial visit as: underweight (BMI < 18.5 kg/m²) normal weight ($18.5 \leq$ BMI < 25 kg/m²) or overweight/obese (BMI ≥ 25 kg/m²). The percentage change in BMI from initial visit to the maximum during follow-up was calculated and classified as: Mild BMI change (-10% to $+9.9\%$), moderate BMI increase ($+10\%$ to $+29.9\%$) and severe BMI increase ($\geq +30\%$). We recorded the occurrence during follow-up of documented SVT on ECG or Holter monitoring and the need for direct current (DC) cardioversion.

Results: There were 292 patients (60% male) including: TOF $n = 156$, Fontan $n = 72$, Mustard $n = 44$, Rastelli $n = 16$ and ccTGA $n = 4$. Median (IQR) duration of follow-up was 9.1 (2.7 - 13.0) years. At initial visit, 12% were underweight 66% were normal weight and 22% were overweight/obese. BMI classification at initial visit did not predict the occurrence of SVT ($p = 0.91$). During follow-up 37% had a moderate and 22% a severe BMI increase and these groups had increased risk of

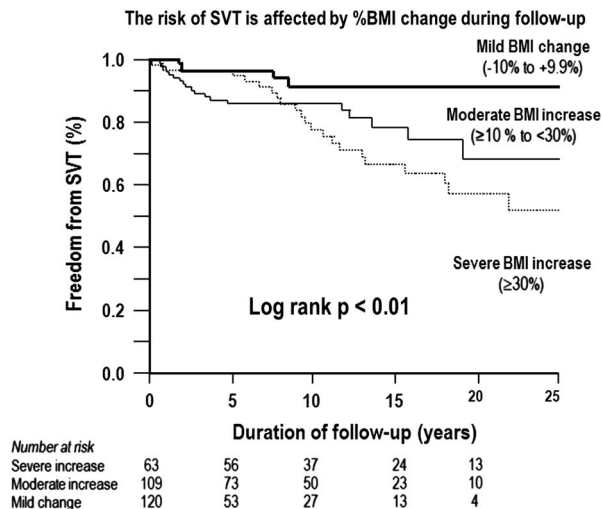


Figure.

developing SVT when compared to the group with mild BMI change (Figure). The highest risk of SVT was in patients with a severe increase in BMI (HR [95% CI]=2.85 [1.14 to 8.64] and 4.35 [1.77 to 13.08] for moderate and severe, respectively, both $p < 0.01$). DC cardioversion was most frequent in the group with severe BMI change (mild change 1.7% vs. moderate increase 16.5% vs. severe increase 32.8%, $p < 0.01$).

Conclusions: Increasing BMI during follow-up is a risk factor for SVT and DC cardioversion in patients with complex ACHD.

MP3-20

Results of cardiopulmonary exercise testing correlate with functional parameters of cardiac MRI in adult Fontan patients - implications for long-term follow-up

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Objectives: To improve long-term outcome of Fontan patients, adequate and continuous follow-up examinations are mandatory. Cardiac MRI (CMR) is a valuable, non-invasive tool to accurately assess cardiac function and quantify Fontan flow. However, CMR is time- and resource-consuming. On the contrary, determination of cardiopulmonary capacity by cardiopulmonary exercise testing (CPET) can easily be performed on a regular basis during follow-up. We therefore analyzed the relationship of CMR measurements with cardiopulmonary capacity in Fontan patients.

Methods: In a retrospective review of our outpatient database covering the last five years, we identified 53 Fontan patients who had both CMR and CPET during regular scheduled visits. At follow-up examination, median age was 18.3 years (range 5.3-48.8) and median follow-up after Fontan operation 11.3 years (range 0.4-22.8). 29 patients were adolescent (>16 years) and adult patients while 24 were children. We compared oxygen uptake capacity (VO_{2max}) determined by CPET with CMR parameters of cardiac function and flow dynamics such as ejection fraction (EF), cardiac index (CI) and venous return (vCI).

Results: In the entire cohort, EF was $53 \pm 13\%$, CI was 3.0 ± 0.7 L/min/m², vCI was 2.8 ± 0.6 L/min/m² and VO_{2max} was $50.6 \pm 14.5\%$ of healthy controls. Except for a higher vCI in children (3.0 ± 0.6 vs. 2.6 ± 0.6 L/min/m, $p = 0.03$) parameters

did not differ significantly between age groups. We found a modest but significant correlation of EF ($r = 0.43$, $p = 0.002$), CI ($r = 0.39$, $p = 0.006$) and vCI ($r = 0.40$, $p = 0.005$) with VO_{2max} . Interestingly, when analyzing age subgroups, a correlation of CI and vCI with VO_{2max} was not confirmed in children ($r = 0.31$, $p = 0.17$ and $r = 0.25$, $p = 0.25$), while clearly present in adolescent and adult patients ($r = 0.45$, $p = 0.02$ and $r = 0.56$, $p = 0.004$, respectively).

Conclusions: Correlation of cardiopulmonary capacity measured by CPET and CMR cardiac function and hemodynamic parameters is modest but significant. In adolescent and adult Fontan patients there is a good correlation of CPET with CI and vCI. Thus, regular CPET should be routinely performed during follow-up, since it allows to estimate deterioration of cardiac function and hemodynamics and might indicate necessity for further investigation or even intervention. In younger Fontan Patients however, CPET seems to be of limited suitability for this purpose.

MP3-21

Pregnancy in women with cardiovascular disease in the Guidelines era: an Italian single centre experience

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Introduction: Risk-tailored multidisciplinary management in specialised centres is recommended for pregnant women with cardiovascular diseases. Migrations and unavailability of specialised clinics make this not always applicable. Our aim was to determine pregnancy outcome with regard to modality of referral to our joint cardiac-obstetric clinic.

Methods: All women referred between 2011 and 2014 were included. Timing and reason for referral were recorded and two groups were identified: women evaluated prior to pregnancy (Group I) and women who were not (Group II). Pregnancies were managed as recommended by ESC guidelines. Maternal death, heart failure, arrhythmias, thromboembolic events and need for urgent surgical or endovascular procedure were defined as cardiovascular events. Gestational age and delivery mode were recorded.

Results: There were 110 pregnancies in 108 women (median age 33 ± 5 years). 51 patients (47.2%) were in Group I, 57 (52.8%) in Group II. Congenital (43.6%) and valvular (26.8%) disease were the most frequent diagnosis. 32% patients were in WHO risk class III or IV. 30% were referred for onset of symptoms, 70% for risk assessment and follow-up planning. Women in Group II were seen later in pregnancy than Group I ($p = 0.009$). 16 (14.5%) cardiovascular events occurred: 8 cases of heart failure, 6 tachyarrhythmias, one patient had mitral surgery due to endocarditis and one had ICD implantation for refractory ventricular arrhythmias. No maternal deaths occurred. Events were more common in Group II ($p = 0.01$), in women in WHO risk classes III-IV ($p < 0.001$) and in patients referred for symptoms ($p < 0.001$). Mean gestational age at delivery was 38 ± 2 weeks, delivery was preterm in 16% of cases, being more common in high risk classes ($p = 0.01$). Cesarean section was performed in 48% of pregnancies, being indication mainly cardiac in high risk patients.

Conclusions: There were no mortality and an overall acceptable incidence of cardiovascular events but a relevant percentage of women were referred late in pregnancy and/or for symptoms. Events were more frequent in this group. Further efforts are needed to optimize referral to specialised centres.

MP4-1

An investigation of scatter removal techniques in paediatric cardiac catheterisation imaging: effects upon radiation dose, image quality and DNA integrity

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Introduction: Paediatric cardiac catheterisations provide immense benefit but also result in a considerable ionising radiation burden to children who are more radiosensitive than adults. The transition from using image intensifiers to flat panel detector technology, as well as wide variations in imaging protocols, indicates that children may be receiving greater radiation doses than necessary. Our ongoing study is investigating the effect of scatter removal techniques upon radiation dose and image quality and DNA integrity.

Methodology: To date, 44 paediatric patients aged 0-15 years have been randomly allocated into one of three imaging protocols prior to cardiac catheterisation: use of an anti-scatter grid (A), removal of anti-scatter grid (B), and removal of anti-scatter grid with a 15 cm air-gap between patient and the image detector (C). For each patient the effective radiation dose and relevant organ doses are being calculated by performing Monte Carlo photon simulation. The number of radiation induced DNA double-strand breaks are being quantified for all examinations using the γ H2AX assay. A visual grading analysis will be performed by blinded clinicians using sample images from each imaging protocol.

Results: Preliminary data from the 44 participants has demonstrated that the mean effective dose of protocol B is lower (5.1 mSv) compared to protocols A (7.5 mSv) and C (5.2 mSv). Increases in mean γ H2AX-foci are greater for protocol A (0.14) compared to protocols B (0.07) and C (0.08).

Conclusion: Preliminary data has demonstrated that the removal of the anti-scatter grid in paediatric cardiac catheterisations may result in reductions in radiation dose (~32%) and radiation induced DNA damage (~50%). Protocols A, B and C have each provided images which were sufficient for successful completion of cardiac catheterisation. Our study will be fully completed for presentation at AEPC 2015 and will also include blood and organ radiation doses and estimated cancer risk calibrated from DNA damage observations.

MP4-2

Real-Time 3D-Echocardiography of the Pediatric Right Ventricle - influence of different quantification software

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Background: Determining right ventricular (RV) volume and function is essential for therapy and follow-up in patients with heart disease. Real-time 3D echocardiography (RT3DE) is a promising method for RV assessment, however there is no systematic study comparing the inherent influence of different evaluation software on the resulting measurements.

Methods: 379 healthy children and adolescents (range, 1 day-216 months) underwent RT3DE imaging of the RV, 6

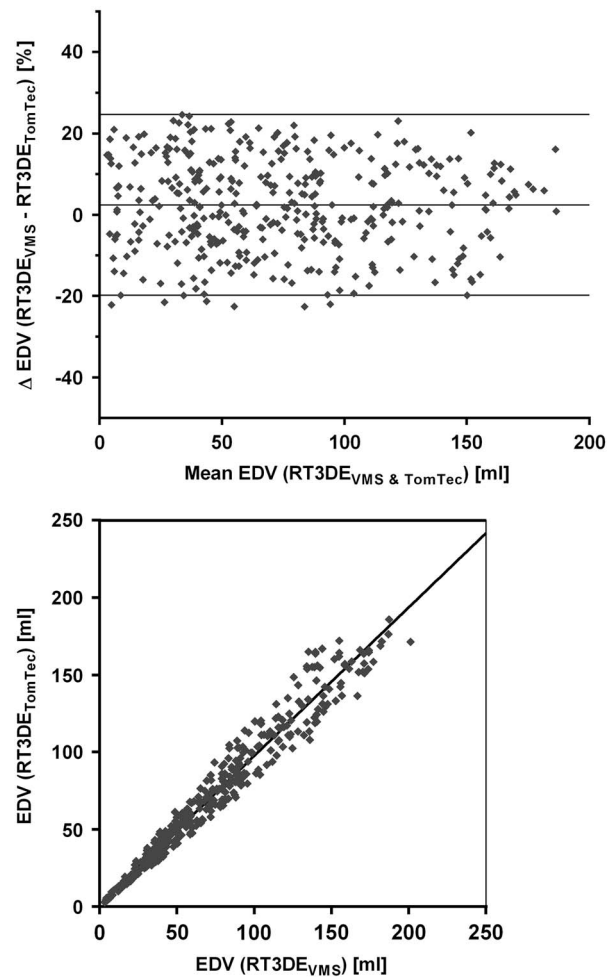


Figure.

Table.

RT3D Volume Analysis of the Right Ventricle in Children Agreement between VMS and TomTec

Para-meter	Bias	Bias (%)	Standard Deviation (ml)	SD (%)
ESV	-0.18	-0.79	5.24	13.8
EDV	1.64	2.52	9.34	11.44
SV	1.82	4.73	6.93	15.42
EF		1.33		4.58

subsequently underwent CMR imaging. RT3DE data sets were quantified using both a knowledge-based reconstruction software (VMS, Ventripoint Diagnostics Ltd., Bellevue, Washington, US) and a semiautomatic border detection software (TomTec RV V1.1, Unterschleissheim, Germany). CMR data sets were quantified by the method of discs (MOD).

Results: Bland-Altman Analysis showed estimations of end-diastolic volume (EDV, Figure) using VMS to be larger than using TomTec, while estimations of end-systolic volume (ESV) were slightly smaller, resulting in larger stroke volume (SV) and ejection fraction (EF) in VMS (Table). Compared with CMR there were trivial volume overestimations using VMS (EDV: Bias: 1.73 ml = 1.69%, SD: 1.82 ml = 1.69%), (ESV: Bias: 1.12

ml = 2.54%, SD: 2.53 ml = 4.85%), (SV: Bias: 0.62 ml = 0.92%, SD: 2.96 ml = 3.59%) and underestimation of EF (Bias: -0.40%, SD: 1.84%) while volumes and EF were underestimated using TomTec (EDV: Bias: -9.0 ml = -8.98%, SD: 13.41 ml = 13.73%), (ESV: Bias: -1.03 ml = -1.44%, SD: 3.55 ml = 6.88%), (SV: Bias: -7.99 ml = -14.72%, SD: 11.87 ml = 21.48%), (EF: Bias: -3.45%, SD: 5.07%). Intra- and interobserver-variation for EDV, ESV and SV were excellent both in VMS and TomTec with intraclass correlation coefficients (ICC) between 0.992-0.998. Regarding EF, ICC for intraobserver variability was significantly lower using TomTec (ICC 0.792; CI [0.829-0.843]) if compared to VMS (ICC 0.973, CI [0.941-0.987]).

Conclusions: 3D-volumetric assessment of the RV is possible using different evaluation software. However, resulting measurements differ depending on the software used. Therefore a different set of reference values is required.

MP4-3

3 years' experience with pre-operative Dual Source CT (DSCT) for initial congenital cardiovascular surgery: Do reduced radiation dose and the option of 3D planned surgery justify an extended use?

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Objectives: Evaluation of feasibility, impact and effective dose (ED) of DSCT with 3D-reconstructions for diagnosis and for planning initial and repeat cardiovascular surgery in first year of live.

Methods: During a three years period 2012-2014, in a total of 123 consecutive patients with CHD less than one year of age imaging was performed to assess the arbitrary anatomic malformation. DSCT scanner second generation SOMATOM Definition Flash (80 kV) and third generation SOMATOM Force at 70 kV [Siemens] were used. 3D-reconstruction with TeraRecon™-software. Image quality; artifacts; utility of iterative reconstructions

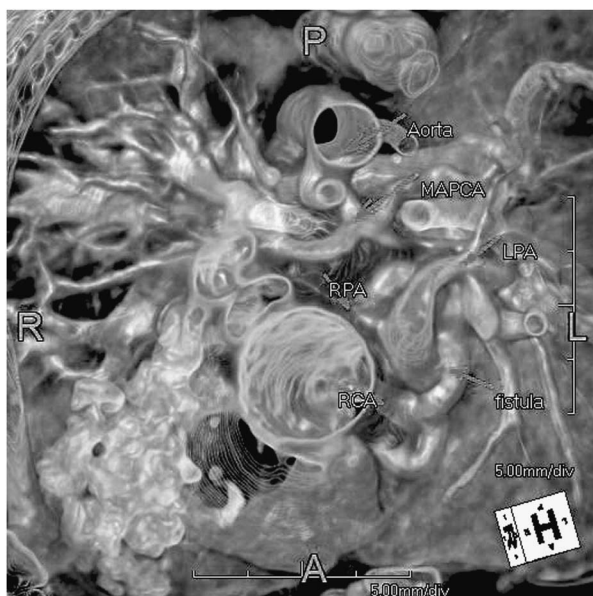


Figure.

Table.

	Number	Age (d)	ED (mSv)	Contrast to noise	Visibility of mammarian artery (%)	Utility for surgery
Neonates Force	14	6 (1-22)	0.24 (0.19-0.31)	18 (11-21)	100	1.8
Neonates Flash	43	3 (0-13)	0.4 (0.3-0.7)	29 (5-78)	90.1	2
Infants Force	15	104 (30-320)	0.21 (0.13-0.49)	14 (8-29)	79	1.9
Infants Flash	51	103 (29-348)	0.4 (0.23-1.2)	24 (6-78)	88	1.8

and radiation dose was assessed. The utility of the 3D-reconstructions was rated using a 5-Point-scale from “essential” (1 P) to “misleading” (5 P).

Results: In all cases imaging was successful without complications. 3D-imaging for pre-operative planning; was rated as very useful or even essential in all cases.

Conclusions: Low dose DSCT is an ultrafast and appropriate imaging modality in the preoperative assessment of complex CHD. 3D-imaging enables an advanced surgical planning without the need of an experienced imaginativeness. Actual ED lower than 0.5 mSv justifies an extended use of this imaging modality in complex cases and in critical ill patients.

MP4-4

Hybrid palliation allows the hypoplastic left ventricle for a chance of biventricular repair: echo-surveillance on left heart growth

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Objectives: We aimed to describe the growth potential of the left ventricle (LV) in patients with borderline hypoplastic left heart, excluding aortic atresia, who underwent hybrid palliation with pulmonary arteries banding and ductal stenting.

Methods: 43 patients (25 males; mean weight 3 kg), born with hypoplastic LV underwent hybrid palliation at 3,6 days between October 2011 and November 2014. We focused our attention on 16 patients with intermediate features. We excluded mitral atresia or stenosis and aortic atresia, in which left heart structures are not expected to grow, and interrupted aortic arch with ventricular septal defect since LV is usually suitable for biventricular repair. The intermediate group included patients with mitral stenosis/aortic stenosis and right unbalanced atrioventricular septal defect, with severely underdeveloped LV (LV mass z-score <2). After interstage (mean of 4 months) 5 patients (group 1) underwent single ventricle staged palliation, 8 patients (group 2) had biventricular repair and 3 patients (group 3) died. Two-dimensional echocardiograms were retrospectively reviewed, comparing examinations at birth and after interstage. Aortic annulus and root z-scores, indexed aortic root diameter, LV long-axis to heart long-axis ratio (LAR), mitral valve (MV) annulus z-score, indexed and z-score LV mass, LV end-diastolic and end-systolic indexed volumes are reported in Table 1. Group 1 was compared to Group 2 at birth and after interstage, using a Mann-Whitney Test.

Results: MV annulus, LV mass and volumes were critically small in both groups at birth. After interstage a significant growth of LV mass and volumes from pre- to post-hybrid procedure was evident only in group 2. Conversely, parameters like aortic annulus, aortic root and LAR were smaller in group 1 than in group 2 at birth and

Table 1.

Parameter	Group1 at birth	Group2 at birth	P value	Group 1 interstage	Group2 Interstage	p value
Aortic annulus Z	-5.47	-3.49	0.071	-3.52	-2.17	0.169
Aortic root Z	-5.13	-3.31	0.053	-3.28	-1.65	0.06
Ind aortic Root (mm/m ²)	2.76	3.33	0.071	2.61	3.31	0.041
LAR	0.67	0.79	0.054	0.73	0.85	0.032
MV annulus Z	-4.21	-4.13	0.880	-3.81	-2.57	0.132
LV mass Z	-5.58	-4.89	0.271	-5.10	-2.48	0.032
Ind LV mass (g/m ²)	20.20	24.00	0.260	29.00	45.38	0.068
Ind End Diastolic Volume (ml/m ²)	7.44	10.08	0.168	11.94	25.69	0.034
Ind End Systolic Volume (ml/m ²)	4.00	3.96	0.970	5.20	11.78	0.023

could be useful in predicting which patient will probably end up in biventricular group.

Conclusions: The goal in the treatment of borderline hypoplastic LV is to postpone the decision for surgical repair until the time the LV has fully expressed its growth potential: then, many more biventricular repairs will be reached than at the time of first evaluation. Echocardiography provides good quantitative analysis for a tight surveillance on LV growth.

MP4-5

Can the subsystemic right ventricle remodel and contract like a left ventricle after atrial switch repair for transposition of the great arteries? A magnetic resonance feature tracking study

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Introduction: D-transposition of the great arteries can be repaired with re-direction of blood flow either by atrial (Senning) or by arterial (ASO) switch operation. The Senning procedure results in a sub-systemic morphologically right ventricle (RV) and a sub-pulmonary morphologically left ventricle (LV), while ASO creates concordant atrio-ventricular and ventriculo-arterial connections. We sought to determine to what extent the RV after Senning can compensate as a sub-systemic pumping chamber.

Methods: Twenty-three patients (14 Senning, 9 ASO) without significant residual lesions underwent cardiac magnetic resonance imaging. Mean age was 27.6 ± 5.5 y for the Senning and 17.6 ± 7.1 y for the ASO patients ($p=0.002$). 2D SSFP cine images were post-processed with a feature tracking software (TomTec 2D CPA). Global strain was measured separately for each ventricle in radial and circumferential directions in short axis mid-ventricular slices, and in longitudinal direction in a four-chamber view.

Results: The sub-systemic RV in Senning patients showed reduced strains compared to sub-systemic LV in ASO patients in both radial (21.7% vs. 32.8%; $p=0.001$) and circumferential (-16.1% vs. -25.5%; $p<0.001$) directions. In contrast, longitudinal strain values were similar for either ventricular morphology. In sub-pulmonary position, RV presented with lower circumferential strains than LV (-16.7% vs. -19%; $p<0.05$). No significant difference was observed for global contractile function between morphologically RV and LV in sub-systemic position (EF% 52 ± 9 vs. 58 ± 7), nor in sub-pulmonary position (EF% 60 ± 8 vs. 54 ± 8).

Conclusions: In spite of preserved global contractile function, the sub-systemic RV can adapt and behave like a LV only in longitudinal direction, but not in radial and circumferential shortening. This may reflect the well described specific fibre arrangement of each ventricle. Correspondingly, higher circumferential deformation was found in the morphological LV than RV in sub-pulmonary position.

MP4-6

Preoperative evaluation of total anomalous pulmonary venous connection in children with low-dose dual-source MDCT angiography

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Objective: To determine the diagnostic accuracy of low-dose dual source 256-detector multidetector computed tomographic (MDCT) angiography in children with total anomalous pulmonary venous connection (TAPVC).

Material and Methods: MDCT angiography images of 43 patients [23 female (53%), mean age: 9 months (4 days-7.1 years)] with TAPVC confirmed by surgery were retrospectively reviewed. In all patients, MDCT examinations were performed using a dual-source 256-MDCT scanner (Definition Flash, Siemens Healthcare) with high pitch protocol. The types of TAPVC, the presence of obstruction and associated cardiovascular anomalies were investigated. According to accompanied cardiovascular anomalies, patients were subdivided into two groups as isolated and complex type. Isolated type was diagnosed if the patient had associated atrial septal defect (ASD) and/or patent ductus arteriosus (PDA) and complex TAPVC was diagnosed if the patient had other cardiovascular anomalies besides ASD and/or PDA.

Results: In 43 patients, 22 of them (51%) were supracardiac, 10 (23%) were cardiac, 6 (14%) were infracardiac, and 5 (12%) were mixed types. Obstruction was detected in 7 (16%) patients. 17 patients (40%) were isolated and 26 patients (60%) were complex type. The patients with complex TAPVC had one or more of accompanied anomalies including atrioventricular septal defect ($n=7$), ventricular septal defect ($n=7$), persistent left superior vena cava ($n=7$), pulmonary stenosis ($n=7$), right atrial isomerism ($n=5$), double outlet right ventricle ($n=5$), right aortic arch ($n=5$), transposition of the great arteries ($n=4$), tubular hypoplasia of the aortic arch ($n=3$), aortic coarctation ($n=3$), pulmonary artery hypoplasia ($n=3$), right pulmonary artery agenesis ($n=1$), cor-triatrum sinister ($n=1$), left ventricle agenesis ($n=1$), pulmonary atresia ($n=1$), hepatic interruption and anomalously drainage of inferior vena cava into left atrium ($n=1$), and an accessory left hepatic vein anomalously draining into left atrium ($n=1$). The diagnostic agreements between MDCT and surgical results were %100 in isolated and complex group. The overall mean effective radiation dose was 0.66 mSv (range, 0.15-1.11 mSv), and it was 0.52 mSv (range, 0.12-0.72) in the patients younger than 1 year old.

Conclusion: Dual-source 256-detector MDCT angiography is a reliable imaging modality allowing comprehensive anatomic imaging of TAPVC in neonates and children with lower radiation doses.

MP4-7**Improving the Role of Echocardiography in the Quantification of Right Ventricular Geometric and Functional Parameters of Corrected Tetralogy of Fallot Patients Awaiting Pulmonary Valve Replacement: Comparison with MRI Findings**

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Introduction: Right ventricular (RV) dilatation is the most frequent complications in corrected tetralogy of Fallot patients (T4F). Correct timing for pulmonary valve replacement (PVR) represents a crucial issue in the correct follow-up of these patients. However, indications for pre-PVR MRI examination in T4F patients in clinical practice is based of qualitative or only semi-quantitative echocardiographic assessment of the RV. The aim of our study was to report the diagnostic accuracy of echocardiography in the definition of RV geometry and function and to develop a model to improve the correct identification of T4F patients undergoing pre-PVR MRI study.

Methods: We studied 174 corrected T4F patients, who performed an echocardiographic examination and an MRI study in order to establish indications for PVR. Indications for PVR were defined as RV end diastolic volume index $\geq 155 \text{ ml/m}^2$ and/or RV EF $< 47\%$. **Results:** 95 patients had echocardiographic exams with adequate quality to derive parameters for the present analysis study. Of the 95 patients ultimately included in the analysis, 41% met MRI criteria for PVR. When analyzing functional parameters we found a strong association between MRI-EF and ECHO-RVFAC ($r=0.42$; $p<0.001$), with no association identified with other routine echocardiographic parameters of RV function. When analyzing geometric parameters a significant association between all echocardiographic parameters (RV area, RV length and diameters, proximal and distal RVOT diameters) and MRI-RV end diastolic volume could be observed (all $p<0.05$), with the strongest association found for RV diastolic area ($r=0.66$; $p<0.001$), and proximal RVOT diameter ($r=0.55$; $p<0.001$). Thus, we defined a regression equation for predicting MRI volume using echocardiography (RV EDV [MRI]=[Coeff.A *RVED area] + [Coeff.B *RVOTprox] - K; $r^2=0.59$; $p<0.001$; model tolerance >0.75). Applying regression equation we were able to correctly identify 64% patients according to PVR criteria, significantly improving the diagnostic ability of echocardiography in these patients.

Conclusions: Echocardiographic assessment of the RV in T4F is limited by exam quality. However, when available, combining parameters assessing the RV inflow and outflow in a simple regression equation, allows an accurate estimate of MRI-RV volume, thus improving the echocardiographic ability to correctly identify T4F patients candidate for pre-PVR MRI study.

MP4-8**Impact of loading conditions on ventricular function in Ebstein anomaly (EA) of tricuspid valve**

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Introduction: In EA, both atrialised and functional components of the right ventricle are exposed to high preload and low afterload.

The latest surgical repair (Cone operation) enables to eliminate tricuspid regurgitation (TR) and reposition tricuspid valve to its anatomic annulus. The aim of the study was to investigate the adaptation of right (RV) and left ventricles (LV) to change in loading conditions after Cone repair.

Methods: A retrospective longitudinal study was conducted from 2009 to 2014. All symptomatic patients with moderate to severe TR were included. Transthoracic advanced echocardiography was performed pre-operatively, at short-term (less than 30 days after Cone repair) and mid-term (2 months to 4 years). Conventional parameters, the systolic peak of longitudinal 2D strain and the time to systolic peak (onset of QRS to systolic peak of longitudinal strain) were measured for LV and RV. Paired t-test analyses were performed using Wilcoxon Matched-pairs signed rank test.

Results: From the 38 patients operated for EA, the echocardiographic data of 17 patients could be analysed. GOSH score was significantly reduced after Cone repair (1.07 ± 0.24 vs. 0.30 ± 0.07 , $p=0.020$) as well as TR (3.53 ± 0.24 vs. 1.18 ± 0.37 , $p=0.003$). However, the TAPSE (26.42 ± 5.79 vs. 5.78 ± 2.00 , $p=0.005$), LV and RV systolic peaks were significantly reduced post-operatively (-19.80 ± 1.05 vs. -15.85 ± 1.13 , $p<0.001$ for LV, -18.50 ± 1.80 vs. -13.53 ± 1.52 , $p<0.001$ for RV), but with no significant reduction between pre-operative and mid-term post-operative period (-19.80 ± 1.05 vs. -21.86 ± 1.90 , $p=0.677$ for LV, -18.50 ± 1.80 vs. $-18.74.50 \pm 1.80$ vs. 3.66 , $p=0.285$ for RV). LV time to peak was significantly reduced in short-term post-operative period (441 ± 9 vs. 415 ± 11 , $p<0.001$) while RV free wall time to peak was significantly prolonged in mid-term post-operative period (445 ± 8 vs. 469 ± 13 , $p=0.001$).

Conclusions: The rapid change from high preload/low afterload to low preload/relatively high afterload following Cone operation reduces myocardial contractility of both ventricles but with the trend to later recovery; LV myocardial mechanics appears better than preoperatively while continuing impairment of RV function may reflect intrinsic myocardial dysfunction likely due to myocardial deficiency.

MP4-9**Ambulatory monitoring of blood pressure and arterial stiffness in patients with aortic coarctation**

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Introduction: After successful correction of aortic coarctation (AoCo), some patients continue to have arterial hypertension. Ambulatory monitoring of blood pressure (AMBp) has been suggested as the ideal method for evaluating these patients. Arterial stiffness may be altered in patients with efficiently corrected coarctation, reflecting the inherent arteriopathy.

Methods and Results: We studied the relation between AMBP and arterial stiffness in patients with aortic coarctation. Arterial stiffness was studied by tonometry (Sphygmocor®), through the augmentation index (Alx) and pulse wave velocity (PWV). To avoid the influence of the AoCo region, arterial stiffness parameters were obtained in the right radial artery. 33 patients (8 women, 18.3 ± 11.2 years of age), 17 after corrective surgery, 10 after balloon dilatation, 4 after stenting and 2 with native AoCo were studied. Antihypertensive medication was found in 8 patients (24%). In the AMBP, 28 patients (85%) were Non Dipper, 19 patients (58%) had systolic 24 h loads above 25% and 9 patients (27%) had diastolic 24 h loads above 25%. Mean Alx corrected to 75 bpm was 9.5% and mean PWV was 7.8 m/s. Several regression models were constructed with AMBP parameters as dependent variables and all arterial stiffness parameters as

independent variables, using R-3.1.0 software. With diastolic 24 h loads (D24Load) as the dependent variable, the best linear regression model was obtained with PWV as the only independent variable, with the formula $D24Load = -16.18 + 4.19 \cdot PWV$. There was a significant relation between D24Load and PWV (t test: $p < 0.01$).

Conclusion: Through regression models, it was possible to establish a relation between diastolic 24 h loads by AMBP and arterial stiffness parameters in patients with AoCo, native and corrected. Although systolic blood pressures are classically the main focus of blood pressure evaluation in coarctation patients, diastolic pressures and loads should be equally addressed in the evaluation of these patients, as they correlated better with changes in arterial stiffness.

MP4-10

Borderline Left Ventricles: morphological differences in papillary muscles compared to normal

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Introduction: Papillary muscles (PM's) are usually organized as two equal-sized muscles holding superolateral (SL) and inferomedial (IM) positions. However, as described in autopsy studies, their morphology may vary. In this study, we describe differences in PM morphology in borderline and normal-sized left ventricles (LV's) using MRI.

Methods: This retrospective study comprised thirty-one consecutive children with borderline LV (heterogenous group with severe aortic stenosis, coarctation with borderline LV and aortic atresia with VSD) and thirty consecutive subjects with a normal-sized LV. PM morphology, location, angle created between them and their length ratio related to LV length was assessed using steady-state-free-precession short-axis (SAX) cine and 3D-whole-heart sequences.

Results: The median age was 4.3 years (range: 4 months–20 years; 36 male). The combination of SAX and 3D-whole-heart imaging demonstrated PM anatomy in all cases.

All patients with normal-sized LV had two PM's whereas eight (24%) borderline LV cases had a single IM muscle, and one had a single SL muscle.

Splitting of PM into groups was found with equal frequency in normal and borderline LV's when considering the SL muscle (40 vs. 24%; $p = 0.21$) but more frequently in the normal LV when considering the IM muscle (64 vs. 36%; $p = 0.03$). Although PM splitting is common in the normal LV, the PM pedicle was narrow or fused in all normal cases. However, 16% of borderline LV's showed at least one PM, which was split and had a broad insertion. SL muscles were shorter in the borderline group (SL/LV ratio 0.31 ± 0.17 vs 0.46 ± 0.09 ; $p < 0.001$) but IM muscles were of similar lengths between groups (IM/LV ratio 0.39 ± 0.15 vs 0.43 ± 0.09 ; $p = 0.2$). The mean angle between PM insertion points was also similar between groups ($118 \pm 42^\circ$ vs $114 \pm 17^\circ$; $p = 0.63$).

Eight children underwent successful biventricular repair in this series of borderline LV. There were no significant differentiating features for PM morphology in this subgroup.

Conclusions: In normal-sized LV, PM's are dual with narrow/fused pedicles. In comparison, the AL muscle is shortened or often even absent in the borderline LV. Additionally, the PM pedicle in borderline LV's can be split with a broad-based insertion. A

combination of 3d whole-heart and short-axis cine is helpful to delineate papillary morphology.

MP4-11

New Diagnostic Methods for Left Ventricular Function of the Patients with Beta-Thalassemia Major

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Introduction: The aim of this study is to determine the early regional and global myocardial functional changes and whether the myocardial changes that cannot be detected by conventional echocardiography could be detected by T2* magnetic resonance imaging (MRI), tissue Doppler imaging (TDI) or speckle-tracking echocardiography (STE) in beta-thalassemia major (BTM) cases without symptoms.

Methods: The myocardial functions were evaluated by conventional echocardiography, TDI and STE in 30 children with BTM and 30 healthy children. Cardiac T2* MRI has been used to measure the amount of cardiac iron deposition.

Results: The mean age of the patients was 13.3 months at the time of the diagnosis. The mean duration of taking erythrocyte transfusion was 13.5 years.

When left ventricular basal lateral wall and basal septum TDI values were compared between the patients with BTM and the control group; it was observed that only isovolumic contraction time (ICT) values were statistically significantly longer in the patients with BTM.

Longitudinal and circumferential strain values were statistically significantly lower in the patients with BTM than the control group globally. In addition, when evaluated as segmental; longitudinal strain of basal interventricular septum, and circumferential strain values of anteroseptal, and anterior segments, were significantly lower in the patients with BTM than the control group. In the patients with BTM, the group who have pathological T2* MRI values (< 20 ms, $n = 10$), longitudinal and circumferential strain values globally were significantly lower than the group who have not. In addition, circumferential strain values in anteroseptal, anterolateral, inferior, and inferoseptal segments were significantly lower in the patients with T2* values < 20 ms than those with T2* values ≥ 20 ms.

Conclusions: Cardiovascular T2* MRI is the most sensitive test in detecting myocardial iron load. However, it is expensive and is not widely available in many centers. Echocardiography is available in many centers and easy to perform. Our results indicate that STE and TDI can be used for screening of patients for myocardial dysfunction. Findings of this study and future longitudinal studies might have an important impact on monitoring the effects of the chelation therapy.

MP4-12

Maternal ingestion of cocoa constricts fetal ductus arteriosus: an experimental study

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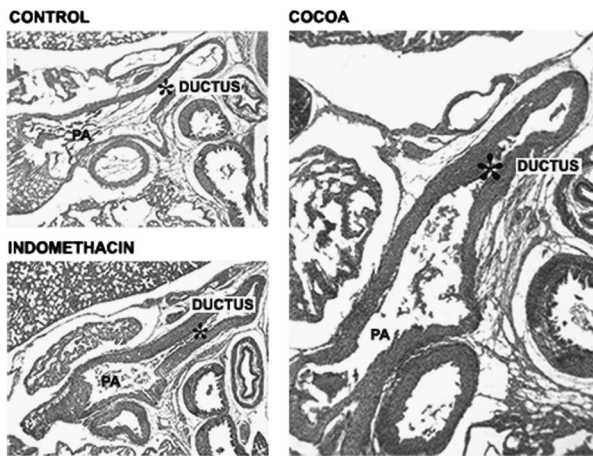


Figure.

Objective: Cocoa has antioxidant and antiinflammatory effects by downregulating COX2 receptors expression in the endothelium and enhancing nitric oxide bioavailability. In this study, we investigated whether cocoa causes ductus arteriosus constriction (DAC) in fetal Wistar rats and if it has antioxidant effects on fetal tissues.

Methods: Wistar pregnant rats at the 21st day of gestation were submitted to administration of cocoa (7.2, 72 or 720 mg/kg) 12 h before cesarean section, indomethacin (10 mg/kg) or water 8 h before cesarean section. Thorax and livers were stored. By digital microscopy, images were digitalized and the diameters were measured. Fetal and maternal liver catalase and superoxide dismutase activities and TBARS levels were analyzed. Statistical analysis used one way ANOVA and differences were compared by Tukey's tests.

Results: Maternal ingestion of cocoa 720 mg/kg and indomethacin 10 mg/kg decreased fetal internal DA diameter when compared to water, (mean: $135 \pm 53 \mu\text{m} \times 92 \pm 59 \mu\text{m} \times 263 \pm 81 \mu\text{m}$, $P < 0.0001$, $n = 7-33$) respectively. Cocoa alone increased DA wall thickness, when compared to indomethacin and control ($200 \pm 56 \mu\text{m} \times 160 \pm 36 \mu\text{m} \times 136 \pm 34 \mu\text{m}$, $P < 0.0001$, $n = 7-33$) respectively. Similar to indomethacin, cocoa decreased the ratio between ductal internal diameter and pulmonary artery internal diameter compared to control group ($0.4 \pm 0.1 \times 0.2 \pm 0.09 \times 0.72 \pm 0.3$, $n = 7-33$), respectively. Sagittal cross sections of the fetal heart showed DAC after administration of both cocoa and indomethacin compared with the control group. Cocoa did not alter oxidative stress markers in fetal nor maternal livers, a similar effect to indomethacin.

Conclusion: These results constitute pharmacological evidence supporting the role of cocoa as a potential cause of fetal ductal constriction, which prompts a note of caution for maternal consumption of chocolate in late pregnancy.

MP4-13

Impact of antenatal echocardiography on outcome of neonates with coarctation of the aorta

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The objective of this study was to analyse the impact of antenatal echocardiography on outcome of neonates with coarctation of aorta (COA).

Methods: Single-center analysis of clinical and echocardiographic data of foetus with suspicion of, and neonates and infants <1 year of age with COA. Groups DAN (antenatal diagnosis) and DPN (postnatal diagnosis) were compared.

Results: 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. PGE was administered in 53% preoperatively and mechanical ventilation in 39%. Among 275 confirmed COA (95 DAN and 180 DPN), 272 underwent Crafoord operation, at mean age 29days (median 12days) and mean time after admission 3.4days. Age at surgery was lower and time to surgery longer in DAN than DPN group. Survival was 98.5%. Restenosis occurred in 20cases (7%). Postoperative morbidity, survival and restenosis were not different between the two groups.

Conclusion: Antenatal diagnosis helps to detect severe postnatal COAO but sensitivity is still low. Antenatal diagnosis impacts on early outcomes of neonates with COA but not on late outcomes and restenosis.

MP4-14

Complex Rhythm Patterns Associated with Fetal Atrial Flutter

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Background: Atrial flutter (AFL) accounts up to a third of all fetal tachyarrhythmias and is believed to present later than 30 weeks gestation. Hydrops fetalis and a high mortality rate have been described. The aim of this study was to investigate the electrophysiologic characteristics of fetal AFL by fetal magnetocardiography (fMCG) from 20 weeks gestation onwards in order to define patterns that might predict sustainability.

Methods: All fetuses were referred due to a suspected diagnosis of AFL by echocardiography, which included an atrial rate $> 300/\text{min}$ and variable AV conduction. Electrophysiologic fMCG rhythm and conduction patterns such as RR interval, cardiac time intervals, AV conduction pattern were defined for presenting and associated rhythms. The fMCG recordings were made using a 37-channel superconducting quantum interference device magnetometer.

Results: 15 fetuses were included in the study with a median gestational age of 30 (Range 20 to 38) weeks gestation. AFL was diagnosed in 13 fetuses (incessant AFL ($n = 3$); intermittent AFL ($n = 10$); Sinus rhythm was found exclusively in two fetuses. Additional rhythm patterns showed SVT ($n = 6$), WPW pattern ($n = 3$) and both ($n = 1$), blocked atrial bigeminy/trigeminy ($n = 7$), atrial fib/flutter ($n = 2$), atrial couplets ($n = 1$), bundle branch block ($n = 1$) and saw tooth P wave pattern ($n = 3$).

Conclusion: Fetal AFL includes variable and complex rhythm patterns and can present earlier than 30 weeks gestation, indeed as

early as 20 weeks gestation. As these rhythm disturbances can lead to severe complications in pregnancy, fetal electrophysiologic assessment may help better define antiarrhythmic therapy and perinatal management strategies.

MP4-15

Fetal atrioseptostomy and stenting – what can be achieved?

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Objective: HLHS with closed or restrictive atrial septum is fatal condition with mortality rate close to 100%. Fetal intervention has been proposed as the rescue procedure. The aim of this study was to evaluate the indication for fetal atrial septum opening and review the outcome.

Method: Since 2011 one team performed 12 fetal interventions in 11 fetuses for opening the interatrial septum. The indication for the intervention was closed foramen ovale in fetuses with HLHS or severe aortic stenosis, with biphasic flow in the pulmonary veins. We evaluated fetal condition after intervention and neonatal outcome.

Results: 6 fetuses had HLHS with closed Fo, 5 closed Fo with critical aortic stenosis and heart failure (AS&HF). 4 of them had aortic valvuloplasty previously (BAV). The mean age of diagnosis was 23 In HLHS and 26 in AS&HF. Atrial balloon septostomy was performed in 3 fetuses. Procedures were not successful, in all atrial septum closed soon after ballooning. One fetus died during 24 hours post intervention, second was life-born with closed IAS, in the third stent was placed. In two fetuses with HLHS the connection between pulmonary vein and SVC was suspected, in one was confirmed after birth, in one disappeared after atrial stenting. In all fetuses after stent placement was immediate improvement of the pulmonary veins flow. There were 2 IUD within 24 hours from interventions. 3 neonates were born prematurely, all died in the early neonatal period despite patent stent (2 HLHS, 1 AS&HF). 6 children were delivered at term, all in good condition. They were operated on within 24 hours – surgical atrioseptectomy was done, in 2 with pulmonary artery banding, in 1 – Norwood procedure. There is just one survivor – the baby with AS&HF.

Conclusions: Interatrial stenting is technically feasible and successful procedure. Fetuses with severely impaired LV function and closed Fo probably had lethal illness, in which prenatal interventions cannot change the natural history. Careful evaluation of neonatal treatment must be established to understand the poor outcome in spite of successful prenatal therapy.

MP4-16

Fetal Cardiomegaly: Looking beyond the Fetal Heart.

A Single-Centre Experience

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Objectives: Fetal cardiomegaly when not associated with structural or rhythm abnormalities, can be a diagnostic challenge for cardiologists. The aim of this study was to review our series of fetal cardiomegaly, focusing on aetiology, with emphasis on cases with no obvious cardiac causes.

Table.

Aetiologies	Cases initially referred to cardiologist Number (%)	Mean gestational age (weeks) at referral
Group 1:		
Fetal Vascular Tumor	0/3	–
Kidney Malformations	0/5	–
Lung Malformations	0/3	–
Anhydramnios	0/15	–
Abdominal Abnormalities	0/6	–
Group 2:		
Vein Abnormalities	4/14 (29%)	26.6
Placental Tumors	3/14 (21%)	24.6
Congenital Infections	6/25 (24%)	21.3
Fetal Anaemia (no infection)	0/19	–
Arteriovenous Malformations	2/5 (40%)	28.5
Skeletal Disorders	3/17 (18%)	28.5
Group 3: Miscellanea		
Group 3: Miscellanea	2/13 (15%)	22.4
Group 4: Unexplained		
Group 4: Unexplained	8/15 (53%)	27.7
Total	28/154 (18%)	–

Methods: This was a retrospective study conducted in a single tertiary referral center (Fetal Medicine and Fetal Cardiology Unit) over a 17-year period. From a computerized fetal database, we searched for all cases with cardiomegaly or where cardiac size was measured. All available images and digital records were reviewed. The diagnosis of cardiomegaly was accepted when the cardiothoracic circumference ratio (CTR = heart circumference/chest circumference ratio at the 4-chamber level) was above the 95th percentile for gestational age. Twins and growth-restricted fetuses were excluded. Cases were grouped according to aetiology and percentage initially referred to the fetal cardiologist was noted.

Results: There were 510 fetuses with cardiomegaly of which 240 (47%) had a cardiac abnormality. In 270 cases (53%), no cardiac reason was found. There were 54 twins and 62 growth restricted fetuses. In the remaining study group of 154 singleton pregnancies, we identified four aetiological groups (Table): Group 1 consisted of cases with an overt aetiology, which was associated with obvious extra-cardiac abnormalities usually detected during routine obstetric scans (n = 32, 21%); Group 2 included fetuses with an apparent isolated cardiomegaly where the covert aetiology was not obvious on routine screening (n = 94, 61%); Group 3 consisted of a miscellaneous group of mixed causes (n = 13, 8%) and Group 4 was truly unexplained cardiomegaly (n = 15, 10%). The percentage of all cases initially referred to the cardiologist was 18%. Most of these belonged to Group 2, none to Group 1.

Conclusions: Fetuses presenting with apparent unexplained cardiomegaly are often referred to cardiologists for initial diagnostic workup. It is important to raise awareness of possible non-cardiac causes of cardiomegaly (search for infection, assessment of the fetal and placental circulation). A multidisciplinary approach is likely to optimize pregnancy outcome.

MP4-17

Fetal Cardiac Imaging Audit – A Revised Scoring System to Assess Quality of Images at Routine Screening

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Introduction: Recent guidelines for screening for congenital heart disease in pregnancy include assessment of five axial planes through the fetal abdomen and chest. The aim of this study was to

Table.

Sonographer	2010 (Views 1-4)	2012 (Views 1-4)	2013 (Views 1-4)	2013 (Views 1-5)
	Max 10)	(Max 10)	(Max 10)	(Max 12)
A	7.6	9.6	9.6	11.6
E	7.2	8.7	8.8	10.2
G	9.0	9.4	9.4	11
K	8.6	9.2	8.4	9.4
L	7.2	7.6	8.4	10
O	7.6	7.8	7.4	9.2
P	7.8	8.2	8.8	9.4
Unit Score	7.86	8.64	8.69	10.1
(Mean, 95% CI)	(7.22–8.50)	(7.91–9.38)	(8.01–9.36)	(9.28–10.9)

revise a previously reported objective scoring method to audit cardiac images obtained as part of the routine mid-trimester pregnancy scan.

Methods: Retrospective analysis of routine cardiac images obtained by obstetric sonographers in a tertiary referral centre. All pregnancy screening images in our Unit are digitally archived. Images used had been previously randomly selected for audits in 2010, 2012 and 2013. A revised imaging scoring system was devised and is based on the five axial planes recommended for screening: (1) situs, (2) four-chamber view with appropriate magnification (axis with symmetry, size and position, septum, and crux/offset), (3) left ventricular outflow tract (4) right ventricular outflow tract/three-vessel view and (5) three-vessel and trachea view/aortic arch view. Two independent auditors scored images representing each view and the average value was used for analysis. Scores for each sonographer are an average of images from five patients. Maximal possible score per patient was 10 (all years, views 1 to 4) and 12 (2013, views 1 to 5). Overall Unit score per year is average of individual sonographers.

Results: Average scores per sonographer and Unit scores are summarised on the Table. Based on views 1 to 4, the Unit scores show stability in the years 2010, 2012 and 2013 (ANOVA, ns). Lowest sonographer score was 7.2 out of 10 (2010) and median Unit scores were 7.6, 8.7 and 8.8 (2010, 2012 and 2013 respectively). In 2013, the three-vessel and trachea view was also available for scoring: the lowest sonographer score was 9.2 out of 12 and median Unit score was 10.0

Conclusions: The new, revised scoring system offers a simple and practical way to assess standards of performance, in compliance with the implementation of the new guidelines for screening. It allows cross-sectional and serial comparisons to be made and can be used to provide feedback on quality of screening. This may improve detection rates.

MP4-18

Timing of Diagnosis and Outcome in Readily Treatable Critical Congenital Heart Disease

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Introduction: Surgical results for repair of d-transposition of the great arteries (d-TGA) and for critical aortic arch obstruction with two ventricles (AAO) are excellent. The time of highest risk may be prior to operation, but the magnitude of this risk is poorly defined. Early detection is beneficial, however screening strategies are evolving and the relative benefits of pre- and postnatal detection are likely to vary dependent on the type of heart disease.

Table.

	d-TGA (n = 93)	AAO (n = 50)
Timing of Diagnosis:		
Before Birth	38 (41%)	10 (20%)
Before Hospital Discharge	41 (44%)	15 (30%)
After Hospital Discharge	14 (15%)	25 (50%)
Mortality:		
Presurgical	8 (9%)	9 (18%)
Surgical	0 (0%)	1 (2%)

Methods: We undertook a population-based review of critical congenital heart disease in New Zealand from 2006 to 2010, prior to the use of postnatal oximetry screening. There were 313,478 births of whom 353 were live-born with critical heart disease. The timing of diagnosis, intervention and death, and any comorbidities were recorded.

Results: In those with d-TGA and AAO the timing of diagnosis differed depending on the heart condition ($p < 0.001$) (Table). d-TGA was usually diagnosed before birth or prior to hospital discharge, while AAO was more often diagnosed after hospital discharge ($p < 0.001$), and included 3 cases where the diagnosis was made at postmortem. Except in one case, mortality occurred before any intervention could be undertaken. In d-TGA pre-intervention death ($n = 8$) occurred in association with birth distant to the surgical centre (4), comorbidity (3) and late diagnosis (1), and for AAO ($n = 9$) late diagnosis (6) and comorbidity (3).

Conclusions: The mortality risk for d-TGA and for AAO is greatest prior to cardiac surgery. The relation between timing of diagnosis and risk is lesion specific. In d-TGA, improved prenatal diagnosis allowing delivery close to the surgical centre could further reduce mortality. In AAO enhanced antenatal detection and the addition of postnatal screening may further reduce mortality, although the currently available techniques may limit the incremental gain.

MP4-19

Restriction of foramen ovale in fetuses with TGA – how to predict it?

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Objectives: Transposition of the great arteries (TGA) can be lethal disorder for the neonate even with prenatal diagnosis. The aim of this study was to establish the optimal monitoring for fetuses with TGA to predict urgent balloon atrioseptostomy (BAS), as published criteria are of unsatisfying specificity.

Methods: We evaluated echocardiographic exams of fetuses with TGA diagnosed between 2003–2014 in the referral center for fetal cardiology. Follow-up data were collected from target pediatric cardiology departments.

Results: There were 85 fetuses (male/female ratio ≈ 3.5) with TGA, including 28 with VSD. Median time of diagnosis was 23 weeks (24 in TGA&IVS, 22 in TGA&VSD, ns), slightly decreasing with time. The number of diagnoses increased from 3 in 2003 to 20 in 2013 and 14 in 2014, with growing proportion of 'abnormal 3-vessel-view' as reason for referral.

Karyotype was checked in 12 cases and was abnormal (46,XX,del(11)(q22.1)) in only one, otherwise dysmorphic fetus (associated

defects motivated karyotype examination). Associated cardiovascular anomalies included: pulmonary stenosis or LVOTO(13), RAA(1), and MAPCAs(3).

Appearance and blood flow across foramen ovale (FO) was assessed during every examination. In 36/73 fetuses, its character changed after 30 week of pregnancy. Limited interatrial blood mixing, usually associated with thick septum, was regarded as marker of restriction and predictor of BAS.

57 children were born in our institution, remaining group was excluded from further study. Follow-up was known in 51 cases. 36 newborns needed urgent BAS, which was predicted on fetal echocardiography in 21 cases (58%). Urgent BAS was more frequent in TGA&IVS vs. TGA&VSD group (75% vs. 63%). Hypermobile septum, sometimes described as predictor of BAS, was unspecific (present in 10/11 no-BAS and 27/40 BAS fetuses) and could coexist with both free and limited FO flow (57% and 43% appropriately).

Conclusions: FO flow in fetuses with TGA should be assessed repeatedly, especially just prior to delivery, as it can change during pregnancy. Neonatologists should be informed of suspected FO restriction in a newborn. Diagnosis of restrictive FO in TGA fetuses is difficult. Hypermobile septum is not specific sign of FO restriction. Thick septum primum and impaired blood mixing observed in color Doppler seems more important.

P-1

Ventricular repolarization intervals in children previously treated with anthracyclines

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Introduction: Anthracyclines used in a wide range of childhood hematologic and solid cancers may cause dose-dependent cardiotoxicity. They inhibit DNA and RNA transcription and replication (blocking topoisomerase II) thereby creating iron-mediated free radicals which damage DNA and cardiac myocytes, mechanism underlying cardiomyopathy. Little is known on the longer term effect of anthracyclines on cardiac repolarization.

We investigated ventricular repolarization indices in children after completing treatment with anthracyclines for neoplastic disease; specifically corrected QT interval (QTc), QT dispersion (QTd), T-wave peak to end (TPE) interval, its dispersion and maximum, TPE/QT, TPE/JT, all these being markers of repolarization heterogeneity and potential for ventricular arrhythmias.

Methods: Twenty patients post- anthracycline treatment for childhood neoplasms were studied. Additionally, 80 healthy age matched subjects served as controls. A 12 lead digital ECG was recorded and stored on a server subsequently retrieved digitally on screen. Intervals measured included RR, QT + dispersion, TPE + dispersion, TPE max/QT and TPE max/JT calculated for heart rate correction. Descriptive and analytical statistics were calculated, significance level set at $p = < 0, 05$.

Results: Mean patient age 13 + 4.3 years, time from last anthracycline therapy 5.8 + 4.6 years. Cumulative anthracycline dose: 185 – 480 mg/m². Control group mean age 13.4 + 2.9 years. Repolarization intervals patients versus controls respectively: QTc 423.6 + 20 vs 408 + 24 ms; JTc 324 + 25 vs 315 + 24 ms; TPE dispersion 35 + 13 vs 16 + 6 ms; QT dispersion 32 + 8 vs 12 + 4 ms, maximum TPE 106 + 19 vs 92 + 14 ms; TPE max/QT 0.29 + 0.04 vs 0.25 + 0.03; TPE max/JT 0.38 + 0.06 vs 0.33 + 0.05. All intervals were significantly longer in patients, $p < 0,05$. Four patients developed a dilated cardiomyopathy, one required heart transplantation, one developed ventricular tachycardia.

Conclusion: These intervals reflect spatial and transmural dispersion in ventricular repolarization and may serve as an arrhythmogenic index. They were prolonged in midterm follow up of some anthracycline receiving patients, signifying potential risk for ventricular arrhythmias. Confounding factors may be responsible in the pathogenesis.

P-2

The risk of arrhythmias after surgical repair of anomalous left coronary artery arising from the pulmonary artery (ALCAPA)

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Background: Anomalous left coronary artery arising from the pulmonary artery (ALCAPA) is a rare defect (0.1% of new cases of congenital heart disease), resulting in congestive heart failure secondary to myocardial ischemia and infarction in early infancy. Surgical strategy to construct a two-coronary system for a patient with ALCAPA has evolved with time. Despite apparent normalization of cardiac function in the first years in survivors with a patent dual coronary system, the scar persists. This may be a substrate for ventricular arrhythmias. This study evaluated the risk of arrhythmias after surgical repair of ALCAPA.

Methods: We designed a retrospective, longitudinal, descriptive study that included 18 patients with ALCAPA operated from January 1991 to July 2014. One patient was lost to follow up early and one in recent years. Surgery was performed with direct coronary reimplantation in 14/17 patients and intrapulmonary tunnel (Takeuchi repair) in 3/17.

Results: Mean follow-up was 12 years and 4 months (5 months – 23 years). There were 2 postoperative deaths (overall mortality 11.1%); one early (30 days), due to ventricular arrhythmia and heart failure and the second due to ventricular fibrillation 20 years after surgical repair. Electrocardiographic abnormalities were noted in 10/16 patients; 8 patients had PVCs or ventricular tachycardia; six patients had repolarization abnormalities in inferior and lateral leads. Three of them were on antiarrhythmics, none was considered for ICD implantation or catheter ablation. Mean preoperative left ventricular ejection fraction (LVEF) was 32% (Teichholz). At last follow-up visit, the LVEF improved in all patients to a mean of 66% (ranging from 55 to 81%), with six patients having regional hypokinesis. Fibroelastosis persists in all but one patient. The presence of scar was confirmed by myocardial perfusion scintigraphy and magnetic resonance imaging. These findings correlated with electrical instability of the myocardium.

Conclusions: Treatment of ALCAPA aims to stop process of myocardial ischemia and to restore the normal anatomy of the coronary arteries but deaths may occur due to severe myocardial injury and malignant ventricular arrhythmias. Further research is needed, aiming at early identification of patients at risk of late arrhythmic deaths.

P-3

Electrocardiographic Properties of Patients Diagnosed with Topsy-Turvy Heart

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Topsy-Turvy heart (TTH) is a special type of superoinferior ventricles which was firstly described by Freedom et al. Topsy-Turvy heart is characterized by approximately 90 degrees clock

Table 1.

Diagnosis	Age*	Rhythm	PR		P	QRS	QRS	T
			HR	Interval	Duration	Duration	axis	axis
1 TTH, APW	4	Normal	150	126	90	70	110	-80
2 TTH, APW	4.3	Normal	110	180	100	80	115	77
3 Topsy-turvy heart, Aortic interruption,	2	Abnormal	144	102	46	90	180	87

APW: aortico pulmonary window, HR: heart rate.

*months

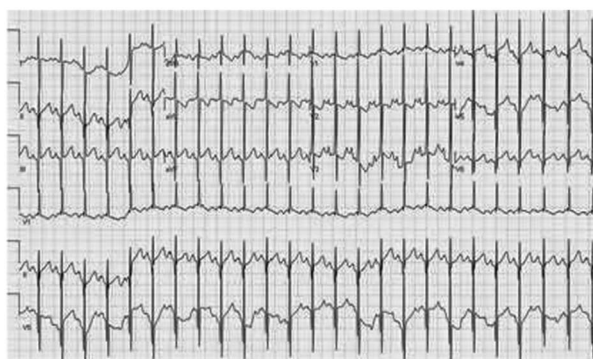
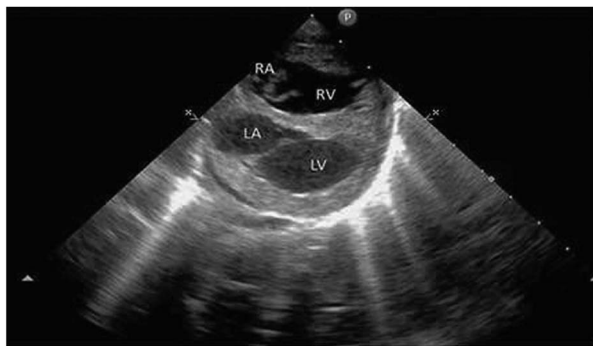


Figure.

wise rotation of the entire heart as a block around the axis from apex to base, leaving the right ventricle in a superior spatial relationship to the left ventricle, while the great arteries are displaced inferiorly and posteriorly in to the distal mediastinum resulting elongation of the brachiocephalic arteries. To date, reports of only six living cases have been published in the literature. There is limited information about the ECG findings of these patients. In this report we discuss three cases of 12 derivatives ECG changes of patients with TTH that shown in table 1.

Different ECG findings were determined in these two different patients with the same diagnosis. Especially the ECG findings of right axis deviation and the QRS-T angle > 60 degrees in all patients were found noteworthy.

P-4

The Utility of Exercise Testing and Adenosine Response for Risk Assessment in Children with Wolff-Parkinson-White Syndrome

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Objective: In Wolff-Parkinson-White (WPW) syndrome, rapid antegrade conduction of atrial tachyarrhythmias can result in ventricular fibrillation and sudden death. Antegrade conduction of accessory pathway (AP) can be assessed through noninvasive testing [exercise stress testing (EST) and adenosine responsiveness of AP] or invasive electrophysiology study (EPS). We aimed to determine the correlation between noninvasive testing and EPS.

Patients and Methods: This prospective, observational study enrolled 40 children (58% male, median age 13 years, and median weight 47.5 kg) with WPW syndrome and candidates for invasive EPS in one year period. EST was performed for all the study participants before EPS, and adenosine administered during the EPS. Conduction through the AP to a cycle length < 250 msec was considered rapid or high risk; otherwise patients were non-rapid or low risk.

Results: The sudden disappearance of the Delta-wave was seen in 10 cases (25%) during EST. AP was found to be high risk in 13 cases (13/40, 32.5%) while the AP was identified to be low risk in 27 cases; however, six (15%) patients had blocked AP conduction with adenosine during EPS. Low-risk classification by EST alone to identify patients with non-rapid conduction on baseline EPS had a specificity of 93%, and a positive predictive value (PPV) of 90% (accuracy 54%). Blocked AP conduction with adenosine as a marker of non-rapid baseline AP conduction had a specificity 93% and a PPV 84%. Finally, AP was adenosine non-responsive in a majority of patients (28/30, 93%) with persistent Delta waves, 40% of those who had sudden disappearance of Delta wave had adenosine responsive AP (p value 0.028).

Conclusion: Abrupt loss of pre-excitation during EST and blocked AP conduction with adenosine had high specificity and PPV for non-rapid and low-risk antegrade conduction during baseline invasive EPS. Successful risk stratification of pediatric patients with WPW is possible through the use of EST and adenosine responsiveness of AP.

P-5

Electrocardiographic and Electrophysiologic Effects of Dexmedetomidine on Children

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Background: Dexmedetomidine (DEX) is a highly selective alpha-2-adrenergic agonist approved for short-term sedation and monitored anesthesia care in adults. Its effects on the electrocardiography and cardiac conduction tissue are not well described in the literature. Therefore, we aimed to characterize the electrocardiographic and electrophysiologic effects of DEX in children.

Patients and Methods: Twenty children (11 boys and 9 girls) ages range between 8 and 17 years undergoing electrophysiological study and ablation of the supraventricular tachycardia, had hemodynamic and cardiac electrophysiologic variables measured before and during the administration of DEX (1 microgram/kg IV over 10 minutes followed by a 10 minute continuous infusion of 0.5 microgram/kg/hour).

Results: A significant decrease in heart rate was seen after the administration of DEX, but the systolic–diastolic–mean arterial pressures, respiratory rate, and end tidal carbon dioxide did not change. Corrected sinus node recovery times and baseline sinus cycle lengths, which are markers of sinus node function, were both lengthened with the administration of DEX. Atrioventricular (AV) nodal function, as evidenced by the Wenckebach cycle length, the ventriculo–atrial block cycle length, and AV node effective refractory periods, were lengthened significantly. We also found that DEX increased the atrial refractory period and diminished atrial excitability.

Conclusions: Dexmedetomidine significantly depressed sinus and AV nodal functions in pediatric patients without significant ECG interval changes, except a trend towards lower heart rates. Although no spontaneous AV nodal block and no clinically significant bradycardia were seen, we recommend that DEX be used with caution in patients at risk for bradycardia and/or AV nodal dysfunction due to its associated comorbidities.

P-6 Evaluation of complete heart block after open heart surgery for congenital heart disease

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Objectives: Cardiac conduction system injury is a cause of postoperative cardiac morbidity following the repair of congenital heart disease. The most important rhythm problem is complete atrioventricular block (CHB) and its frequency varies from 0.7 to 3. Pacemaker implantation is a standard recommendation for patients with persistent CHB more than 7 to 10 days. Our goal was to define the incidence of permanent and transient CHB and compare patients characteristics.

Method: The hospital records of all pediatric patients with early postoperative heart block following congenital heart surgery at our institution between January 2010 and June 2014 were reviewed retrospectively. Patients divided in to two groups. Group-I (transient CHB) and Group-II (permanent CHB). Preoperative diagnosis, peroperative and postoperative clinical status and variables were compared.

Results: Among 1450 congenital cardiac surgeries, 87 (6%) patients had postoperative CHB. There were 59 (4%) patients in Group-I and 28 (2%) in Group-II. All of the patients in Group-II had permanent pacemaker implantation. There were 21 males (36%) in Group-I and 17 (61%) in Group-II. The weight (Group-I: 6 kg (2.9–30); Group-II: 7.6 kg (2.8–50)) and the operation age (Group-I: 6 months (0.1–84); Group-II: 10 months (1–124)) were similar in two groups. The most common diagnosis in Group-I were tetralogy of Fallot (TOF) (n = 14), atrioventricular septal defects (AVSD) (n = 11), ventricular septal defect (VSD) (n = 7) and transposition of great arteries (TGA) (n = 6) and in Group-II TOF (n = 8), VSD (n = 6), AVSD (n = 4), and left ventricular outflow obstruction (n = 4).

Temporary AVB recovered to sinus rhythm within a median of 3 days (1–21 days) after surgery. Permanent pacemakers were implanted at a median of 10 days (7–21 days) after surgery. There were significant differences in ICU stay (Group-I; 6 days (2–25 days) Group-II; 13 days (4–90)) and hospital stay (Group-I; 10 days (2–33 days) Group-II; 20 days (10–90)) between two groups ($p < 0.05$).

Conclusions: CHB is an uncommon complication of congenital heart surgery. TOF and AVSD are at highest risk for CHB postoperatively. Heart block can recover in 2/3 of patients usually at the postoperative ninth day.

P-7 Clinical Spectrum of Primary Cardiomyopathy Associated Ventricular Arrhythmia in Children: 5-year Single Center Experience

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Objectives: The aim of our study was to evaluate the characteristics of ventricular arrhythmias in children with primary cardiomyopathies (CMPs).

Methods: Patients with cardiomyopathy who had visited pediatric cardiology department from January 2010 to December 2014 were retrospectively reviewed. Patients with secondary cause of cardiomyopathy were excluded.

Results: A total of 163 patients with cardiomyopathy was enrolled and grouped according to the echocardiography and MRI findings as follows; Hypertrophic cardiomyopathy (HCMP, n = 71, 43.6%), dilated cardiomyopathy (DCMP, n = 50, 30.7%), isolated noncompaction cardiomyopathy (IVNC, n = 32, 10.6%), restrictive cardiomyopathy (RCMP, n = 7, 4.3%), arrhythmogenic right ventricular cardiomyopathy (ARVC, n = 3, 1.8%).

Twenty-five (15.3%) of 163 patients had premature ventricular contractions (PVCs) and/or ventricular tachycardia (VT) (17 male, 68%; 8 female, 32%). Frequency of ventricular arrhythmia with cardiomyopathies was as follows; HCMP (n = 8, 11.3%), DCMP (n = 9, 18%), IVNC (n = 4, 12.5%), RCMP (n = 1, 14.3%) and ARVC (n = 3, 100%). The mean patient age and the mean follow-up period were 5.3 ± 11.5 years and 12 ± 12.3 months, respectively.

Exercise testing was performed in 4 (16%) patients and sustained/nonsustained VT was not induced in any patient. Atrial fibrillation/flutter was developed in 2 patients with ARVC. PVCs and VT characteristics of patients were mild monomorphic PVCs (n = 7), mild polymorphic PVCs (n = 2), moderate monomorphic PVCs (n = 1), moderate polymorphic PVCs (n = 1), frequent monomorphic PVCs (n = 1), sustained monomorphic VT (n = 1), sustained polymorphic VT (n = 3), nonsustained monomorphic VT (n = 4), nonsustained polymorphic VT (n = 5).

Antiarrhythmic treatment was instituted in 20 cases (80%). ICDs were implanted in 10 patients (2 cardiac arrest, 1 sustain VT, 7 primary). One patient died suddenly, 2 with ARVC, 1 with IVNC and 1 with DCMP patients died for incessant VT, and 1 with ARVD died for multiorgan dysfunction after implantation of biventricular assist device.

Conclusion: VT is a potentially life-threatening arrhythmia because it leads to VF and sudden cardiac arrest especially in cardiomyopathic patients. Consequently, these patients should be monitored closely and carefully for ventricular arrhythmia.

P-8 Premature Ventricular Contractions in Children With a Structurally Normal Heart: A Single Center Experience from Turkey

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Objectives: Premature ventricular contractions (PVCs) in a structurally normal heart generally are a benign condition, especially if

they disappear during exercise. The aim of our study was to evaluate the clinical characteristics and outcomes of otherwise healthy children with PVCs.

Methods: We retrospectively evaluated patients with PVCs by 12 lead ECG, echocardiography, 24-h Holter monitoring (three channel, and 12lead if necessary) and exercise test. Patients were excluded if PVCs was considered to be secondary to any systemic disease, structural cardiac abnormality, or inherited arrhythmias. And also patients with ventricular tachycardia were not included in the study.

Results: Between January 2012 and November 2014, 10260 24-h Holter monitoring was performed. Among them 256 patients had PVCs (135 male (52,7%)/121 female (47,3%)) with a structurally normal heart. Age at the first visit was $11,2 \pm 4,5$ (range: 5 days-17,8 years). 118 patient (46%) were symptomatic. The most commonly associated symptoms were palpitation (n = 70; 27,3%), chest pain (n = 34; 13,3%), syncope (n = 14; 5,5%).

Patients were grouped according to the burden of PVCs as follows; <500 PVCs/24-h (n = 82; 3%), 500-5% PVCs/24-h (n = 93; 36,3%), 5%-10% PVCs/24-h (n = 26; 10,2%), >10% PVCs/24-h (n = 55; 21,5%). In 243 patients (94,9%) uniform PVCs was noted. Ventricular couplets were seen in %16,8 of patients (n = 43).

PVC with left bundle branch block was seen in 60,9% of the patients (n = 156); with right bundle branch block in 8,9% (n = 23); and in 30,1%(n = 77) the origin of the PVC could not be determined.

Antiarrhythmic treatment was instituted in 64 cases (25%). Of the 55 patients with frequent and symptomatic PVCs(>10%/24-h), 4 patients (7,3%) underwent an electrophysiologic study; 3 had inducible ventricular tachycardia and had ablation. The mean period of follow-up was $13,3 \pm 9,6$ months (range: 1-34 months), none of the patients developed left ventricular dysfunction. PVCs disappeared in 34,3% of patients, while PVC frequency decreased in 35%, increased in 9,5%, and was unchanged in 28,5% of the patients.

Conclusion: PVCs in structurally normal hearts have benign prognosis and during follow-up, they may disappear or decrease in time.

P-9

Electrocardiographic and 24 h-Holter characteristics of patients with Ebstein anomaly

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Objectives: A retrospective cohort study was conducted in a tertiary cardiac surgery centre, to determine the electrocardiographic and 24-Holter characteristics of patients with Ebstein anomaly.

Patients and Methods: Echocardiographic, electrocardiographic and 24-Holter characteristics of 41 patients with Ebstein anomaly patients diagnosed between January 2011 to November 2014 were retrospectively reviewed.

Results: Twenty one patient were male. median age at referral to our clinic was 2 years. Among 41 patient 13 were referred during newborn period, 5 were between 1 month and 1 year and 23 were between 1 year and 18 years. All of the patients had either atrial septal defect or patent foramen ovale. Echocardiographic accompanying pathologies were ventricular septal defect (3/41), corrected transposition of the great arteries (3/41), anatomically corrected transposition of the great arteries (1/41) and all newborn patients had ductus arteriosus. Electrocardiography revealed first degree heart block (2/41), atrial flutter (1/41), nonspecific intraventricular conduction delay/block (5/41), pre-excitation-WPW pattern (5/41), right bundle branch block (3/41), complete heart block (1/41), and supraventricular extrasystole (1/41). Additionally 24-hour-Holter

monitoring revealed in 4 patients intermittent pre-excitation (total number of pre-excitation 9), 6 patients first degree heart block (total number of first degree heart block 8), 1 patient ectopic atrial rhythm. Eleven of the 24-hour-Holter monitoring were in normal limits.

One patient died in the neonatal period due to intractable supra-ventricular tachycardia (atrial flutter) resulting in heart failure. Pacemaker implantation was performed to another patient with complete AV block. Her diagnosis was corrected transposition of the great arteries with Ebstein anomaly. Five patients underwent six catheter ablation of an accessory pathway procedure. One of the patients had accessory pathway with Mahaim-type preexcitation. Also one patients ablation procedure was unsuccessful due to epicardial accessory pathway.

Conclusions: In our series most common diagnosis were first degree heart block 8/41 (20%) and preexcitation and/or Wolff-Parkinson-White syndrome prevalence was 9/41 (22%). Intermittent rhythm disturbances can be diagnosed with 24-hour-rhythm-Holter recordings. We suggest 24-hour-rhythm-Holter monitoring for all patients diagnosed with Ebstein anomaly with regular intervals.

P-10

Arrhythmia in Fontan Patient

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Introduction: Arrhythmias may develop in the Fontan patient soon after the procedure is performed. With longer duration of follow-up, as many as 50% of Fontan patients will develop atrial tachycardia.

The incidence and type of arrhythmia occurrence are examined in our patients with Fontan and risk factors for development of these arrhythmias are considered.

Methods: We reviewed the outcomes of 162 consecutive patients who underwent the Fontan operation between 1975 and 2005 (75 patients had an atriopulmonary connection (mean follow up 15 years) and 87 patients had an bicavopulmonary connection (TCPC)(Mean follow up 8,85 years)).

Results: In atriopulmonary connection the most frequent arrhythmia is the atrial re-entry and In the TCPC surgery is the sinus node dysfunction.

In our series, arrhythmias were not associated to a higher rate of transplantation, death, protein-losing syndrome nor plastic bronchitis and associated to worse functional class III and IV (χ^2 7,87: p<0,01) Factors enhancing the possibilities of arrhythmia were significant: Systemic atrioventricular valvular regurgitation (AVR) (χ^2 26,97: p<0,001) and ventricular dysfunction (χ^2 20,86: p<0,001). No significant association was found related to subaortic stenosis, mean pulmonary arterial pressure, pulmonary vascular resistance, ventricular end-diastolic pressure, pulmonary artery sizes or fenestration and age at the time of surgery.

Table.

	Atriopulmonary connection (63%)	Bicavopulmonary connection (16%)
No arrhythmia	27 (36%)	73 (83%)
Perioperative arrhythmia	5 (6.6%)	2 (2.3%)
Arrhythmia in the immediate postoperative	16 (21%)	1 (1%)
Late arrhythmia	27 (36%)	11 (12%)

Conclusions: Arrhythmias are one of the main complications in univentricular surgery associated to worse functional class and leads to an unavoidable impairment of ventricular function that favors arrhythmia maintenance.

Arrhythmias are evidently more frequent in the atriopulmonary connection.

We only found significant association related to systemic atrioventricular valvular regurgitation and ventricular dysfunction.

P-11

Electrocardiographic abnormalities and rhythmic events in a pediatric cohort of myotonic dystrophy I

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Introduction: Myotonic dystrophy (MD1) is an inherited muscular dystrophy responsible for myocardial degeneration of the cardiac conduction system, with a high risk of sudden deaths in adults. Long-term clinical course and ECG findings in a pediatric cohort have yet to be assessed.

Methods: We retrospectively collected clinical and electrocardiographic data in patients with proven MD1 followed at our neuromuscular disease center. An arrhythmic event was defined as syncope or sudden cardiac death of presumed arrhythmic etiology. PR interval, QRS width, and axial deviation are compared to standard data described by Davignon *et al.* and defined as abnormal when values are superior to the 98th percentile for PR and QRS durations, and out of the 1st and the 98th percentiles for axial deviation.

Results: A total of 56 patients were included with a mean age at diagnosis of 5.3 ± 4.9 years and a follow-up duration of 7.2 (range 1.9–15.6) years. Of these patients, 27 cases (48.2%) were congenital MD1 with a reported diagnosis in the first year of life. At baseline, 21.4% (n = 12) had PR values superior to the 98th percentile and 78.5% (n = 44) had a widening of QRS. By the end of a follow up, these conduction abnormalities do not regress with PR prolongation notified in 25.0%, and widening of QRS in 87.5%. Only 12.5% (n = 7) in the cohort had normal ECG findings at the end of follow-up.

Over the course of follow-up, 5 patients had syncope (mean age 15.1 ± 3.7) and 2 patients died suddenly (mean age 17.5 ± 1.9), corresponding to an event-free survival rate of 98% at 7.2 years. All of these patients had abnormal ECG findings but, in univariate analyses, only the width of QRS is associated with arrhythmic events (110.3 ± 8.2 ms in the event group vs. 98.1 ± 1.7 ms in the no-event group, $p = 0.03$).

Conclusion: During a follow up nearly 16 years, more than 85% of pediatric patients who suffered from MD1 had ECG abnormalities, which increased over time. Rhythmic events in pediatric population are rare, and occur in patients with conduction abnormalities on basal ECG.

P-12

Using of the 12-lead electrocardiogram to determine the site of ventricular arrhythmia in children

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Introduction: There are a number of studies focusing on determining the site of ventricular arrhythmia (VA) origin.

However, electrocardiographic (ECG) patterns of VA for children have not been reported yet. The goal of the study was to determinate the ECG characteristics of the most common sites of VA in children.

Methods and Materials: 12-lead ECG of 246 children aged from 5 to 17 years (mean age 13.4 ± 3) with VA were investigated. The origin of VA was confirmed during the endocardial mapping. ECG characteristics of ventricular complexes such as QRS axis, the QRS morphology, the ratio of R and S waves in precordial leads, the QRS duration in II and V2 were assessed.

Results: According to endocardial mapping VA originated from right ventricular outflow tract (RVOT) in 58,5% patients (pts), left aortic sinus of Valsalva (LVS) in 17,9% pts, right ventricular inflow tract (RVIT) in 12,6% pts. Other loci had 11% of patients. RVOT loci characterized with both normal and right axis deviation. The morphology of QRS in this group was QS or rS/RS in V1, R \leq S in V3, amplitude of R wave ≤ 30 mm in lead III and QRS duration ≥ 120 ms in II. The ECG morphology of LVS showing only inferior axis together with Rs or rS/RS pattern in V1 and R > S in V3, amplitude of R wave > 30 mm in lead III. The origin in RVIT was associated with left axis deviation of ventricular complex, RS or rS/QS pattern in V1 and QRS duration > 120 ms in V2.

Conclusion: The origin of ventricular arrhythmia using surface ECG can be localized with the accuracy 91%.

P-13

Comparative analysis of transmural dispersion and atrial natriuretic peptide (ANP) concentration in plasma as risk marker of arrhythmogenic dysfunction in children with hypertrophic cardiomyopathy

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Objective: Assessment of ANP interrelation in plasma with the index of transmural dispersion of repolarization at treadmill testing in children with HCM.

Patients and Methods: 46 children (aged 14 to 17 years) with left ventricular hypertrophy were examined. The control group consisted of 20 healthy children. Functional diagnostics and laboratory tests included ECG, Holter, echocardiography, treadmill test with transmural dispersion (TDR) (normal 69–92 ms) assessment. All the patients underwent laboratory evaluation of ANP concentration in plasma.

The Results: HCM was diagnosed in 26 of 46 patients (mean age 15.7 ± 2.2 years). The patients were divided into two subgroups. The 1st subgroup (n = 11) - patients with asymmetrical obstructive LVMH (wall thickness > 20 mm). The 2nd subgroup (n = 15) - patients with nonobstructive symmetric LVMH (wall thickness < 20 mm).

In the 1st subgroup (n = 11) the level of ANP was higher than normal (1,92nmol/l) in 7 of 11 patients (64%). In the 2nd subgroup (n = 15) increased values of ANP were observed in 5 of 15 patients (33%). In the 1st subgroup ANP concentration was 47% higher than in the 2nd one (3,34nmol/l of 1,57nmol/l). In the control group ANP concentration corresponded to the norm (0,89nmol/l) in 100% of patients. When comparing the average values of ANP in all the groups significant differences of these parameters ($p = 0,002$) were found.

In the 3rd minute of a cool down period the episodes of TDR elongation were recorded in 7 of 11 subjects (64%) in the 1st subgroup and in 11 of 15 patients (7.3%) in the 2nd subgroup. TDR values and ANP in the control group corresponded to normal levels during the whole study period. The correlation coefficient between TDR and ANP values was equal to 0,46 ($p = 0,002$).

Conclusions: Significantly positive correlation between concentration level of ANP and TDR value was revealed at the 3rd minute of a cool down period during stress testing, which allows to identify prognostic risk of life-threatening arrhythmias and sudden cardiac death in patients with HCM.

P-14

Growth-related change in effect of electrotonic interaction between dual nodal pathways in children with atrioventricular nodal reentrant tachycardia

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Background: Atrioventricular nodal reentrant tachycardia (AVNRT) is less common in children, possibly due to immaturity of the atrioventricular node (AVN). Electrotonic interaction between fast and slow pathways plays an important role in the development of AVNRT, especially in slow-fast type and slow-slow type, by lengthening the refractory period of fast pathway. We hypothesized that the interaction significantly affected older children but not infants. In the present study, we sought to find out the relationship between body size and the involvement of the interaction.

Methods: Retrospective observational study was conducted. Patients who were diagnosed with AVNRT and received radiofrequency catheter ablation with successful slow pathway discontinuation were included in the study. Patients with comorbid congenital heart diseases or with fast-slow type were excluded from the study. Fast pathway effective refractory periods (FP-ERPs) before and after the procedure were compared. Δ FP-ERP was defined as the difference between the FP-ERPs. Correlation coefficient between Δ FP-ERP and body surface area (BSA) of the patients was obtained to describe the influence of growth on the interaction between dual nodal pathways.

Results: Eleven cases with median BSA of 1.22 m² (range 0.78 – 1.51 m²) were included in the study. Fast pathway effective refractory period was significantly shortened after slow pathway disconnection (403.8 ± 95.8 ms vs. 305.6 ± 91.5 ms, $P < 0.001$). Furthermore, strong positive correlation was found between Δ FP-ERP and BSA ($r = 0.659$, $P = 0.028$).

Conclusion: The results of the study indicated that FP-ERP lengthened as body size increased. That might explain why AVNRT is more common in adolescents and adults than in younger children.

P-15

Catheter Ablation in Pediatric Permanent Junctional Reciprocating Tachycardia

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Background: Permanent junctional reciprocating tachycardia (PJRT) is an infrequent type of atrioventricular reentry tachycardia, which substrate is a right posteroseptal accessory pathway (AP) with decremental properties. Almost incessant in infancy, the risk of tachycardia-induced cardiomyopathy is high. Management of this arrhythmia is challenging because of its frequent refractoriness to drug therapy. Catheter ablation (CA) has arisen as a safe and effective therapeutic option.

Objective: To describe the characteristics and results of CA of PJRT in children.

Methods: We retrospectively reviewed 21 CA in children with PJRT at our institution between July 2009 and December 2014.

Results: PJRT was diagnosed within the first year of life in 57% of the cases. Antiarrhythmic medications were used as first line therapy in 85%, while CA was performed as first line therapy in only 3 patients. Amiodarone was the most frequent used drug: 16 patients were or have been treated by long term Amiodarone prior to CA procedure. Amiodarone was associated with at least one other antiarrhythmic drug in half of the patients. 23 CA procedures were performed in 21 patients with a success rate of 86%. The mean age at CA procedure was 9 years and mean weight 30 kg. No major complications were reported. All AP were localized to the posteroseptal area. CA was performed through the coronary sinus with a 4 mm irrigated-tip radiofrequency catheter. Angiography of the left coronary artery was performed prior and after radiofrequency delivery for all children weighting less than 20 kg. Recurrence occurred in 3 patients: one year after the CA procedure for one patient, and within 24 hours for 2 patients. Overall, at one-year follow-up, all by one lost of follow up patient were on sinus rhythm without drug therapy.

Conclusion: CA is an effective therapeutic option for PJRT in children. Success rate is high, and despite recurrence antiarrhythmic medications can be stopped in all patients.

P-16

Effect of biventricular pacing in young patients evaluated by Three-Dimensional Echocardiography

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Introduction: The aim of this study is to evaluate long-term results of Cardiac resynchronization therapy (CRT) in a pediatric and CHD (Congenital Heart Defect) population and utility of 3-D echocardiography (3DE) to optimize VV interval.

Methods: We retrospectively analyzed patients who underwent CRT between 2006 and 2012 in our institution. For individualized VV interval optimization 3DE full-volume datasets of the left ventricle were obtained and analyzed to derive a systolic dyssynchrony index (SDI). Response to CRT was predefined as a minimum 5% proportional increase in EF over baseline measurements. We performed a multiparametric echocardiographic assessment of LV dyssynchrony using standardized parameters: IVMD, M-mode Pitzalis index, Morphology of the pulsed Doppler mitral valve. Routine baseline 2DE and 3DE, were performed in all patient according to institutional guidelines. 3DE guided optimization of the CRT device was performed the day after implantation. The device was programmed for the VV interval with the lowest SDI. Follow-up was at 1 month, 3 months and 6 months.

Results: Twenty patients (12 M- 8 F), age at implantation 14 years (5 – 31 years), have been followed-up for 3.4 years (1.2 – 8.1 years). All patients had advanced or complete AV block (CAVB). Thirteen patients (65%) showed severe systolic LV dysfunction (LVD) without CHD and 7 patients (35%) with CHD and LVD. Individualized optimization significantly reduced SDI from 12.3% to 5.3%. Four patients (20%) with CHD and LVD were non-responders. Among the 16 responders (80%), 12 patients had CCAVB and previous RV pacing (67%), 3 had complex CHD and LVD and the last s/p ablate and pace. Partial results after 1 month: SDI significantly reduced in 4 patients (25%) and EF in 2 patients (12.5%). Partial results after 3 months: SDI significantly reduced in 7 patients (44%) and EF 3 patients (19%). Partial results after 6 months: SDI significantly reduced in 12 patients (75%) and EF 10 patients (62.5%).

Conclusions: CRT could improve the LV function in young patients with heart failure and SDI quantification could predict of LVEF after CRT. We can use in both acute and late phase to optimize the VV delay.

P-17**Characteristics of pediatric Brugada Syndrome**

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Introduction: Brugada syndrome (BS) is a primary cardiac electrical disorder, which manifests as coved-type ST-segment elevation, either spontaneously after the administration of intravenous class I antiarrhythmic drugs. The syndrome predisposes to ventricular arrhythmias eventually leading to sudden cardiac death (SCD).

However, diagnosis of BS is sometimes difficult in children.

Methods: Children for the evaluation and diagnosis of BS, and definite diagnosis of BS were included in this study.

Results: The consecutive 9 patients with BS (mean age 10.4 ± 5.6 years, M:F = 8:1) were included in this study. The initial onset was syncope in 3 patients (1 was due to vasovagal syncope, and the other 2 were due to VT), heat screening in 3 patients, coincident recording of electrocardiogram in 2 patients, and familial study in 1 patient. In these patients, 5 of 6 patients had SCN5A mutation and 1 patient was under investigation of genetic anomaly. Electrophysiological Study (EPS) was performed in 4 patients, ventricular tachycardia (VT) was induced in 2 patients, and ventricular fibrillation (VF) in 1 patient. Associated arrhythmias were: sick sinus syndrome (SSS) in 4 patients, VT in 2 patients, and atrial tachycardia (AT) in 2 patients, and supraventricular tachycardia (SVT) in 1 patient. Implantable cardioverter defibrillator (ICD) was implanted in 2 patients. Catheter ablation for atrial tachycardia was performed in 2 patients. During mean follow up of 6.7 ± 5.0 years, 1 patient detected VT by remote monitoring of ICD, however no other patients developed VF, syncope or sudden death.

Conclusions: In this study, none of the patient documented spontaneous syncope due to VF. Incidence of SCN5A anomaly is higher (5/6 patients, 83%) than adult patients. Supraventricular arrhythmia (SSS, AT, SVT) was noted more than half of the patients. And also, monomorphic VT was noted 22% of the patients. In children, fibrosis of right ventricular outflow tract may not be profound, that may reduce the possibility of the occurrence of VF or polymorphic VT. Catheter ablation of AT, or SVT may be useful to prevent inappropriate discharge of ICD in children.

P-18**RFA of drug-refractory arrhythmias in small children under one year old**

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The aim of the study was to evaluate the safety and efficacy of RFA in critically ill small children (<1 year of age) with drug resistant tachycardia accompanied by arrhythmogenic cardiomyopathy and heart failure.

Material: The study included 18 patients aged 4.8 ± 3.7 months. Wolff-Parkinson-White syndrome and atrial tachycardia were detected in ten (53.3%) and eight (46.7%) patients, respectively. Patients with structural heart pathology, including congenital heart diseases and laboratory-confirmed myocarditis, were excluded from the study.

Results: The indication for RFA was drug refractory supraventricular tachycardia (SVT) accompanied by arrhythmogenic cardiomyopathy and heart failure. Unsuccessful ablation was observed in two 1-month-old patients who underwent successful ablation 3 months later. The follow-up periods ranged from 0.5 to 9 years (average 4.1 years). Only two patients (11.1%) had tachycardia recurrence 1 and 2 months after RFA, respectively. The RFA success rate was 88.8%. The study did not show any procedure-related complications. Heart failure disappeared within 5–7 days after RFA. Complete normalization of cardiac chambers sizes was documented within 1 month after effective RFA. A three-dimensional CARTO system was used in three patients with body weight > 7 kg. The use of the CARTO system resulted in a remarkable decrease of the fluoroscopy time without vascular injury or other procedure-related complications in all cases.

Conclusions: Our study suggested that RFA may be considered as the method of choice for SVT treatment in small children when drug therapy is ineffective and arrhythmogenic cardiomyopathy progresses. **Key words:** supraventricular tachycardia, infants, radiofrequency ablation.

P-19**Heart rate variability in single ventricle patients- before and after Glenn surgery**

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Introduction: Atrial arrhythmias and sinus node dysfunction are well-known complications in patients with Fontan circulation. In the Glenn procedure, preceding TCPC, the SVC is divided at the entrance to the heart. In this area cardiac autonomic ganglia are located. Thus, there is a risk of damage to the autonomic ganglia at the procedure, potentially affecting cardiac autonomic innervation which theoretically would raise the risk for arrhythmia. Heart rate variability (HRV) can be used to evaluate the autonomic nervous control.

Our aim was to investigate HRV in ambulatory 24-hour ECG recordings (Holter- ECG) in a cohort of children with TCPC, focusing on HRV changes after Glenn surgery.

Methods: Patients with single ventricle physiology underwent 24-hour ECG during daily activity; 35 before and 27 after Glenn surgery. HRV was analyzed with power spectrum analysis. Total variability and RR-intervals were measured from Holter-ECGs performed before and after Glenn surgery. Data were expressed as z-scores based on the age development in the control group (n = 36 healthy children). We used Kruskal-Wallis test for comparison of ranks in the three groups.

Results: Group means and standard deviations are presented in Table 1. In patients before Glenn-surgery: RR-interval was normal compared to controls. Patients had lower HRV than controls ($p < 0.05$). Twelve patients showed reduced HRV (< 2 z-score).

In patients after Glenn-surgery: RR interval was longer than controls and patients before Glenn-surgery ($p < 0.05$). Patients had lower HRV than controls ($p < 0.05$). Nine patients showed reduced HRV (z-score < 2).

Conclusions: We found a significant reduction in RR-intervals in patients with single ventricle physiology after Glenn surgery. This finding is interesting since low heart rate may indicate sinus node dysfunction. We also found a reduced HRV in single ventricle patients compared to health controls both before and after Glenn surgery.

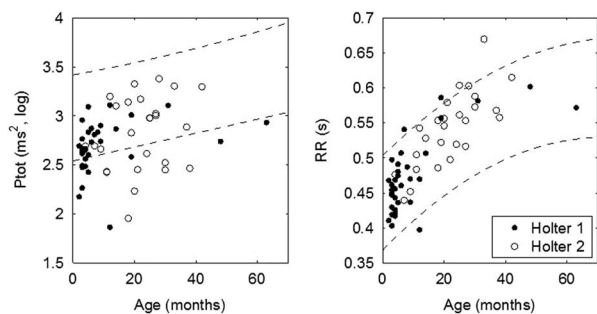


Figure 1. HRV as represented by total power. Lines indicate regression lines for controls and 95% confidence intervals.

Table 1.

	Controls (n = 36)	Before Glenn (n = 35)	After Glenn (n = 27)
RR (s)	0.52 SD = 0.07	0.47 SD = 0.05	0.54 SD = 0.05
Plot (ms ² , log)	3.15 SD = 0.26	2.69 SD = 0.26	2.83 SD = 0.3

P-20

The prevalence of Turner Syndrome in girls who underwent surgical repair for Coarctation of Aorta; A 10 year single centre UK study

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Objective: To identify whether genetic tests were performed in female infants who underwent surgical repair of Coarctation of aorta (CoA) and evaluate the occurrence of Turner syndrome.

Study design: A total of 38 females who underwent surgical repair for CoA as a primary procedure in the last 10 years were identified. Patient data between March 2004 and January 2014 were analysed for genetic test results, availability of genetic counselling and prevalence of Turner syndrome in the patients.

Results: Of 38 females with CoA, 21 (55%) had karyotype analysis done. 3 out of the 21 patients were found positive for Turner syndrome. Genetic tests were not done for the rest 17 patients. This translated into a prevalence of 14.2% in the cohort with a minimum prevalence of 7.8% should the rest females be negative for Turner syndrome. All the patients who underwent genetic tests were offered counselling but 5 patients (28%) attended counselling.

Conclusion: Our study demonstrated for the first time in Europe that a minimum of 7.8% of girls presenting with CoA were found to have Turner syndrome on karyotyping. Turner syndrome has a huge spectrum of preventable and treatable health problems which can be addressed more effectively if diagnosed early. We propose that all girls with CoA should have a karyotype analysis at the time of diagnosis of CoA.

P-21

Evaluation of GATA4, NKX2.5, TBX5, CRELD1 and BMP4 genes with MLPA technique in patients with congenital heart disease

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Background and objective: Congenital heart diseases (CHD) are the most common birth defects. Nevertheless, the etiology of the

majority of these illnesses remains unknown. Genetic and environmental factors are important in the etiology of CHD. Cardiac development is a complex and multifactorial biological process. Heterozygous mutations in some genes were defined in CHD.

Methods: In this study, we investigated the presence and frequency of mutations in the NKX2.5, GATA4, BMP4, TBX5 and CRELD1 genes on 255 patients with CHD. Mutational analysis was performed using multiplex ligation-dependent probe amplification –MLPA– technique.

Results: There was ventricular septal defect (VSD) in 144 patients, atrial septal defect (ASD) in 60 patients, complete atrioventricular septal defect (AVSD) 14 the patients and cyanotic congenital heart disease in 37 patients. We identified total 24 mutations in the 20 patients with CHD. We did not find any mutations NKX2.5 and CRELD1 genes in the patients with CHD. Mutations of the GATA4, BMP4 and TBX5 genes were detected in the five patients (VSD:3 and ASD:2), five patients (VSD:4, ASD:1) and a patient with VSD, respectively. There was MSR1 gene mutation in the six patients (Tetralogy of Fallot: 3, VSD: 3).

Conclusions: GATA4, BMP4, TBX5, MSR1 gene mutations could be important in congenital heart disease pathogenesis. However there was need the studies performed in larger numbers of patients used with CHD used the MLPA technique in this issue.

P-22

Genetical and clinical features in inherited pediatric LQT syndrome: single center data

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Introduction: Long QT syndrome is an inherited disorder characterized by Qt interval prolongation. Often first manifestation of the disease is sudden cardiac death.

More than 1000 pathogenic genetic variations have been identified in 17 genes, encoding ion channel proteins or functional subunits associated with ion channels, most frequent genes involved are KCNQ1(LQTS1), KCNH2(LQTS2) and SCN5A(LQTS3). We present clinical data and genetic mutations in our pediatric LQTS.

Methods: Retrospectively, we revised clinical and genetic features of 52 pediatric patients with diagnosis of LQTS: epidemiological data, clinical features, age at diagnosis, QTc interval, modified Schwartz score, Holter/exercise stress test, genetic counseling and pharmacological and non-pharmacological therapy.

Results: 52 pediatric patients with diagnosis of LQTS (1 m-18 years, mean 7 years). In 6 cases (11.5%), pediatric patient was the index case and relatives were diagnosed later. Most frequent symptoms at onset were palpitations and syncope.

In 4 cases (7.6%) the first manifestation was sudden cardiac death (2 not recovered), one of the recovered patients during exercise (swimming) with KCNQ1 mutation and the other one during a minor non-cardiac surgery with SCN5A mutation. Genetic testing was performed for any index case in which LQTS was suspected. All patients with modified Schwartz score ≥ 3 (48%) and those patients included in the cascade screening had a genetic testing, in order to identify of all affected family members.

Genetic features: 42% of our patients had genetical mutation identified (59% KCNQ1, 22% KCNH2, 18% SCN5A, 9% CACNA1C). Propranolol and Bisoprolol was preferred in LQTS1 and LQTS2. In two patients with LQTS3 Flecainide treatment was started. Despite pharmacological treatment, a left cardiac

sympathetic denervation was performed in two cases due to cardiogenic syncope, with clear clinical improvement.

In our cascade familial screening, we detected 9.6% silent mutation carriers and 3.8% of patients were negative for the known parent's mutation. At data, no ICD was needed in our pediatric patients.

Conclusions: Sudden cardiac death related to LQTS can be the first manifestation, in our data 7.6% of all patients.

More than 80% of genetic mutations were KCNQ1, KCNH2 and SCN5A, and accordingly to published literature.

It's important to determine genetic mutations detecting both the non-carriers and silent carriers who must be followed to detect ECG changes or clinical symptoms and avoid malignant arrhythmias and sudden death.

P-23

GATA 4 sequence variation in Egyptian children with congenital heart disease

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Introduction: GATA 4 is a transcription factor that is expressed in the heart and is essential for cardiac development. Recent studies show relation between GATA 4 sequence variation and congenital heart disease.

The aim of this work is to investigate relation between GATA 4 sequence variation and congenital heart disease (CHD) among Egyptian children.

Methods: A total of 48 cases 38 cases of CHD of different types and 10 control cases were included in our study. Exon 1 of GATA 4 was investigated for sequence variants using denaturing high-performance liquid chromatography or conformation-sensitive gel electrophoresis. Samples showing peak or band shifts were reamplified from genomic DNA and sequenced.

Results: Novel mutation in Exon 1 in GATA 4 was found in 5 cases of CHD 13% (3 cases of Isolated Ventricular septal defect and 2 cases of Ventricular septal defect with atrial septal defect). Mutation was found in position 193 in exon 1 with base substitution (A) instead of (C) resulting in Amino acid change Histidine instead of Proline.

Conclusion: There is a significant correlation between Ventricular septal defect and GATA 4 sequence in Egyptian Children. We need to further investigate this relation by having larger scale of cases and focusing on Ventricular septal defect.

P-24

Involvement of Proline-rich tyrosine kinase 2 (Pyk2) in Kawasaki disease-like murine vasculitis

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Introduction: Although the pathogenesis of Kawasaki disease (KD) is still unclear, some studies suggest the involvement of the innate immune to the vasculitis. Pattern recognition receptor (PRR) plays an important role in the immune system. Mice of vasculitis induced with *Candida albicans* water-soluble fraction (CAWS) is widely used as an established model of KD, and CAWS is already known as a ligand of dectin-2 which is one of PRR. Furthermore,

proline-rich tyrosine kinase (Pyk2) activates NF- κ B in macrophage via the PRRs signaling. Hence, we examined whether Pyk2 involves in the onset of KD-like vasculitis by CAWS.

Methods: Pyk2-knock out (Pyk2-KO) and wild-type C57BL/6 mice (WT) were administered CAWS to induce KD-like vasculitis. At 1 and 28 days after CAWS injection, mice were sacrificed. We determined immunohistochemically the onset of experimental vasculitis. NF- κ B activation was evaluated by quantifying nuclear translocation of NF- κ B p65 subunit in peritoneal macrophages isolated from mice *in vitro*. Serum cytokines and chemokines across each mice were compared by cytokine array.

Results: Pyk2-KO mice didn't show any apparent defective phenotype. While marked inflammation was observed in the aortic root including coronary bifurcation of CAWS-treated WT mice on day 28, such vasculitis was barely detected in CAWS-treated Pyk2-KO mice. On day 1, it is before the onset of vasculitis, some cytokines/chemokines involved in migration of lymphocyte or vascular smooth muscle cells increased in WT mice and decreased in Pyk2-KO mice. CAWS-induced NF- κ B activation was also less observed in macrophages from Pyk2-KO mice.

Conclusions: It is possible that Pyk2 is involved in the pathogenesis of KD through producing cytokines/chemokines related with migration of inflammatory cells. Pyk2 may have therapeutic potential for the treatment of KD.

P-25

Human cardiac progenitor cell seeded-collagen patches for cell therapy applied to right ventricular dysfunction:

Preliminary results in a large animal model

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Background: Cell therapy using intramyocardial injections of cardiac progenitors differentiated from human embryonic stem cells showed benefits on overloaded right ventricular (RV) tissue remodelling and arrhythmic susceptibility but this delivery mode failed to improve RV function. Our aim was to evaluate in a porcine model of overloaded RV dysfunction a new delivery mode of such therapy.

Methods: A combined overloaded RV dysfunction was obtained in piglets using a surgical procedure mimicking repaired tetralogy of Fallot. After 4 months, cell therapy was surgically administrated using 2 types of human NKX2.5+ cardiac progenitor cell-seeded collagen patches applied on the epicardium: QGel® and pressured-patches. Myocardial function was measured 1 month after transplantation by conductance catheter technique and echocardiography (standard and strain). The fate of progenitors was studied using antibodies directed against human Nkx2.5, human mitochondria, GFP, CD31, actinin and Islet1.

Results: All pigs survived without any complication. Pressured-patches allowed human progenitors to survive well and to migrate away from the epicardium while QGel® patches restricted the cell migration. Progenitors differentiated close to the epicardium toward the cardiac lineage as assessed actinin expression. NKX2.5+ cells within pressured-patches migrated between myocardial fibers.

Only pressured-patches (N=3) tended to improve the contractility (Emax slope). By contrast, this parameter decreased in QGel® patches animals (N=2). Moreover, in 2 pressured-patch animals, standard echocardiographic functional parameters (FAC, TAPSE, s'wave) were maintained while 2D strain and strain rate values increased.

Conclusion: Cell therapy using seeded-patches was more conservative for engrafted cells than intramyocardial injections but only pressured-patches seemed to give benefits on overloaded RV function and contractility. These first promising results need to be checked at longer term.

P-26

Endothelial function in adolescents with newly diagnosed essential hypertension

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Introduction: Hypertension is associated with higher risk of atherosclerosis. Microvascular endothelial dysfunction is considered to be an early indicator of atherosclerotic changes preceding the morphological alterations, commonly associated with elevated blood pressure. However, the findings regarding the cause-effect question remain controversial and the pathophysiology of this relation is still not fully understood. Therefore, we aimed to study the endothelial function in adolescents at the onset of essential hypertension using peripheral arterial tonometry.

Methods: Fifteen nonobese adolescent males (age: 17.4 ± 0.4 years, BMI: 22.9 ± 0.6 kg/m²) with newly diagnosed essential hypertension and 15 age-matched healthy adolescents as a control group were examined. Continuous recording of the pulse wave amplitude was performed using peripheral arterial tonometry device EndoPAT 2000 (Itamar Medical, Israel) under standard conditions during three 5-minute phases: baseline period, ischemic phase, postischemic phase. Reactive hyperemia index (RHI) was calculated automatically by the EndoPAT software.

Results: Statistical analysis revealed no significant differences in the RHI between hypertensive and control group (2.12 ± 0.14 vs. 2.18 ± 0.10 , $p = 0.384$). Both values were within physiological range (RHI 1.67 and above).

Conclusions: Microvascular endothelial function indexed by RHI was not altered in adolescents with newly diagnosed essential hypertension compared to healthy controls. We suggest that endothelial production of nitric oxide likely remains at the physiological level in newly diagnosed hypertension or other endothelial substances could compensate its deficit. Therefore, these findings could support the hypothesis of endothelial dysfunction being rather the effect than the cause of hypertension. Large longitudinal studies are needed to confirm this assumption.

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P-27

Non invasive ultrasonic chordal cutting

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Objective: Chordal cutting targeting leaflet tethering has been described to improve the efficacy of annuloplasty during ischemic mitral regurgitation surgery. Histotripsy is a novel ultrasound based technique for tissue fragmentation through the cavitation generated at the

focus of a very intense ultrasonic pulse. In this study we investigate the feasibility of using histotripsy for chordal cutting to avoid cardiopulmonary bypass and invasive surgery in infarcted heart.

Methods: Experiments were performed in vitro in explanted sheep heart (N = 5) and in vivo in sheep beating heart (N = 3, 40 + /- 4 kg). In vitro, the mitral valve basal chordae was removed, fixed on a holder in a water tank. The ultrasonic pulses were emitted from the therapeutic device (1-MHz focused transducer, pulses of 8µs duration, peak negative pressure of 17 MPa, repetition frequency of 100 Hz) placed at a distance of 64 mm.

In vivo, we performed sternotomy and the device was applied on the thorax cavity was filled out with water. We analysed MV coaptation and chordae by real time 3D echocardiography before and after chordal cutting. The animals were sacrificed at the end of the procedure, for postmortem anatomical exploration to confirm the section of the basal chordae and the integrity of the remaining marginal chordae.

Results: In vitro, all the basal chordae were completely cut. The mean procedure time was 6 (+ /-3) minutes. The thickness of the chordae was the main criteria affecting the duration of procedure. In the sheep, central basal chordae of anterior leaflet were completely cut. The mean procedure time was 19 (+ /-9) minutes. By echography, the sectioned chordae was visible and no mitral valve prolapse was found. All the postmortem anatomical exploration of hearts confirmed the section of the basal chordae. No additional lesions were objectified.

Conclusions: Noninvasive ultrasound histotripsy succeed to cut mitral valve basal chordae in vitro and in vivo in beating heart. Future investigations would be needed to test this noninvasive technique on others applications, such atrioseptostomy or valvular stenosis. If positives, this will open the door of completely noninvasive therapeutic technique for these situations.

P-28

Cardiac mitochondrial ultrastructure and respiratory chain enzymatic activities in patients with pediatric hypertrophic cardiomyopathy

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Introduction: Hypertrophic cardiomyopathy (HCM) is known to have impaired energy metabolism due to inappropriate myocardial energy demand or decreased mitochondrial ATP production. We speculate that cardiac mitochondrial homeostasis is one of the significant factors affecting prognosis in patients with HCM. Here, we studied cardiac mitochondrial ultrastructure and respiratory chain enzymatic (RC) activities in patients with HCM.

Methods: Five patients with HCM (or HCM/RCM overlap) (Aged 10–28 y/o) were studied retrospectively. Genetic study showed sarcomeric gene mutation (TNNT3, MYL2) in 3 cases and mt.DNA mutation (mt.3243 A > G) in 2 cases. Myocardial samples were obtained by right ventricular endomyocardial biopsy (EMB). Mitochondrial ultrastructure and RC activities in EMB samples were studied by transmission electron microscopy (TEM) and RC assay, respectively. One case (mt.3243 A > G) was lost due to heart failure two years after EMB at 12 years old, and another case (TNNT3) was lost suddenly 10 months after EMB at 11 years old.

Results: TEM revealed increased number of mitochondria in all patients. In patients with mt.3243 A > G, mitochondria appeared quite irregular in size and contour with aberrant cristae. RC assay revealed decreased activity of complex I and I + IV, respectively. In patients with sarcomeric gene mutation, mitochondria appear

almost regular in size with lamellar cristae, however some swollen mitochondria with a paucity of cristae were found. Notably, A case of sudden death with TNNI3 mutation had increased number of swollen mitochondria and decreased activity of complex IV, while others revealed almost normal lamellar cristae in EM and normal RC activity.

Conclusions: Mitochondrial evaluation in patients with HCM using EMB may be a critical role in not only diagnosis of primary mitochondrial cardiomyopathy, but assessment of secondary mitochondrial dysfunction in HCM with sarcomeric gene mutation.

P-29

Changes of blood pressure, visceral fat tissue and gene in fetal programming induced rat model after Cozaar XQ treatment

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Purpose: The purposes of this study were to investigate the effects of fetal under-nutrition during pregnancy and lactation on blood pressure, visceral fat, and several genes and evaluate changes after Cozaar XQ treatment.

Methods: Pregnant rats were studied from the 10th day to term gestation and through lactation. The rats were divided into three groups: the control (C) group and study group. The study rats were put on a 50% food restricted diet (FR). The study rats were further divided into the FR group and Cozaar XQ (CX) group, which was treated with Cozaar XQ after birth. Serum lipid profiles and glucose were determined in offspring at 1, 2, 3, 4 and 5 months. Masson's Trichrome staining was performed in order to observe the degree of fibrosis in the heart tissues. The amount of abdominal visceral fat was measured in the offspring. Microarray analysis was performed in visceral fat tissues. Western blot analysis regarding five genes such as endothelin (ET)-1, ACE (angiotensin converting enzyme), angiotensin II receptor type IA (ATIA), troponin (Tn) I and endothelial nitric oxide synthase (eNOS) were performed.

Results: Blood pressure and body weight were significantly higher in FR group in the 5 month old FR group compared with the C group and significantly lower in CX group compared with the FR group. The amount of visceral fat was significantly higher in the 2, 4 and 5 month old FR group compared with the C group and significantly lower in CX group compared with the FR group. Fat acid synthase, leptin, ET, angiotensinogen, ET receptor A, adiponectin, carnitine palmitoyltransferase-1 and insulin-like growth factor binding protein-3 genes were significantly expressed in visceral fat by microarray analysis. Expressions of ACE, ATIA, Tn and eNOS proteins in the CX group were lower than FR group at 5 month.

Conclusion: Fetal under-nutrition only during pregnancy resulted with obesity, high blood pressure and gene changes in offspring. Cozaar XQ treatment prevented obesity, hypertension and gene changes.

P-30

Neonatal cardiomyopathy caused by double mutation in RAS pathway genes

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Introduction: Severe myocardial hypertrophy in children, and especially, in infants, needs more extensive diagnostic work up due

to the wide spectrum of inborn errors of metabolism, monogenic syndromes and microchromosomal aberrations. Pediatric patients with hypertrophic cardiomyopathy diagnosed in the first six months of life represent a group with the poorest prognosis and very limited treatment options. Every such case needs the personalized approach in terms of arrhythmia management, surgical interventions and genetic counseling.

Results: A 5-month boy was hospitalized due to obstructive hypertrophic cardiomyopathy diagnosed right after birth. He was born in term with Cesarean cessation due to the mother's age. Myocardial hypertrophy was revealed on week 28 of gestation during scheduled ultrasound. The patient had no signs of dysmorphism, lactate level and other biochemical parameters were normal. The inborn metabolic disorders were excluded by mass spectrometry metabolite tests and standard cytogenetic analysis confirmed 46XY karyotype. Serial echocardiography performed during the first year demonstrated severe increase in myocardial thickness with critical left ventricular outflow tract obstruction that gradually deteriorated with time. Morphological examination of the myocardial sample obtained during Ross-Cohn operation revealed gross myocardial fibrosis and myofibrillar disarray. To identify a genetic cause of severe inborn hypertrophic cardiomyopathy we performed a target sequencing of 108 genes associated with cardiomyopathies, arrhythmic disorders and several metabolic disorders using NGS approach on MiSeq instrument after Haloplex target enrichment. Two mutations in the genes of RAS signaling pathway were detected. The first one, Y279C in PTPN11 was previously reported in a patient with LEOPARD syndrome. The second one is a new R585L substitution in CBL gene, which is associated Noonan-like syndrome. Both genes participate in the intracellular RAS signaling pathway that contributes to the cell proliferation and hypertrophy in response to extracellular growth factor stimulation. Mutations in the RAS-pathways genes lead to the group of clinically overlapping disorder called "RASopathies" which often involve myocardial hypertrophy, congenital heart disorders, ectodermal abnormalities and tumors. To our knowledge this is the first report of combined double mutations in the genes of RAS-pathway in a patient with hypertrophic cardiomyopathy.

Conclusions: We conclude, that severe neonatal myocardial hypertrophy with left ventricular tract obstruction may be caused by double mutations in the genes of RAS-pathway in spite of absence of facial dysmorphism and other organ abnormalities.

P-31

Regional cerebral oxygen saturation (rScO₂) in a cardiac muscarinic receptor overexpression rabbit model

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Introduction: Near infrared spectroscopy (NIRS) can be used as a noninvasive monitoring technique for regional cerebral oxygenation (rScO₂).

We studied basal rScO₂, after atropin blockade, and after hypertensive test by phenylephrin in a cardiac muscarinic receptor overexpression rabbit strain (H) with severe cardiac pauses compared to a normal rabbits strain (N).

Results: 1) rScO₂ values are systematically higher in H rabbits compared to N rabbits:

(H = 75.6% ± 5.6 vs N = 60.7% ± 5.7, n=6 in both groups, p < 0.05).

2) Atropin decrease rScO₂ in both groups, but this reduction is more marked in H rabbits.

3) During the phenylephrin test, the cardiac pauses in H group are longer than in N group (H: 24 285 ms ± 8 837 (n = 6) vs N: 3 566 ms ± 1 455 (n = 8)).

We observed a progressive hypoxia with rScO₂ decrease in both groups during the test. Unexpectedly, the rScO₂ reduction is less in H rabbits with severe cardiac pauses than in N group.

Conclusion: These data support the hypothesis that:

A higher rScO₂ by NIRS could reflect a higher central muscarinic receptor density, protecting brain against hypoxia.

RScO₂ could be a non invasive muscarinic receptor over-expression marker, useful in vasovagal syncope study.

P-32

Evaluation of heart function In children with intestinal failure on long-term parenteral nutrition

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Objectives: Children with intestinal failure receiving long-term parenteral nutrition have several risk factors potentially deteriorating heart function, such as volume overload, the presence of permanent central catheter with the tip in right atrium, recurrent anemia, carnitine deficiency, and often, catheter-related sepsis in medical history.

Methods: The aim of this cross-sectional study was to evaluate heart function in children on long-term parenteral nutrition with the use of biochemical parameter – NTproBNP value, echocardiographic cardiac function and anatomy parameters: SF, E/A wave ratio, LVEDD; LVESD; IVSd; IVSs, LVPWd, LVPWs; calculated LV mass, valves 2D measurement; and cardiac-thoracic index assessed on a chest X-ray. Results were compared with normal values determined by epidemiological studies with respect to body surface area [Heart 2000;83:667–672] and age [Pediatr Cardiol (2009);30:3–8]. Sixty seven children with intestinal failure aged 1–19 years (median:5.84 years) received parenteral nutrition for 9 months to 14,75 years (median: 3 years) before the study.

Results: Mean percentiles of echocardiographic parameters were respectively: LVEDD – 46th, LVESD–46th, IVSd–70th, IVSs–45th, LVPWd – 49th; LVPWs–34th, LV mass calculated on the basis of M-mode parameters – 70th, mitral valve – 86th, tricuspid valve – 29th, aortic valve – 86th, pulmonary valve–73th. Shortening fraction was within normal values (28–44%) in almost all of the patients (range: 28,6–49,5%; mean 37%), except for 5 children, in which the SF was augmented over 44%, the tricuspid valve flow pattern expressed as E/A waves ratio was normal in most of the patients (range 0,65–2,35; mean: 1,5) except for 3 children who had E/A ratio <1; E/A waves ratio for mitral valve was normal in all of the patients (range: 1,2–2,8; mean: 1,66).

46 of 67 children had significantly (over 95th percentile) augmented NTproBNP level, 25 patients (of 59 in which chest X-ray was performed) had an elevated (over 0,5) CTI.

Conclusions: Systolic and diastolic heart function assessed by shortening fraction and E/A wave ratio were in the normal values. IVSd, aortic, mitral and pulmonary valves measurements were slightly augmented (70–86th percentile). Cardiac-thoracic index and NTproBNP values were elevated in most of the patients. Long-term parenteral nutrition and/or intestinal failure might influence cardiac function and anatomy.

P-33

Pharmacogenetic approach in treatment of arterial hypertension in children

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Background: Hypertension affects up to 5% of all children and the incidence of pediatric hypertension is increasing, mainly due to an increase in primary (essential) hypertension. Evidence suggests that blood pressure response and outcomes associated with anti-hypertensive drugs are influenced by genetic variation. Most of medications used for adults can be also used for children. However, efficacy data is limited for the pediatric population.

Objective: To analyze the efficacy of hypertension treatment approach in children, based on evaluation of specific genetic polymorphisms responsible for hypertension.

Methods: The study subjects consisted of 54 adolescent patients with essential hypertension [mean age 13.2 ± 2.9 years]. We identified the following genetic variants of ADD1 1378 G > T, AGT 704 T > C (Met235Thr), AGT 521 C > T (Thr174Met), AGTR1 1166 A > C, AGTR2 1675 G > A, CYP11B2 344 C > T, GNB3 825 C > T, NOS3 786 T > C, NOS3 894 G > T (Glu298Asp) in all subjects. Gene DNA was extracted from blood samples and amplified by polymerase chain reaction (PCR). We divided all patients into two groups. First group consisted of 26 children who received treatment based on their genetic profiles. Subjects from the second group (29 subjects) have been treated in strict accordance with the fourth report on the diagnosis, evaluation and treatment of high blood pressure in children and adolescents without taking genetic profile into consideration.

Results: All children had more than five genetic polymorphisms of genes, associated with arterial hypertension in the homozygous or heterozygous state. In our study genetic-based treatment was statistically more effective than traditional. As a result of treatment systolic blood pressure lowered more significantly in first group (14,4 mmHg) vs second group of patients (6,1 mmHg), p < 0,05.

Conclusions: Our study has shown that hypertension treatment based on evaluation of individual's genetic profile is more effective than traditional. The study suggests that current management strategies for treatment of essential arterial hypertension in children need to be changed, and pharmacogenetics may be a tool to help achieve a goal.

P-34

Carvajal syndrome – rare entity of cardiocutaneous syndrome in a child from the Mediterranean part of Croatia – report on a new mutation of a desmoplakin gene

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Introduction: Within the arrhythmogenic right ventricular cardiomyopathy (ARVCM) there are two entity forms referred to as the Naxos syndrome and the Carvajal syndrome. If ARVCM also has a palmoplantar keratoderma with distinctive hair features (wooly hair) it is described as the Naxos syndrome, but if cardiomyopathy is spread on both ventricles with even more severe changes on the left ventricle, the entity is referred to as the Carvajal syndrome.

Objective: To describe a child with a biventricular dilated cardiomyopathy with a dominantly left sided cardiomyopathy with

palmoplantar hyperkeratosis and wooly hair, and to present a molecular-genetic analysis of a gene which codes desmoplakin; one known database mutation as a non sense mutation was found: c.3791 G > T, p.Glu1265X on exon 23 and one variant not yet presented in the database (<http://www.arvcdatabase.info>): variant p.Asp297His, c.889 G > C on exon 7.

Case study: A 4.5 years old boy from the Croatian Mediterranean basin was hospitalized for clinical symptoms of global biventricular cardiac insufficiency (hepatomegalia, positive venal pulse, pulmonary edema), with extreme cardiomegalia and high pro-BNP values. He had palmoplantar hyperkeratosis and peculiar hair (wooly hair), and echocardiography revealed biventricular dilatation with severe LV hypocontractility. After a short-term stabilization using conventional therapy (diuretics, ACE inhibitors, digitalis), he became dependent on inotropic support. He was candidate for heart transplant, but died before available donor. Child's mother also has dilated cardiomyopathy which extends on both ventricles with decreased contractility (EF 35%). Due to arrhythmogenic episodes which commenced when she was 40, an ICD was implanted in the mother. A molecular-genetic analysis was performed for the child. We also presented a pathological anatomical finding with special emphasis on presenting the histological analysis of right and left ventricle.

Conclusion: We present a patient with a triad of symptoms (biventricular cardiomyopathy, palmoplantar hyperkeratosis, wooly hair) making the Carvajal syndrome which is a rare entity form of the Naxos syndrome, i.e. arrhythmogenic cardiomyopathy of the right ventricle. Detailed molecular-genetic analysis has confirmed a new mutation for desmoplakin synthesis on exon 23 and a peculiar pathohistological finding on right and left ventricle.

P-35

Wound Infections in Children with Ventricular Assist Device Support

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Introduction: Children with end-stage heart failure often require circulatory support with a ventricular assist device (VAD). Wound infections may lead to significant morbidity and mortality. The incidence and outcome of wound infections is still unknown in children supported on VADs. We aimed to describe the predictors, incidence and outcomes of early and late wound infections in pediatric VAD recipients.

Methods: A retrospective review of all patients from 2005–2014 aged <18 years supported on VADs was completed. VAD-related infection was defined using INTERMACS definition and wound infection was further categorized using Center of Disease Control (CDC) classification as superficial or deep, and early (<30 days) or late (>30 days post implant).

Results: 70 children (28 female) underwent implantation of VADs (58 Berlin Heart®, 12 HeartWare®) of which 12 had biventricular support. Diagnosis included cardiomyopathy/myocarditis (n = 55) and congenital heart disease (CHD) (n = 15). Wound infections occurred in 20 patients (28.6%); 3 deep infections and 17 superficial infections. Late infections occurred in 18 of 20 patients; with positive wound cultures identifiable in 16, and 5 of these patients with blood cultures matching wound culture. Mortality in patients with wound infections was 25% (5 patients) and was not different to overall mortality of 27%. Type of device (intracorporeal or

extracorporeal), BiVAD, LVAD and diagnosis were not predictors of late wound infections. Both patients with early wound infections had CHD and were supported with extracorporeal LVADs. These 2 children died within 2 weeks of the wound infection.

Conclusion: Wound infections occurred in 29% of children supported on VADs. Early wound infections are infrequent, but resulted in death of the children. Type of device, age, and diagnosis were not risk factors for the development of wound infections.

P-36

Neurodevelopmental outcome, behaviour difficulties and quality of life in School children after heart transplantation in early childhood

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Objectives: The aim of this study was to examine neurodevelopment, behaviour difficulties and health-related quality of life (HRQoL) in school aged children after heart transplantation during infancy/early childhood.

Methods: 28 children (13 females, 15 males) with CHD after HTx were examined at a median age of 9 years 2 months (range 5 y 8 mo–11 y 9 mo). Outcome was assessed with standardised test of intelligence (CFT 1), selective testing of attention (d2), neurological and fine motor testing. Parental questionnaire (CBCL 4-18, Short-form health survey) were used to measure behaviour difficulties and health related quality of life (HRQoL). Results were compared with those of 28 age- and sex-matched healthy comparison individuals.

Results: Outcome with regard to full-scale IQ was poorer in patients after HTx than in the comparison group ($p < .001$). Patients with CHD also had lower scores on all motor domains ($p < .001$). In the attention test the group after HTx scored also poorer than the healthy comparison group ($p < .01$). Parental estimation revealed also a higher rate of behavioural problems ($p < .01$). Quality of life was similar to that of typically developing peers.

Conclusions: Children after HTx have persistent cognitive and motor impairments, while parental-reported HRQoL are mostly described as normal. Consequently long-term neurodevelopmental evaluations/follow up are necessary to provide early educational and therapeutic support.

P-37

Egr-1 identifies neointimal remodeling and relates to disease progression in human pulmonary arterial hypertension

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Introduction: Pulmonary arterial hypertension (PAH) is hallmarked by the development of neointimal lesions. The transcription factor

Egr-1 plays a critical role in neointimal formation in experimental PAH. This study investigated whether Egr-1 is also associated with neointimal-type vascular remodeling in different forms of human PAH.

Methods: Using immunohistochemistry, we studied pulmonary vascular Egr-1 expression in lung tissue of 58 PAH patients with either advanced (PAH-CHD, 12 explants, 9 biopsies; iPAH, 18 explants), or early stage disease (PAH-CHD, n = 19 biopsies), and compared with non-neointimal PH (hypoxic PH; n = 4) and controls (n = 10).

Results: Pulmonary vascular expression of Egr-1 was increased in PAH compared to non-neointimal (hypoxic) PH and controls, and expression was more abundant in PAH-CHD than in iPAH. Egr-1 expression was more pronounced in advanced PAH-CHD compared to early disease stages.

Conclusions: Pulmonary vascular Egr-1 expression is increased in patients with PAH and is specifically associated with neointimal-type vascular remodeling. These data add to the accumulating evidence that Egr-1 plays a critical role in the development of neointimal lesions in PAH and identifies it as a putative target for PAH therapy. In addition, the level of pulmonary vascular Egr-1 expression may be supportive in assessing disease stage in patients with PAH-CHD.

P-38

Biventricular versus monventricular mechanical circulatory assistance in children

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Experience in Pediatric Mechanical Circulatory Support has shown that prognosis is worse in biventricular (biVAD) than LV (LVAD) support.

Objectives: To assess outcomes of children who had biVAD compared to LVAD in a single center experience.

Material and Methods: From 2005 to 2014, 27 patients aged <18 years, were implanted with Ventricular Assist Device. Their clinical and biological data, echocardiographic records and outcomes were reviewed. Patients were divided in biVAD (21 cases) and LVAD (6 cases) groups.

Results: Thirteen females and 14 males, aged 4 ± 4.8 years: 0.29 to 16.1 years (median 1.4) received a left ventricular (n=6) or biventricular (n=21) support, for cardiogenic shock or cardiac arrest in 8, or uncontrolled low cardiac output in 19, due to cardiomyopathy (24), acute myocarditis (2) or ischemic cardiomyopathy (1). Age at VAD was lower in LVAD than BiVAD (median: 0.9 vs 2.5 y). Duration of support was 46 ± 44 days: 3 to 182 days (median 35: 31 days in biVAD, 65 days in LVAD). Four patients less than 1 year, experienced a severe stroke (14%), 6 died on support (22%), of whom 5 were <1 year, 3 were weaned off support (11%), 18 were transplanted (67%). Mortality was 19% in biVAD vs 33% in LVAD. Survival to support was 78%. Death decreased from 28.5% in 2005-09 to 15.4% in 2010-14 ($p=0.22$), significant stroke from 28.5% to none ($p=0.05$) and infections from 71.4% to 30.8% ($p=0.02$). Significant stroke on VAD was more frequent in LVAD (33%) than in biVAD (9%), $p=0.1$. Infections rate was 33% in LVAD compared to 57% in biVAD ($p=0.5$).

Conclusion: Survival to mechanical circulatory support in children increased over time despite biVAD still being the most frequently used mode. Stroke and mortality was lower in biVAD than LVAD groups.

P-39

Chronological change of left ventricular systolic function in children with fulminant myocarditis supported on ECMO

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Introduction: Aggressive hemodynamic support including extracorporeal membranous oxygenation (ECMO) would be beneficial for patients with fulminant myocarditis in the acute phase, serving as a bridge to the recovery of LV function. However, few studies reported the optimal duration of ECMO support for children with fulminant myocarditis. We aimed to describe the time course of LV ejection fraction (EF) recovery in children with fulminant myocarditis supported on ECMO.

Methods: We identified 15 children supported on ECMO out of 21 children with fulminant myocarditis admitted to the pediatric intensive care unit at our institution from December 2008 to December 2014. Of these, 13 patients were successfully weaned from ECMO support without further treatment with ventricular assist device implantation or heart transplantation, and included in this study. The medical records of the patients were retrospectively reviewed.

Results: The median age of the patients was 5 years (range, 4 days-11 years), and the median duration of ECMO support was 8 days (range, 5-14 days). Indication for ECMO support was severe LV systolic dysfunction in 10 patients (group A, LVEF $28 \pm 13\%$), and 3 patients required ECMO support because of uncontrollable arrhythmia despite the preserved LVEF prior to ECMO introduction (group B, LVEF $47 \pm 8\%$). Overall, LVEF was $32 \pm 15\%$ prior to ECMO introduction, and decreased drastically after the initiation of ECMO support. The recovery of LVEF started 4.5 ± 1.5 days (range, 1-7 days) after the ECMO introduction. The patients restored LVEF of 60% 14 ± 9 days (range, 5-38 days) after the ECMO introduction. Children in group A showed later recovery of LVEF (16 vs 9 days) and longer duration of ECMO support (9 vs 6 days) than those in group B, although the differences did not reach statistical significance.

Conclusions: In children with fulminant myocarditis successfully weaned from ECMO, LV systolic function started to improve within a week after ECMO initiation and was restored to normal mostly in a month. Further treatment with ventricular assist device implantation or heart transplantation should be considered in patients without an evidence of LVEF recovery after 1 week's ECMO support.

P-40

Heart transplantation in children in Croatia – our first experiences

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Introduction: According to the ISHLT register between 350 and 400 pediatric heart transplantations (PHT) are performed worldwide each year (10% of all). Basic criteria for PHT are: stage D cardiac insufficiency (need for continuous use of inotropes, mechanical ventilation), stage C (abnormal cardiac structure or function with inherent symptoms of cardiac insufficiency) with reactive pulmonary hypertension (PH); PVR <6 Wood Units

(WU); TPG <15 mmHg. There are three main indication areas for PHT: 1. Cardiomyopathies, 2. Congenital heart defects (with no possibility of anatomical correction, high operative fatality, and after palliation; e.g. failing Fontan), 3. Retransplantation.

Objective: Presenting first experiences with PHT in Croatia.

Results: The first (adult) HT in Croatia (Zagreb) was performed in 1988, heart transplantation in children are performed since 2011. Indications for PHT had been set for 8 children. All had cardiac insufficiency due to cardiomyopathies. Four of the transplanted patients had a dilated cardiomyopathy as a result of earlier myocarditis and two had restrictive cardiomyopathy (RCMP) of undetermined etiopathogenesis. Two patients age 6 with DCMP (one had Carvajal CMP) died before HT. Mean age was 10 (4–12 years). All patients with DCMP had stage D heart failure and PVR below 6 WU. Two with RCMP had stage C (NYHA III) with PVR around 6 WU. Average time between making the diagnosis and need for HT was 18 months. Early post transplantation complications were: diastolic dysfunction of left ventricle with PH needing ECMO and one patient died due to septic shock and right ventricle failure. Late complications included chronic renal insufficiency (stage III), transitory PH, bradycardia needing permanent pacemaker and transitory iatrogenic Cushing syndrome. Immunosuppression is performed with corticosteroids, tacrolimus and mycophenolate mofetil. Myocard biopsy for rejection analysis is performed 1, 2, 3, 6, 12 months and yearly after. Time of monitoring is 6 months to 3 years, status of all five patients is NYHA I.

Conclusion: Pediatric heart transplantation, when all the transplantation criteria are strictly adhered to, saves lives, improves the quality of life and allows normal physical activity and a normal “schoolchild” life.

P-42

Echogenic Focus in Fetal Heart; Is it still a dilemma?

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Objectives: The significance of fetal intracardiac echogenic foci (IEF) still remains uncertain. The aim of our study was to investigate the clinicopathologic significance of IEF by detailed fetal echocardiography (DFE).

Methods: We performed 760 DFE (including 2D, M – Mode, Doppler and Tissue Doppler imaging - heart size, ventricular diameters and ratios, ventricular wall thicknesses, pulmonary and aortic diameters; peak velocities, mitral and tricuspid E/A ratios; left ventricular fractional shortening and myocardial performance indexes-) during 18 and 34 weeks gestational aged pregnant between January 2013 and November 2014. Fetal heart structures and ventricular functions (using both conventional and doppler techniques) were evaluated. A questionnaire were taken from the patients including general health and obstetric history.

Results: The number of fetuses have single IEF in left ventricle were 206; right ventricle 72 and in both ventricle were 48. The overall incidence was 42% in our study. When compared fetuses without IEF, only the multiparas showed significant difference (n=161, 49,3%, p<0.05). No significant differences were found in fetal cardiac dimensions, Doppler indices and left ventricular fractional shortening and myocardial performance indexes of both ventricles of fetuses who have IEF in left, right or both ventricle when compared with fetuses without IEF. Congenital cardiac malformations and chromosomal anomalies like trisomy 21 were same (p>0.05 for all).

Conclusions: Isolated IEF in fetal heart; even multiple and in both ventricles has no association between fetal cardiac structural and functional abnormality and thus no need for further evaluation for this benign situation.

P-43

Incidence of Congenital Cardiac Disease, detected by Fetal Echocardiography, in Azerbaijan Experience of three centers during a thirty month period

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Aim: To present the incidence of Congenital Heart disease (CHD), detected by fetal echocardiography. This is the experience from 3 centers during a period of 2.5 years in Baku, Azerbaijan.

Population-Method: Between July 2012 and December 2014 we have initially studied in 3 centers, 372 fetuses that were referred with different indications. Most common were:

1. Inbreeding (first and second degree relatives among parents) (29.17%) 2. Abnormal cardiac screening examination (24.72%) 3. IVF conceived fetuses, (14.86%) 4. Abnormal heart rate or rhythm (9.50%) 5. First-degree relative of a fetus with congenital heart disease (6.81%), 6. Combination of maternal age and increased nuchal translucency (6.40%) 7. Maternal Metabolic disorder (Diabetes Mellitus)(4.71%) 8. Others (1.63%). The initial scans were done mostly after the 21st week of gestation, with a mean gestational age (23 + /-5 weeks). The assessment was done using color Doppler – 2D echocardiography, by a General Electric Vivid 7 device, with an appropriate cardiac fetal probe and software program. The findings were verified with echocardiography an MRI studies were indicated, post-natal, and postmortem examination in pregnancies that were interrupted.

Results: We detected 72 (CHD), (19.35%).

Complex (CDH) were: Complete AV channels (8/11.11%), Tetralogy of Fallot (7/9.72%), Double Outlet RV (6/8.33%), Tricuspid valve atresia (6/8,33%), combination of a (pm)VSD with Aortic arch hypoplasia (3/4.17% and or coactation (5/6.95%), hypoplastic LV Syndrome (4/5.55%), (4/5.55%) of complex VSD's, Simple TGA's (4/5.55%), (1/1.39%) congenitally corrected TGA with additional VSD and Ebstein type Tricuspid valve defect, (1/1.39%) Ebstein Anomaly and (1/1.39%) Truncus Arteriosus with a criss-cross, heart. The commonest simple defects were VSD's (15/20.83% mostly muscular (11/15.27 and pulmonary (5/6.95%) aortic valve stenosis (2/2.78%). Additionally, detected (4/5.48%) Fetal Arrhythmias from which 2 were transitional and (1/1.37%) permanent (complete A–V block).

Conclusion: The incidence of detected (CHD) is high, even without including 1st trimester screening. Of all detected (CHD), 21 (29.17%) were products inbreeding, by first degree related parents.

P-44

The abnormal fetal growth pattern in fetuses with different anatomic and haemodynamic profile of heart defects on the base of analysis the selected anthropometrical parameters

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Background: the publications show that peripheral especially cerebral blood flow may impact brain development and pattern of

head and body growth in fetuses with heart defects. This possibility is supported by identification of neuropathological abnormalities in neonates with heart anomalies prior to surgery.

Aim: Definition of the connection between haemodynamical types of heart defects and anomalies of anthropometric parameters in the fetus and the newborn.

Material: 185 fetuses diagnosed at the Prenatal Cardiology Departments in Lodz and in Cracow were analysed in the three groups:

1. R; n = 67 - with "reversal" flow in ascending aorta (R1; n = 58: hypoplastic left heart syndrome, HLHS, R2; n = 9: critical aortic stenosis, SA)
2. A; n = 54 - with "antegrade" flow to the ascending aorta (A1; n = 42 mixed defects, A2; n = 12 transposition of great arteries, TGA)
3. N-control group; n = 50.

Methods: The two time-points data were analysed:

Prenatal period - estimated fetal weight (EFW), head circumference (HC) biparietal diameter (BPD), femoral length (FL), abdominal circumference (AC), HC/EFW and HC/AC indexes. Postnatal period - birth weight (BW), birth length (BL), head and thorax circumference (HC, TC) HC/TC and HC/BW indexes.

Data were standardized to make results independent from the gestational age using covariance analysis and of the Z-score indexes for EFW and HC. The LSD multiple comparison and post-hoc GT2 Hochberg analysis were used. Fetal parameters: there were no significant differences in the main groups. Higher parameters of AC were observed in the fetuses with TGA (P = .032) and lower of HC values in the R group (borderline significance: P = .055).

Neonatal parameters: BL was the lowest in the fetuses with HLHS and SA (P = .004) and the highest in fetuses with TGA (P = .001), BW was lowest in the SA subgroup, (P = .021); HC/AC indexes were lowest in the newborns with TGA, and lower in the newborns with HLHS compared with SA subgroup (P = .008).

Conclusions: The abnormal growth pattern probably due to abnormal model of aortic flow may concern fetuses and newborn with some heart defects. The mechanisms, physiologic implications and neurodevelopmental impact of abnormal cerebral flow in fetuses with CHD require further study.

P-45

Ductal and Aortic Arch Anomalies Detected in Fetal Life by Echocardiography

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Introduction: Ductal and aortic arch anomalies can be diagnosed by echocardiography and computed tomography in neonates. However prenatal diagnosis is rarely reported. Here we present our experience in detecting ductal and aortic arch anomalies by fetal echocardiography.

Methods: The records of patients with ductal and aortic arch anomalies from 2007 to 2014 were evaluated retrospectively. Autopsy findings, postnatal evaluation and karyotype analyses were enrolled in the study.

Results: In 8 patient right arcus aorta + left ductus arteriosus + aberrant left subclavian artery; in 4 patients right arcus aorta; in 1 patient left arcus aorta with abnormal left subclavian artery; in 1 patient bilateral ductus + right arcus aorta + aberrant left subclavian artery; in 2 patients ductal aneurysm and in 1 patient double arcus aorta were detected. Trizomi 18 was detected in one patient with bilateral ductus arteriosus. Di George syndrome was determined in one patient with right arcus aorta and Williams syndrome was detected in one patient with ductal aneurysm. Atrioventricular septal defect and ventricular septal defect were

accompanied right arcus aorta in two patients. Ventricular septal defect and isthmus hypoplasia were detected in one patient with ductal aneurysm. Postnatal echocardiographic evaluation demonstrated right arcus aorta in 7 patient without any symptom. Ductus arteriosus was shrunk and closed spontaneously in one baby with the diagnose of ductal aneurysm during postnatal period. The other baby with the diagnose of ductal aneurysm + Williams syndrome + ventricular septal defect + isthmus hypoplasia underwent surgery for ductus arteriosus closure and ventricular septal defect and coarctation repair. Intrauterine fetal death accrued in 2 patient and the diagnosis were verified by autopsy. Postnatal echocardiographic evaluation of one patient with aberrant left subclavian artery revealed left arcus aorta. Clinical follow up was considered due to no symptom was detected during the neonatal period. The baby with diagnose of double arcus aorta underwent surgery because of respiratory distress after delivery.

Conclusion: Ductus and arcus anomalies on fetal ultrasonography may indicate chromosomal abnormalities that may complicate postnatal management. When arcus anomalies are identified, fetal karyotype analysis is warranted.

P-46

Single Umbilical Artery: An Important Marker for Prenatal Suspicion and Detection of Fetal Cardiac Anomalies

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Objective: To determine the frequency of associations between the single umbilical artery (SUA) and congenital heart disease in two tertiary centers.

Methods: The fetuses diagnosed with SUA at mid-trimester detailed ultrasound examination between May 2001 and March 2014 were included in the study. Colour Doppler was used to visualize the umbilical arteries adjacent to the fetal bladder and in a section of the free loop of cord.

Results: A total of 265 fetuses with SUA were identified. Complete data were available in 197/265 pregnancies (74.3%). The mean maternal age was 29 years and the average gestational age at diagnosis was 23 weeks. A cardiac anomaly was detected in 58 of these fetuses (29.0%): 34.3% in center-1, and 18.3% in center-2. Detected cardiac abnormalities include 19 ventricular septal defect (14 perimembranous, five muscular), eight tetralogy of Fallot (TOF), seven complete atrioventricular septal defect, five hypoplastic left heart syndromes, five double outlet right ventricle, three coarctation of the aorta, three hypoplastic right heart syndrome, two dextrocardia, and one for each of the following: absent pulmonary valve, aorta-pulmonary window, left atrial isomerism, double aortic arch, aortic stenosis and transposition of the great arteries. The only chromosomal abnormality was trisomy 18 that was detected in eight cases. Among eight twin pregnancies with SUA, only two had cardiac anomaly, both exhibited TOF. Termination of pregnancies was performed in seven cases, and intrauterine death occurred in five cases.

Conclusions: Our study confirms that the frequency of cardiac anomalies is very high in fetuses with SUA. Albeit the incidence is variable from one center to the other, there is a pressing need to consider the finding of SUA among the main referral indications for a detailed fetal echocardiography.

P-47

Introducing Pulse Oximetry as a screen for Critical Congenital Heart Defects in the district general hospital setting

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Introduction: Critical congenital heart defects (CCHD's) are associated with significant mortality and morbidity. Early diagnosis and treatment could reduce this significantly. Currently there is a diagnostic gap in identifying CCHDs even after prenatal screening and postnatal examination. Pulse Oximetry as part of newborn examination is increasingly being used to minimise this gap. There is a growing body of evidence to suggest that it is a valid screening tool for identifying CCHDs, which is reflected by its uptake in many neonatal units. Our aim was to demonstrate the feasibility of implementing pulse oximetry screening in a busy DGH setting and outline the financial and practical considerations.

Methods: All newborn examinations performed by paediatricians in July/August 2014 included pulse oximetry screening. A portable pulse-oximeter with disposable wraps was used to measure pre-ductal and post-ductal saturations. Outcomes were divided into three groups. Those with both readings $\geq 95\%$ were deemed negative. Those with either reading between 90-94%, or $>2\%$ difference of saturations were deemed as a borderline positive test. These required re-measuring after 2 hours if the child remained asymptomatic. Three borderline positive tests, or a positive test (either saturation $<90\%$ or a symptomatic child) required comprehensive assessment and echocardiography where clinically indicated. Exclusion criteria were: Midwife performed newborn examinations, pre-term infants (<37 weeks gestation), infants admitted to the neonatal intensive care ward.

Results: 170 infants had their pre and post ductal saturations measured over a 1-month period. Average time to testing was 38.6 ± 27.5 hours. The average duration of the test was $3.98 (\pm 3.41)$ minutes. All tests done over the trial period were negative. There was no increased burden on the echocardiography workload or SCBU beds. Feedback from doctors, nurses and parents was positive.

Conclusion: Implementing pulse oximetry screening for CCHD is a simple and cost affective strategy, without significantly increasing the time taken for newborn examination. It has a reassuring effect on doctors performing newborn examination. As reported in other studies, delaying the screening until after 24 hours maximizes the specificity, which may explain all tests being negative. There was no increase in demand for echocardiograms or admissions onto the neonatal unit.

P-48

Congenital heart disease in fetuses conceived after assisted reproductive technology

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Background: In the last decade, assisted reproductive techniques (ART) are increasingly used in couples with infertility and there is a concern about outcomes of children conceived, both for the preterm delivery and low birth weight, and for a possibly increased risk of congenital anomalies. Non certain data are available regarding the prevalence of congenital heart disease (CHD), however, in consideration of this risk, the women pregnant after ART are often referred for fetal echocardiography, at least in certain areas.

The aim of this study was to analyse the prevalence of CHD in fetuses conceived after ART, referred to our center, specifically for evaluation of the fetal heart.

Method: observational prospective study regarding consecutive fetuses conceived after ART referred to our Center in the period Jan 2010- Dec 2014.

Population: Three hundred two women (aged 30-49 yrs, median 36) that became pregnant after ART underwent fetal echocardiography at median gestational age of 21 week's gestation: 171 pregnancies resulted after in vitro fertilization (IVF) and 131 after an intracytoplasmatic sperm injection (ICSI); 24 and 21 were twins, after each procedure respectively (one triplet). Cases with known chromosomal anomaly or major extracardiac anomalies were not considered, only a few cases presented minor anomalies (of the chord vessels or chorioid plexus cysts). In two cases suspicious cardiac features were reported at the 1st level scan.

Results: CHD was found in 16 pregnancies (4 twins of healthy co-twins); 9 cases were products of IVF and 7 of ICSI procedure. Prevalence of CHD in the whole population was $16/302 = 5.3\%$, in Fivet $9/171 = 5.2\%$ and in ICSI $7/131 = 5.3\%$. Cases with suspicious heart features at the 1st level scan - were found normal. CHD found after FIVET were 2 TGA, 1 PAtr. + VSD, 1 VSD.1 CoA + AS, 2 Ebstein/Non-Ebstein, 2 PS, after ICSI 3HLH, 1 AVSD, 1 PAtr. + VSD, 1 DORV, 1 TF.

Conclusions: The data of this observational study show an increased prevalence of CHD after both techniques of ART, with respect to the data in normal population and with no difference between two techniques. Obviously, small numbers of this series present a possible bias.

P-49

Outcomes of patients with fetal heterotaxy syndrome in the current era

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Background: Patients with heterotaxy syndrome (HS) have a wide range of outcomes depend on their associated anomalies. Previous reports had shown that patients with asplenia syndrome (AS) had worse outcome than polysplenia syndrome (PS) and live-born patients with AS were associated with survival rate of 21-60%. Considering progress in management and surgical procedure for these patients, we reassessed outcomes of patients with fetal HS in this study.

Methods: From October 2010 to December 2014, 22 consecutive patients were diagnosed HS in utero. We excluded a patient associated with congenital diaphragmatic hernia from this study. We obtained clinical data of these patients from clinical records and assessed.

Results: Fetal diagnoses were made at a median gestational age of 27 weeks (range 19-36). Of 21 patients, 11 (52%) were diagnosed with AS and 10 (48%) were diagnosed with PS. 2 patients with advanced atrioventricular block (AVB) were elected termination of pregnancy. All AS patients were live birth and had single-ventricle physiology. Of 11 AS patients, 8 (73%) were alive. Of 10 PS patients, 8 were live birth and 5 were considered to be candidates for biventricular repair. 4 (50%) patients were alive and 3 were waiting for biventricular repair. 4 patients (3 AS and 1 PS) were associated with total anomalous pulmonary venous connection and 2 were died soon after birth because of pulmonary venous obstruction (PVO). The other 2 were alive, but no patient had undergone Glenn or Fontan procedure. There were 4 PS patients with advanced AVB who had undergone pacemaker implantation shortly after birth. 2 were delivered prematurely at gestational ages of 28 and 35 weeks because of hydrops fetalis. Of these 4 patients, 3 were died because of pacing failure,

postoperatively, and sepsis after operation for congenital biliary atresia. There was no patient who suffered from severe bacterial infection except for an above-mentioned patient.

Conclusions: PVO and AVB remained to be solved problems for HS patients. Outcomes in fetal HS patients without these associated defects were acceptable.

P-50

Right ventricle involvement in foetal critical aortic stenosis with heart failure - autopsy findings following prenatal intervention

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Introduction: Foetal aortic stenosis came into focus since it started to be managed interventionally in utero. However, little is known about the developmental mechanisms underlying the spectrum of the disease. It can be stable throughout the foetal life, evolve to become the hypoplastic left heart or progress to the foetal heart failure. Some authors suggest the involvement of second heart field cells responsible for the formation of compact myocardium and fibrous framework of the cardiac musculature and postulate the secondary role of the aortic valve itself. The aim of the study was to assess the histological features of left and right ventricular wall in autopsy of hearts obtained from the foetuses after in utero aortic valvuloplasty and correlate the results with previously obtained echocardiographic data.

Methods: In years 2011–2014 37 foetal aortic valvuloplasties in 34 fetuses were performed at our institution. 3 fetuses died in utero, 3 in early neonatal period, 2 of them after premature delivery. The autopsy was performed in 3 cases. Samples of the right and left ventricular walls were taken and stained with hematoxylin and eosin, Van Gieson trichrome for connective tissue and orcein for elastic fibers. Foetal echocardiographic examinations of those patients before the foetal aortic valvuloplasty were reviewed.

Results: Left ventricular samples of all the cases presented the endocardial fibroelastosis, myocardial disarray and increased amount of interstitial connective tissue. In the right ventricular samples, myocardial disarray was present in all cases and increased amount of interstitial connective tissue in 2 of them. On echocardiography the foetuses presented signs of heart failure with severe impairment of left ventricular function. In all the cases there were signs of the right ventricular failure: shortened or monophasic inflow to the right ventricle and mild tricuspid regurgitation.

Conclusions: Both left and right ventricular myocardium presents microscopic structural abnormalities in foetal hearts with critical aortic stenosis and foetal heart failure. Post mortem findings are consistent with foetal echocardiographic features. The results support the role of formation of the compact myocardium in development of foetal critical aortic stenosis presenting with heart failure.

P-51

The efficacy and safety of Amiodarone treatment for fetal tachycardia

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Although there is a little literature, Amiodarone is recommended as third-line therapy for fetal tachyarrhythmia due to more toxicity profile than other antiarrhythmic drugs.

The goal of this study was to establish the effectiveness and safety of Amiodarone treatment.

We reviewed outcome of 107 fetuses treated for tachyarrhythmia between 2002 and 2014 in our institution. Basing on experience in neonates we prospectively introduced Amiodarone in 64 fetuses. Amiodarone was first choice in fetuses with hydrops and/or long VA time during tachycardia. In fetuses with short VA time Digoxin was first choice, if not effective after 7 days Amiodarone was added. Amiodarone was given orally in all but 3 mothers. Daily dose was between 900–1200 mg, if effective reduced and continue throughout gestation. The treatment was effective when sinus rhythm was restored. The TSH level were checked in mothers and in newborns.

There were 53 fetuses with supraventricular tachycardia (gr.I), 11 with atrial flutter (gr.II). In gr.I 21 fetuses had NIHF. The CVS was measured in 50. Before treatment 10 fetuses had score between 0–4, 26 between 5–7 and 14 between 8–10. It has improved to 1 with score 4, 9 between 5–7 and 40 between 8–10. Amiodarone as the only drug was used in 20 effective in 16 (80%), Amiodarone with Digoxin was used in 28, effective in 24 (85%). Combined therapy Amiodarone + Metoprolol -1, Amiodarone + Digoxin + Metoprolol-3, Amiodarone + Digoxin + Propafenone-1, were effective. In gr.II 2 had NIHF. The CVS was measured in 9 fetuses. Before treatment there were 1 fetus with score 4, 3 between 5–7 and 6 between 8–10. After treatment all fetuses improved CVS and had between 8–10. Amiodarone alone was used in 5 pts, effective in 4 (80%), Amiodarone with Digoxin in 5, effective in 1 (20%), Amiodarone + Digoxin + Propafenone in 1 and was not effective. For both group, the side effect of Amiodarone were seen in 11 (17%) mothers: venus thrombosis in 1, rash in 2, increased skin pigmentation 1, only elevated TSH level in 6, hypothyrosis in 1. In all but 4 newborn TSH level was normal. Remaining 4 (6%) newborns developed hypothyrosis and required short-term thyroid hormonal substitution.

Amiodarone is highly effective for fetal tacharrhythmias. The complication rate is low.

P-52

The Evaluation of Patients with Borderline Left Ventricle: Single Center Experience

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Introduction: Left heart hypoplasia is a spectrum of left heart structures varying from complete atresia to mild hypoplasia in which the favorable end of the spectrum, namely borderline left ventricle (BLV) may progress to undergo biventricular repair. We conducted a review of data of patients on follow up with the initial diagnosis of BLV.

Methods: The records of 15 patients with the initial diagnosis of BLV were analysed retrospectively. Age, gender, body weight, initial and follow up echocardiographic measurements and additional cardiac abnormalities, operative and interventional history and outcomes were recorded.

Results: The median age at diagnosis was 7 days (1–80 days). Cardiac abnormality was diagnosed prenatally in 3 of the 15

patients (20%). Twelve patients were operated, remaining 3 have still been on follow up.

Echocardiographic analysis: Left atrial isomerism was detected in a patient and 5 patients had persistent left superior caval vein with large coronary sinus. EFE was not demonstrated in any of our patients. Median mitral valve annulus z scores and aortic annulus z scores were statistically insignificant in between patient groups of non operated, single ventricle palliation and biventricular correction performed.

Surgical procedures and outcome: 12 patients were operated at the median age of 38 days (7–120). Biventricular correction could not be performed in 3 patients, and they were directed to single ventricle palliation. Nine patients had pulmonary banding or coarctation repair or arch reconstruction together with additional procedures (VSD closure, pulmonary banding, coarctation repair etc) depending on their initial additional diagnosis. Four patients died postoperatively. Three of them were patients in whom single ventricle palliation was performed and the last patient was the one with LVOT and RVOT obstruction. Postprocedural last ECHO was performed at a median age of 16 months (3–46). There were no statistically significant difference of echocardiographic measurements between patients not operated and biventricular correction performed.

Conclusion: The choice between single or biventricular management pathway is difficult in infants with BLV and very close clinical and echocardiographic monitoring seems to be required. Early orientation to single ventricle palliation is also important for desirable outcome.

P-53

Assessment of arterial functions and cardiovascular risk in long-term follow-up of Kawasaki Disease

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Objective: Kawasaki Disease (KD) is a vasculitis involving coronary arteries. It is not clear if the patients with Kawasaki disease have an increased risk for atherosclerotic heart disease in adulthood. Carotid intima media thickness (CIMT), arterial distensibility an elasticity, flow mediated dilatation (FMD) of brachial artery and serum levels of high sensitivity C-reactive protein (hsCRP) are indicators of atherosclerotic heart disease. We searched for atherosclerosis risk in our patients with prior diagnosis of KD.

Material and Method: Study group consisted of 25 patients with prior KD, and control group consisted of 25 healthy children. Left ventricular systolic and diastolic functions; aortic anulus, sinus valsalva, sinotubular junction, asendan aorta, arcus aorta, isthmus and abdominal aorta diameters in systole and diastole were measured by M-mode and 2-D echocardiography. Aortic strain, distensibility and stiffness were calculated. CIMT and FMD of brachial artery were obtained using a linear transducer. Blood samples were taken for hsCRP levels.

Results: Study group included 4 girls, and 21 boys, control group included 4 girls and 21 boys. Age of the patients ranged between 4–19 years (Mean \pm SD = $8,32 \pm 3,7$ years) and control group between 4–16 years (Mean \pm SD = $9,1 \pm 3,5$ years). Follow-up period of study group was $3,48 \pm 2,47$ years, 10 patients (40%) had cardiac involvement during acute phase. There was no nt

difference in terms of left ventricular M-mode and two-dimensional measurements or systolic and diastolic functions between the groups. Aortic stiffness index was found to be higher (Mean \pm SD = $0,346 \pm 0,20$, Mean \pm SD = $0,196 \pm 0,10$; $p = 0,001$) and FMD of brachial artery at 3rd minute was lower in patients with prior KD comparing the healthy children (Mean \pm SD = $7,08 \pm 3,86$, Mean \pm SD = $10,40 \pm 4,00$; $p = 0,006$ respectively). HsCRP levels were higher, aortic strain and aortic distensibility were lower, CIMT were higher in the patients, however these differences were not statistically significant. ($p = 0,40$, $p = 0,143$, $p = 0,260$, $p = 0,726$ respectively).

Conclusion: Higher aortic stiffness and lower flow mediated dilatation of brachial artery suggests impaired arterial functions and an increased risk for atherosclerotic coronary artery disease following KD. Further investigation about long term results, close monitorization during adult age and elimination of other preventable risk factors are needed in this patient group.

P-54

ECG and Holter findings of transcatheter closure of peri-membranous ventricular septal defects with Nit-Occlud® Lê VSD coil

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Introduction: This study aims to investigate presence of arrhythmia, QT dispersion and heart rate variability (HRV) by performing ECG and Holter monitorization in patients whose VSD was closed with a transcatheter method using a Nit-Occlud® Lê VSD coil.

Patients and Method: The ECG and Holter recordings of 24 cases who presented to our clinic between January, 2010 and December, 2014 and whose ventricular septal defects were closed using a Nit-Occlud® Lê VSD coil were evaluated retrospectively. The 12-lead ECGs and Holter ECGs of the cases were taken before the procedure and at day 1, in month 6 and month 12 after the procedure. The presence of arrhythmias, HRV variables and QT dispersion were checked.

Results: In total, 24 cases were included in the study. Among these cases, 12 (50%) were male. Their average age was $7,9 \pm 4,6$ years (1.3–17.2 years) and average weight was $29,3 \pm 13,9$ kg (10–66 kg). regressed as seen in their Based on a comparison between the values before the procedure and in month 6 and month 12 after the procedure, no significant differences were identified between their QT min, max, dis, QTcmin, max, dis values and heart rate variability values at Holter ECG follow-ups (according to time domain and frequency domain analyses) (for every value, p value $> 0,05$). On the other hand, 3 patients had ventricular premature beats at a very rare frequency prior to the transcatheter closure of VSD with a Nit-Occlud® Lê VSD coil. After the procedure, the ventricular premature beats persisted in these patients and 2 (8%) patients had non-sustained ventricular tachycardia of 3 and 5 beats. However, this pathology regressed as seen in their Holter readings in month 6 and month 12. None of the patients developed atrioventricular block.

Conclusion: Following the transcatheter VSD closure with a Nit-Occlud® Lê VSD coil, no impairments in atrioventricular conduction, HRV changes or increased QT dispersions are observed. Although there were very brief non-sustained ventricular tachycardias following the procedure, they are resolved in the follow-up.

P-55

Platelet indices and their changes on the follow-up in children with pulmonary arterial hypertension

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Introduction: Inflammatory cells and platelets may also play a significant role in pathogenesis of pulmonary arterial hypertension (PAH). Previous studies have demonstrated that altered platelet functions occurs in patients with PAH. Nevertheless it remains unclear platelet dysfunction is a cause or consequence of the disease. Platelet indices include mean platelet volume (MPV) and platelet distribution width (PDW) is potentially useful markers for assessing the platelet function. We aimed to evaluate the platelet indices how it has changed before and after the treatment in PAH patients.

Method: Our study group consisted of 40 patients (19 females, 21 males, and median 5.5 years) with PAH. An age and gender matched control group was composed of 37 healthy peers (17 females, 20 males with a median age 5.0 years). Mean platelet volume and PDW values before and 6 months after the PAH therapy were obtained retrospectively from hospital records.

Results: Initial mean MPV value was significantly higher in PAH group when compared with control group (8.21 ± 1.29 vs 7.34 ± 0.788 fl, respectively; $P < 0.05$). There was no significant difference was observed between study and control group in terms of initial PDW value (16.28 ± 0.97 vs 16.55 ± 0.47 respectively; $P > 0.05$). After the 6 months of specific therapy with PAH-approved drug therapy significant difference was persisted between the study and control group in terms of MPV (8.27 ± 1.24 vs 7.34 ± 0.788 fl, respectively; $P < 0.05$). On the other hand no difference were detected between the initial and follow-up results of MPV ($p < 0.05$). Likewise we observed no significant difference in PDW values when compared before and 6 months after the PAH therapy (16.28 ± 0.97 vs 16.22 ± 1.14 , respectively; $P > 0.05$).

Conclusions: We have observed that MPV is significantly elevated in pediatric PAH patients. Medical therapies with PAH-approved drugs have no effect on the MPV values. This result suggested that specific PAH treatment have no positive effect on platelet function in PAH.

P-56

Evaluation of Ventricular Arrhythmogenesis In Patients With Rheumatic Carditis

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Acute rheumatic fever (ARF), is an important public health problem and leading cause of acquired heart disease in children and young adults worldwide. Carditis is the most important complication of RF that is associated with permanent disability. The electrocardiographic (ECG) findings of ARF have been well described. First degree atrioventricular block is the most common characteristic conduction disturbances of RF. In addition to these ECG findings, increased QT and QTc dispersions that represent the heterogeneity of ventricular depolarization and repolarization were found in patients with RC. Beside the QT and QTc

dispersions Tp-e interval which is the interval between the peak and the end of the T wave on an ECG, can be used as an index of transmural dispersion of ventricular repolarization. In this study, we aimed to assess Tp-e interval and Tp-e/QT ratio in children with RC and to investigate its relationships with inflammation and severity of valvular involvement.

A total of 139 patients, diagnosed with acute rheumatic carditis (RC) and 153 healthy controls were enrolled into study. The mean age of patients was 10.9 ± 2.4 years. In patients group p, QT and QTc dispersions, Tp-e intervals, Tp-e/QT and Tp-e/QTc were found significantly higher than controls. Isolated mitral or aortic regurgitation was present in 79 of the patients. When patients with isolated mitral (MR) or aortic (AR) regurgitation were compared to patients with both valvular (MR + AR) involvement. There was no any differences found in p and QT dispersions and Tp-e intervals, Tp-e/QT and Tp-e/QTc measurements. However, QTc dispersion was significantly higher in patients with isolated valvular involvement. Erythrocyte sedimentation rate was significantly higher in patients with both valvular involvement. However there was not found any correlation between WBC, ESR and p, QT and QTc dispersions, Tp-e intervals, Tp-e/QT and Tp-e/QTc ratios.

RC is considered to be pancarditis. Although the valvular component is much more important on the prognosis than myocardial or pericardial involvement, myocarditis is the most important element of the RC. Therefore, patients with RC may susceptible to ventricular arrhythmias. However, ventricular arrhythmogenesis seems to be independent from severity and number of valvular involvement and acute phase reactants.

P-57

An epidemiological study of pediatric pulmonary hypertension in Turkey

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Objective: The present study aims to evaluate the nationwide epidemiological characteristics of pediatric pulmonary hypertension (PH) within whole Turkish population over a period of five years using the registry of the National Health Insurance System.

Methods: All individuals aged less than 18 years who were admitted to a Turkish hospital for the first time between 2009 and 2013 with a discharge diagnosis of primary PH (ICD-10 code I27.0) and secondary PH (ICD-10 code I27.2) were identified.

Results: The overall annual prevalence of idiopathic PH at pediatric age was 11.7 cases per million whereas the overall annual prevalence of secondary PH during childhood was 9.5 cases per million. There was a gradual and significant increase in the annual prevalence of idiopathic PH and that of secondary PH during the five-year study period ($p = 0.001$ for both) (Figure 1). In the years 2012 and 2013, idiopathic PH was significantly more frequent in children aged less than two years ($p = 0.002$ for both). The frequency of secondary PH did not change significantly with respect to patient age at each admission year (Table 1). The male to female ratio was 1.2:1 for idiopathic PH at pediatric age while the female to male ratio was 1.1:1 for secondary PH during childhood. The frequency of idiopathic PH and secondary PH at pediatric age remained statistically similar for both sexes within the study period.

Conclusion: These findings demonstrate that the prevalence of pediatric PH in Turkey is higher than those reported for the Western populations. Moreover, no female dominance could be

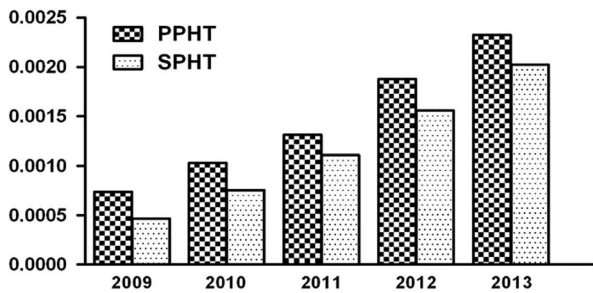


Figure 1 Annual prevalence of primary and secondary pulmonary hypertension during the study period (PPHT: primary pulmonary hypertension, SPHT: secondary pulmonary hypertension).

Table 1. Distribution of pediatric pulmonary hypertension cases according to diagnostic classification, admission year and age

Year of admission	Primary pulmonary hypertension			Secondary pulmonary hypertension			Total
	0-2 years	3-17 years	Total	0-2 years	3-17 years	Total	
2009	230 (61.2%)	146 (38.8%)	376	120 (50.6%)	117 (49.4%)	237	613
2010	364 (63.9%)	206 (36.1%)	570	229 (54.9%)	188 (45.1%)	417	987
2011	525 (66.6%)	263 (33.4%)	788	404 (60.8%)	261 (39.2%)	665	1453
2012	870 (72.4%)	332 (27.6%) †	1202	584 (58.6%)	413 (41.4%)	997	2199
2013	1033 (73.0%)	383 (27.0%) †	1416	669 (54.3%)	564 (45.7%)	1233	2649

† Primary pulmonary hypertension was significantly more frequent in children aged less than two years ($p=0.002$).

noted. These discrepancies may be attributed to the differences in the study design, study cohort, timing of the study and the definitions adopted for PH classification.

Key words: epidemiology, pediatric, pulmonary hypertension

P-58

Risk Factors for Out-of-Range International Normalized Ratio in Patients after Total Cavopulmonary Connection Receiving Warfarin

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Introduction: Warfarin generally prevents thrombosis in patients after total cavopulmonary connection (TCPC); however, out-of-range international normalized ratio (INR) is often occurred. This study sought to identify risk factors for out-of-range INR in patients after TCPC receiving warfarin.

Methods: Patients after TCPC who underwent INR monitoring for dose optimization of warfarin were recruited from 2004 to 2013. INR measurements which were performed <1 week after initiation of warfarin, those from patients who used combined medicines having an interaction with warfarin except aspirin and heparin, and those from patients with liver or renal dysfunction were excluded from the study. No patients experienced pharmacogenomics of warfarin. Age, gender, body weight (BW), warfarin dose, warfarin dose/BW and INR were analysed. INR values were divided into 3 groups; optimal (INR 1.5-3.0), under- (INR <1.5) and over-anticoagulation (INR >3.0).

Results: A total of 602 INR measurements from 19 patients fulfilled the criteria. There were neither thrombotic nor haemorrhagic events during the period. Median age, female gender, median BW, median warfarin dose, median warfarin dose/BW

and median INR values were 7.5 years (range 1.6-32.8), 58%, 20.5 kg (range 7.8-59), 2 mg (range 0.3-4.75), 0.081 mg/kg (range 0.026-0.219) and 1.63 (range 1.07-4.48), respectively. Out-of-range INR was observed in 38% of measurements; under- and over-anticoagulation was 35% and 3%, respectively. INR values showed significantly negative correlation with age and BW. The over-anticoagulation group comprised significantly lower age and BW and less female patients compared with the optimal anticoagulation group (median, 4.3 vs. 7.7 years, 12.7 vs. 20.4 kg and 50% vs. 74%, respectively). In addition, the former showed significantly more warfarin dose/BW compared with the latter (median, 0.10 vs. 0.08 mg/kg). Receiver operating characteristic curve analysis determined the sensitivity and specificity for predicting over-anticoagulation to be 58% and 90% for age <6.5 years and 62% and 95% for BW <16.5 kg, respectively.

Conclusions: Out-of-range INR often occurred in patients after TCPC receiving warfarin. Lower age and BW were the risk factors for over-anticoagulation.

P-59

Left Superior Vena Cava Accompanying Congenital Heart Diseases in Children, Experience of a Tertiary Care Center

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Objective: Determination of concomitant persistent left superior vena cava (PLSVC) in patients with congenital heart diseases.

Materials and Methods: Between May 2005 - November 2012, 2663 patients with congenital heart disease and 88 (3.3%) of them, diagnosed with PLSVC, were evaluated retrospectively. The demographic characteristics of patients, concomitant congenital heart diseases, clinical and radiographic findings, echocardiography, cardiac catheterization and angiography results which obtained from the patients' records were evaluated.

Results: The median age of the patients was 9.5 months (1 month-14 years); 46 (52.3%) patients were female. The most common concomitant congenital heart diseases were ventricular septal defect (23.9%), double outlet right ventricle (14.8%) and tetralogy of Fallot (11.4%), respectively. When the patients according to their heart disease were compared with the whole group; PLSVC frequency was significantly higher in patients with double outlet right ventricle ($p < 0.001$), tetralogy of Fallot ($p = 0.04$), patent ductus arteriosus ($p = 0.01$) and atrial septal defect ($p = 0.03$). PLSVC were opening into the coronary sinus in all cases (100%). Eighty-three (94.3%) of 88 patients with PLSVC were found to have right SVC, and 5 (5.7%) were found to have agenesis of the right SVC. Twenty-seven (32.5%) of 83 patients with double SVC had intercaval connection. In all cases (100%) left SVC was observed to open into the coronary sinus. Right aortic arch association was seen in 14 (15.9%) patients. Eighty-four (95.4%) of the patients were evaluated by echocardiography prior to catheter angiography.

Conclusion: Gaining awareness about the congenital heart diseases that frequently associates with PLSVC and a careful echocardiographic examination can simplify the diagnosis of PLSVC. The early diagnosis is important to prevent the complications during catheterization and surgery.

P-60

The evaluation of mutation analyses results of patients with familial hypercholesterolemia by Simone-Broome criteria

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Objective: Familial hypercholesterolemia (FH) is an autosomal dominant disease characterised with defective cholesterol removal from circulation and hypercholesterolemia caused by mutations in *LDLR* gene. Although definitive diagnosis is made by mutation analysis, some diagnostic criteria have been developed due to the impracticality of routine mutation analysis. This study aims to compare mutation status of familial hypercholesterolemia patients and their diagnoses according to the Simone Broome criteria (SBC).

Material and Methods: All patients with primary hyperlipidemia followed up in the Metabolism and Nutrition Unit of Hacettepe University Medical School were classified according to the Fredrickson classification system. Patients with type IIa hyperlipoproteinemia was assessed by SBC for the diagnosis of FH. Denaturing high-performance liquid chromatography (DHPLC) and DNA sequencing was performed on the most frequently mutated exons (9 and 12). Mutation analyses of this and previous cohorts were compared with SBC in terms of diagnostic compatibility.

Results: 118 of 163 patients with primary hyperlipidemia were type IIa hyperlipoproteinemia according to Fredrickson classification. According to the SBC, 48% of these patients were in the “probable FH”, 26% were in “definite FH”, 17% were in “no FH” group, and 9% could not be classified due to insufficient information. On exon 9 heterozygous I420N (C.1322 T > A) mutation was seen in 2 patients and on exon 12 and one heterozygous and one homozygous W556R (C.1729 T > C) mutation were observed. When we combined these results with 29 patients results that were previously screened, mutations were seen in 10 out of 10 patients (100%) whose mutation statuses were analyzed in the definite FH, 12 out of 16 (75%) patients in the probable FH and 1 out of 3 (33%) patients in the no FH group.

Conclusion: When routine mutation screening is not a feasible choice or when selective screening is performed, diagnosing

familial hypercholesterolemia based on the Simone Broome criteria is a suitable approach.

P-61

Effect Of Pulmonary Hypertension Related Congenital Heart Disease With Left To Right Shunt On The Serum Levels Of High Sensitive Cardiac Troponins In Children

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Introduction: Anatomical anomalies of the heart are generally associated with abnormal hemodynamic load and neurohumoral activation. Significant hemodynamic overload due to a left-to-right shunt can induce myocardial injury. Sensitive cardiac troponin I (cTnI) and high sensitive cardiac troponin T (cTnT) are currently considered to be the most sensitive and specific biochemical marker of myocardial injury. However, few reports have described the use of cTnI, cTnT assays for evaluating abnormal volume and pressure load in children with congenital heart disease. For that reason we want to evaluate cTnI and cTnT levels in children with left-right shunt congenital heart disease and effect of pulmonary hypertension on serum levels of cTnI and cTnT.

Methods: We prospectively enrolled 69 clinically stable subjects, consisting of 51 patients with left to right shunt and 18 healthy children. The first group was also been divided two; 20 patients with pulmonary hypertension, and 31 patients without pulmonary hypertension.

Results: The mean age was 12.1 months (4 months and 3 years). Cardiac catheterization was performed in 25 children with ventricular septal defect prior to surgery and 4 children for percutaneous closure of ductus arteriosus. Serum cTnI and cTnT levels were found to be significantly higher in pulmonary hypertensive patients. In particular, cTnT levels were significantly elevated in patients with left to right shunt without pulmonary hypertension (p <0.001).

Conclusions: Significant volume and pressure overload due to a left-to-right shunt induce myocardial injury. The serum cTnI, cTnT levels are useful biomarkers for evaluating myocardial damage associated with pulmonary hypertension in congenital heart disease left to right shunt children. We also suggest that the use of cTnI, cTnT as bio- markers might aid in the earlier detection of myocardial injury in patients with congenital heart disease.

Table. Mean value of biochemical parameters

	Control group	Group with PH	Group without PH
c Tn I	0.008 ± 0.007	0.039 ± 0.06	0.47 ± 2.37
cTn T	6.22 ± 2.21	18.98 ± 12.2	13.03 ± 13.8

P-62

Incidence of respiratory diseases in hemodynamically significant congenital heart disease (hsCHD) children in Italy: the SINERGY study

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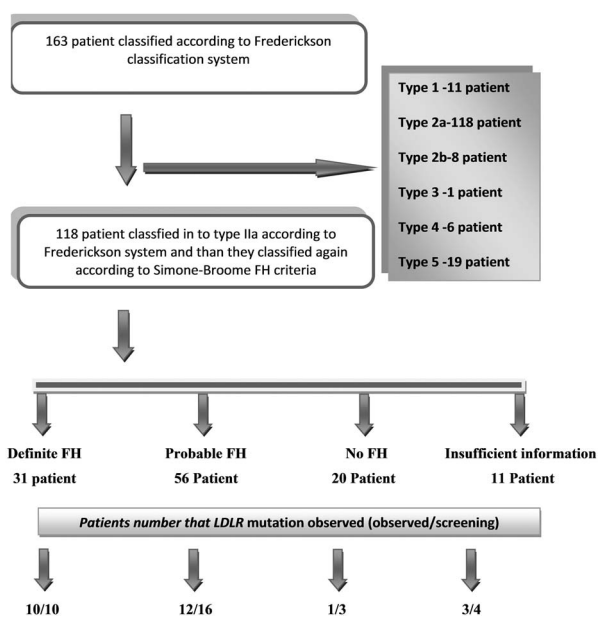


Figure.

Introduction: Children affected by hsCHD are likely to experience severe respiratory complications that increase the number of hospitalizations. Objectives of the SINERGY study were to describe the incidence of respiratory diseases and to collect information on the active and passive immunoprophylaxis during the first two years of life.

Methods: This retrospective, multicenter, epidemiological study enrolled 420 hsCHD children in 11 Italian sites according to Italian Guidelines for hsCHD diagnosis. Subjects affected by HIV or malignancies were excluded from the study.

Observation occurred in children born between 31/12/2007 and 31/12/2012. Data have been collected through hospital databases search and parents' interviews.

Results: 420 children were enrolled in the study: 52% female; 80% full-term born, 78% weighed >2500 g at birth, 14% presented with a genetic syndrome.

Ninety-four percent of the population showed 1 or 2 heart defects: the most frequent were interventricular septal defects (23% overall; 11% in combination with other cardiopathies), interatrial septal defect (14%; 11% in combination), and coarctation of the aorta (12%; 8% in combination).

The major risk factors were hospital discharge during epidemic season (54%), presence of older siblings (44%) and no breast feeding (41%). The overall incidence of any respiratory disease was 63.1%.

The most frequent respiratory diseases which didn't require hospitalization were acute rhinopharyngitis, bronchitis and influenza (27%, 27%, 14%); while required hospitalization mainly bronchitis and bronchiolitis cases (21%) primarily in December and January during the first year of life. The infection's etiology was identified only in 23 children (5,5%).

Active immunoprophylaxis was applied with wide compliance (Diphtheria/Pertussis/Tetanus: 99.5%; Haemophilus influenzae b: 72.5%; Pneumococcus: 80% Meningococcus: 77%), while only 54% of the study population received a respiratory syncytial virus (RSV) passive prophylaxis (palivizumab). In the bronchiolitis hospitalized population (33 children), 26 children (78%) were not prophylaxed against RSV or received only the first dose before the event, while 7 children (22%) were fully prophylaxed ($p < 0.001$).

Conclusions: hsCHD children are at major risk for respiratory diseases: passive immunoprophylaxis can help to prevent hospitalizations for bronchiolitis.

Active immunoprophylaxes showed different degrees of compliance, probably because DTP vaccine is mandatory in Italy, while other vaccines are at parents' discretion.

P-63

Towards improved Rheumatic Heart Disease control and prevention in Fiji Islands

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Introduction: Rheumatic Heart Disease (RHD) is a significant health problem in Fiji Islands with the Pacific region having among the highest reported RHD prevalence in the world. It is common for children to present late with severe RHD. Episodes of acute rheumatic fever (ARF) are often unrecognised by families and health practitioners. The Fiji Islands RHD Control and Prevention project is a four year initiative led by Cure_Kids, a New Zealand (NZ) based charity that funds child health research, in collaboration with the Fiji Ministry of Health (MOH), Auckland District Health Board (Starship Hospital and Auckland Regional

Public Health Service), and the Centre for International Child Health at Murdoch Children's Research Institute). Joint funding of NZD \$3,109,564 is provided by the NZ Ministry of Foreign Affairs and Trade, and Cure_Kids.

Methods: The Project has four key areas: (1) a national register-based secondary prevention programme; (2) development of best practice guidelines for clinical care; (3) a national model for early case detection; and (4) ARF primary prevention and health promotion strategy. The Project will facilitate co-ordination of all ARF/RHD activities, integration into existing services and models of care, and development of new models of care with the aim of creating sustainable and effective ARF/RHD control in Fiji. Fundamental to the Project outputs is an effective national co-ordination structure for the Fiji RHD Control Programme (Fiji MOH). Increasing capacity at this level will provide a governance model that can continue beyond the life of the Project. The effectiveness of the Project will be evaluated via monitoring outcomes associated with each of the areas. These include the effectiveness of the register based approach to deliver benzathine penicillin, monitoring of ARF/RHD incidence and prevalence, improved diagnosis and management of patients with ARF/RHD, improved public awareness of ARF/RHD and understanding of appropriate health seeking behaviour. Additional long-term outcome measures include incidence, prevalence, morbidity, and mortality. Year 1 focuses on establishing a baseline against which programme effectiveness can be measured.

Conclusions: The Project, with its multiple partners, has the potential to contribute substantially to the development of a sustainable ARF/RHD programme and infrastructure in Fiji Islands.

P-64

An Audit of Patients with Muscular Dystrophy Attending a UK Regional Paediatric Cardiology Department

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Objective: The aim was to examine the current practices employed by the cardiology team in the investigation, monitoring and treatment of patients with muscular dystrophy.

Methods: A search for all patients aged 0-16 years with a diagnosis of Muscular Dystrophy (MD) attending the paediatric cardiology department was performed using the Heartsuite Database. Patients who did not have a confirmed diagnosis were excluded. Electronic medical records were then accessed and examined. Details extracted included patient demographics, age at time of referral to cardiology, initial investigation findings, current cardiac status, and drug therapy. In particular, attention was paid to what measurements were being taken during echocardiogram (ECHO) assessment and the timing and choice of drug when medication was first introduced.

Results: 36 patients aged between 4-16 years were identified. 77% of patients had a diagnosis of Duchenne's Muscular Dystrophy while Becker's Muscular Dystrophy accounted for 6%. A wide range of ages was seen at the time of initial referral to Cardiology (1-11 years). At initial assessment 94% had a normal 12 lead electrocardiogram (ECG) and 100% a normal ECHO (78% at this time had ejection fraction/shortening fraction (EF/FS) calculated while 3% had tissue doppler performed). Review intervals for patients were found to be variable ranging from 6 monthly to 3 yearly. At their most recent review, 83% had a normal ECG, while 87% had normal ECHO findings. At this stage 30% had tissue doppler performed and 60% had EF/FS calculated. Of those with abnormal ECHO findings, all had EF/FS calculated, while 75% had tissue doppler performed. Nine patients were found to be on medical therapy. Seven different drugs were prescribed including 3 different types of beta-blocker (5 patients), 3 different ACE-inhibitors (7 patients) and digoxin (1 patient). 3 patients required multi-drug therapy.

Conclusions: This audit of current practice highlighted that there was little consistency in the management of patients with MD. As a result the department has drawn up a new proposed guideline for the management of this patient group.

P-65

Exercise Tolerance and Participation in Sports in Patients with Congenital Heart Disease

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Introduction: Exercise tolerance in patients with congenital heart disease (CHD) varies depending on the heart disease, general physical condition and other factors. Data concerning this group is scarce.

Methods: In 2014 199 patients were examined by cardiopulmonary exercise testing (VO₂max, ml/min/kg) and the predicted VO₂max (%) was calculated according to the data of Cooper (1984, 2000). The results were evaluated in the diagnostic subgroups (table): valvular disease, TGA after arterial switch operation (ASO), simple lesions (ASD, VSD, coarctation, PDA), Ebstein anomaly, other CHD, TGA after atrial switch (Senning), Fontan circulation and complex cyanotic CHD. The patients filled out a questionnaire on physical activity and results were compared with 2 studies on healthy German children (KiGGS n = 17,000) and adults (DEGS1 n = 8152).

Results: 173 patients' exercise data was sufficient for analysis. Patients' median age was 26, 8-69 years; 88 patients were female (50.8%) and the median VO₂max was 23.7 ml/(kg*min) (70% of predicted). The maximal exercise capacity was found in patients with valvular disease followed by corrected TGA (ASO) and the lowest capacity was in Fontan patients and those with complex cyanotic disease. According to the questionnaire, patients with CHD were less active in sports than healthy subjects. This gap is larger in male patients. The largest differences were seen in women aged 18-29 years and men of 30-39 years.

Conclusion: VO₂max in the total population and in the subgroups is comparable to the data published so far. Even CHD patients with simple lesions show impaired exercise tolerance. It stands to reason that not only hemodynamics, but other factors, e.g. attitude of caregivers, patients and physicians contribute. CHD patients should be actively motivated to participate in sports.

Table.

Diagnosis	Number	Age (years)	VO ₂ max (ml/min/kg)	VO ₂ max %
total	173	26	23.7	70
valvular disease	43	24	27.9	77
TGA (ASO)	7	17	27.0	76
simple lesions	22	31.5	24.8	75
Ebstein	16	48.5	19.5	71
other CHD	26	23.5	23.5	71
TGA (Senning)	16	31.5	22.3	68
TOF	47	28	23.4	66
Fontan	18	21	21.3	58
complex cyanotic	4	38	17.7	40

Median values for age and VO₂ max

P-66

Representativeness of the German National Register for Congenital Heart Defects. A clinically oriented analysis

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Introduction: The German National Register for Congenital Heart Defects (NRCHD) is a clinical register comprising data from approximately 48,000 members (largest patient database in Europe). Approximately 6,000 children are born with congenital heart disease (CHD) in Germany each year. They die from their chronic illness increasingly rarely. In the present data analysis, birth years recorded in the NRCHD are compared to results of the PAN study and a meta-analysis by van der Linde et al. with respect to prevalence rates and sex distribution.

Methods: A descriptive data analysis was performed by using a minimal data set. The demographic data include sex and birth year; the medical data comprise the cardiovascular diagnosis according to the IPCC Shortlist. Two birth years recorded in the NRCHD (2004/2005) and two birth years recorded in the PAN study (2007/2008), were compared with each other, as well as with the results of van der Linde et al., with respect to the prevalence rates of CHD.

Results: The two birth years of 2004/2005 as recorded in the NRCHD deviated from each other by an average 0.8% (minimum 0.0% to maximum 1.7%) regarding the diagnosis groups. In three of the nine diagnosis groups (ASD, TOF and Other), the prevalence difference between the two years was < 1.0%. The prevalence rates as recorded in the NRCHD can thus be assessed as very accurate. The birth years of 2007/2008 as recorded by the PAN study show an average deviation of 0.7% (min.: 0.0%, max.: 1.9%) with respect to the prevalence rates. Being a clinical register, the NRCHD primarily includes clinical cases/cases relevant to health care. The prevalence values and sex ratios recorded therein are closer to the values given in the literature than those determined by the PAN study. Severe CHD are slightly overrepresented in the NRCHD as compared to van der Linde et al. Deviations with respect to prevalence values are within acceptable range.

Conclusions: The patient population registered in the NRCHD can be considered as representative for CHD in Germany and Europe. Samples from the NRCHD can thus be expected to represent the gold standard for future studies.

P-67

Research priorities in the field of congenital heart disease as viewed by patients and physicians: A survey by the Competence Network for Congenital Heart Defects

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Introduction: Patients and their relatives, as well as physicians of paediatric cardiology, cardiology and cardiac surgery were surveyed to identify current domains in the field of congenital heart disease (CHD) requiring further research and to thus consider the needs of those affected.

Methods: Over a period of 30 days, an online survey concerning four groups of CHD (Fontan circulation, transposition of the great

Table. Overview of the topics rated highest

Fontan circulation	<ul style="list-style-type: none"> • "Failing Fontan" • Quality of life • Career choices/pension • Diagnostic imaging • Anticoagulation
TGA after atrial switch	<ul style="list-style-type: none"> • Catheter ablation, ICD implantation, sudden cardiac death • Diagnostic imaging • Diagnostics and treatment of heart failure • Catheter based intervention, re-operation
TGA after ASO	<ul style="list-style-type: none"> • Dilatation of the great vessels, AV valve related complications, subsequent interventions/surgery • Catheter ablation, ICD implantation, sudden cardiac death • Problems related to the coronary arteries • Diagnostic imaging • Cognitive function
TOF	<ul style="list-style-type: none"> • Catheter ablation, ICD implantation, sudden cardiac death • Catheter intervention, re-operation • Diagnostics and treatment of heart failure • Diagnostic imaging

arteries [TGA] after atrial switch, TGA after arterial switch [TGA-ASO], tetralogy of Fallot [TOF]) was promoted. The online questionnaires comprised twelve issues each. For evaluation, a scale from 1 (no need for research) to 6 (urgent need for research) was used. Patients/relatives were asked questions specific to their diagnosis. Patients were recruited from the German National Register for Congenital Heart Defects.

596 patients/relatives took part (Fontan circulation: n = 189; TGA after atrial switch: n = 64; TGA-ASO: n = 90; TOF: n = 253), as well as 75 physicians (57.3% paediatric cardiologists, 28.0% cardiologists, 10.7% cardiothoracic surgeons and 4.0% other).

Additionally, the patients' age, sex, educational background and employment status and the physicians' sex, work areas and function were recorded. The total response rate of patients/relatives was 57.2%. **Results:** Apart from a few exceptions, patients rated all fields higher than physicians, thus assuming a greater need for research in all areas. With the exception of the group of TGA after atrial switch, far more females (68%) than males from the patients/relatives group took part in the survey. Female physicians assumed a greater need for research into psychosocial issues than their male counterparts. Regarding the assessed need for research, at times significant differences were found between physicians of the different subspecialties. Patients and relatives in large part – albeit with different emphasis on and ranking of single topics – agreed with the physicians regarding the choice of CHD related topics primarily requiring further research.

Conclusions: The topics considered to be the most important by both patients/relatives and physicians point to the everyday challenges of offering health care to named patient groups. Special emphasis should thus be placed on investigating these topics in the coming years.

P-68

A German Biobank for Congenital Heart Defects

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Universitätsklinikum Erlangen, Dep. of Pediatric Cardiac Surgery, Germany (4); Heart Center Universität Leipzig, Dep. of Pediatric Cardiology, Germany (5); Universitätsklinikum Erlangen, Dep. of Pediatric Cardiology, Germany (6); Universitätsklinikum Schleswig-Holstein/Kiel, Dep. of Congenital Heart Disease and Pediatric Cardiology, Germany (7); German Heart Center Munich, Dep. of Cardiovascular Surgery/Division of Experimental Surgery, Germany (8); HDZ-NRW Bad Oeynhausen, Dep. of Pediatric Cardiology & Congenital Heart Disease, Germany (9); German Heart Institute Berlin, Dep. of Surgery for Congenital Heart Disease/Pediatric Cardiac Surgery, Germany (10); Universitätsklinikum Freiburg, Dep. of Congenital Heart Disease and Pediatric Cardiology, Germany (11); DZHK (German Centre for Cardiovascular Research) Partner Site (12)

A multicentre biobank infrastructure was established in order to support current and future scientific investigations in the field of congenital heart defects (CHD) and cardiovascular diseases. The biobank is an integral part of the German Registry for congenital heart defects and collects DNA from patients plus parents (trios) and affected families, and cardiac tissue from patients undergoing heart surgery. So far, eight ethics committees in Germany have approved the CHD-Biobank concept that implies indefinite storage of samples and their use in on-going and in still undefined future studies. Compliance with data privacy regulations have been confirmed by the Berlin Official for Data Protection. Currently, pediatric cardiology/heart surgery departments of eight hospitals in Germany are involved.

The CHD-Biobank is centrally managed by the Registry team that is responsible for the validation of clinical/phenotype data, the compliance with legal and ethical regulations, the implementation of state-of-the-science protocols for sample acquisition, processing and storage, and the implementation and efficient use of software solutions for data and biomaterial management.

The CHD-Biobank operates

1. a central ID management using uniform coding systems (including barcoded sample containers) and a central online-accessible database platform
2. a central facility for blood/DNA processing, storage and dissemination of samples (partnership with the Charité-Berlin centralized Biomaterialbank ZeBanC)
3. a decentralized sample collection structure for cardiac tissue samples that need to be stored in liquid nitrogen gas phase (below -150°C).

The DNA collection currently comprises samples from approximately 3,600 participants covering a wide range of CHD phenotypes. The collection includes 420 trios and 150 families with more than one affected member. The cardiac tissue collection comprises approx. 1200 tissue samples from 500 patients with open heart surgery.

The rising number of requests for collaboration from numerous research institutions (in addition to various German institutions also Newcastle Genetic Medicine, WT Sanger Institute Cambridge) can be regarded as an indication for the high quality of the CHD-Biobank infrastructure, sample logistics and phenotype database.

P-69

Carotid and Subclavian Aneurysms in Infants Following Neonatal Norwood Surgery: Report of Seven Cases

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Introduction: Following incidental finding of a left common carotid aneurysm during diagnostic cardiac catheterisation prior to cavopulmonary shunt surgery, we sought to investigate the occurrence of head and neck artery anomalies following neonatal Norwood surgery.

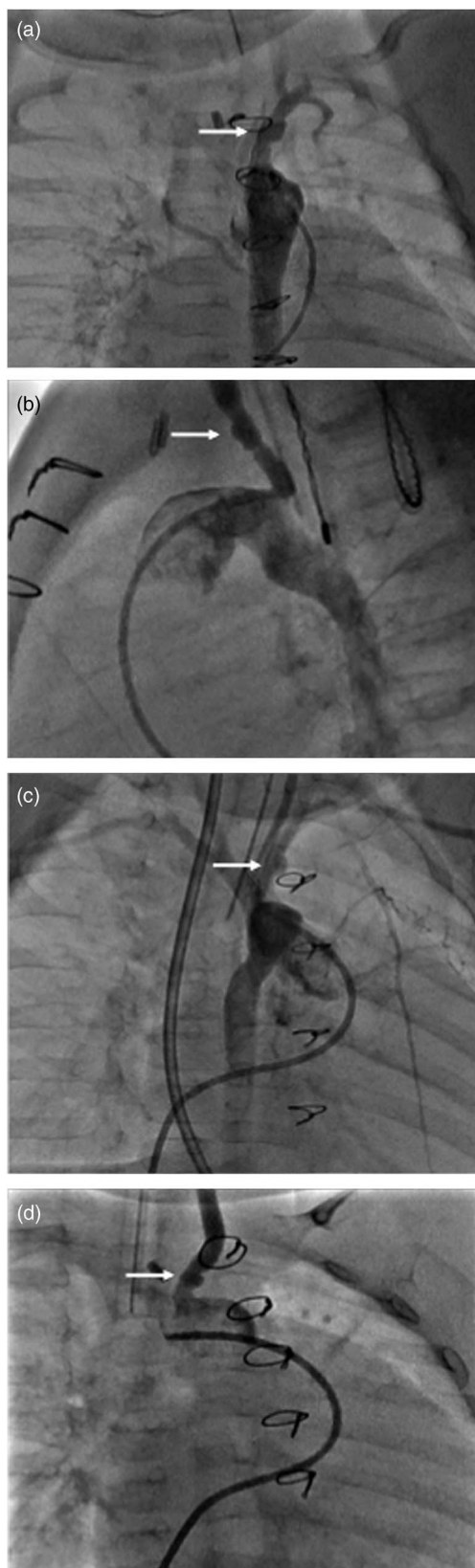


Figure 1.

As the only patient group to undergo both neonatal bypass surgery and routing advanced imaging of the aortic arch, those selected for this analysis were exclusively post-Norwood palliation.

Methods: Retrospective data review for 80 sequential patients post Norwood surgery and diagnostic catheterisation at a single tertiary referral center (May 2010–March 2014).

Results: Carotid or subclavian arterial abnormality was identified in seven patients (8.8%). See figure 1 for example angiography. In all patients the lesion was proximal to the arch. Lesions consisted of mixed stenosis and aneurysm. The largest aneurysm measured 4 mm. No patient had any clinical manifestation of their abnormality.

Discussion: The relevance and natural history of such abnormalities is unreported. The adult literature for extra-cranial carotid aneurysms predominantly describes thrombo-embolism as the most commonly reported manifestation. In a case series of four symptomatic children with carotid artery aneurysms from a variety of aetiologies, one patient had CVA with focal neurological deficit, local signs were identifiable in all. We hypothesised as a mechanism, the use of silk snuggers' for head and neck vessels during circulatory arrest and selective head and neck perfusion during cardiac bypass.

Patients undergoing neonatal bypass surgery with circulatory arrest other than Norwood do not routinely undergo advanced imaging, thus the true incidence of these anomalies remains undefined. Patient numbers in this report were insufficient for statistically significant delineation of risk factors for aneurysm.

This report highlights the importance of assessing proximal arch vessels during advanced imaging for complex patients who have undergone selective head and neck perfusion as part of arch reconstruction. The incidence, mechanism and natural history of these lesions remains undefined. Further evaluation and follow-up of these patients will be required to guide future practice.

P-70

Assessment of myocardial deformation with 2-dimensional speckle tracking echocardiography in children with isolated subaortic stenosis after surgery

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Introduction: Here, we aimed to investigate whether strain and strain rate could detect subtle left ventricular dysfunction and impairment in regional myocardial functions in children with repaired isolated subaortic stenosis after mid-term follow up.

Methods: We studied 20 patient (%40 female, mean age was 15.2 ± 6.86 years) and 31 healthy subjects (%40 female). Electrocardiography was performed in all subjects. NT-pro BNP levels were determined in patient group after exercise ECG. By using two dimensional speckle tracking echocardiography, strain and strain rate were measured in all subjects. Multiple linear regression analysis was used to determine independent variables on longitudinal and radial strain.

Results: Operation age was between 2 and 24 years (mean 8.05 ± 5.57 years), and mean postoperative follow-up time was between 2 and 14 years (mean 7.15 ± 3.26). In the patient group, preoperative left ventricular outflow tract gradient was 73.35 ± 23.70 mmHg, and postoperative left ventricular outflow

gradient was 22.80 ± 19.31 mmHg. In the patient group, mean NT-pro BNP level was 193.89 ± 611.13 pg/mL. Relative to the healthy subjects, interventricular septal wall thickness in diastole (IVSd) was significantly higher in the patient group ($p=0.001$). EF, FS and VCFc were found to be significantly lower in patients than in controls ($p=0.001$). In the patient group, mitral E' ($p=0.03$) and septum E'/A' ($p=0.03$) were significantly lower than in controls. Septum MPI, mitral MPI and aortic strain (using M mode) were found to be significantly higher in patients than in controls ($p=0.03$, $p=0.02$, $p=0.001$, respectively). Aortic distensibility was significantly lower in patient than in controls ($p=0.034$). Longitudinal strain of LV was found to be significantly lower in patient than in controls ($p=0.025$). Radial and circumferential strain indices were not statistically different between two groups. Multivariable analysis showed positive correlation between aortic strain and longitudinal strain ($\beta=0.138$, $p=0.034$; 95% CI: 0.010-0.265).

Conclusions: Longitudinal strain of left ventricle is significantly impaired in children with subaortic stenosis after surgery. Longitudinal strain shows correlation with aortic strain. Longitudinal strain seemed to be superior to radial and circumferential strain of left ventricle in the follow-up of children with subaortic stenosis.

P-71

Symptomatic myocardial bridging in a young soccer player without hypertrophic cardiomyopathy

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Introduction: Myocardial bridging, muscle surrounding the segment of the epicardial coronary artery, is almost always associated with hypertrophic cardiomyopathy and left ventricular hypertrophy. Here, we report a 14-year old boy with presyncope and chest pain during exercise related with myocardial bridging of the middle segment of left anterior descending coronary artery (LAD).

Case: A 14-year-old boy was admitted to our pediatric cardiology department for exertional chest pain and presyncope. The attacks generally lasted up to 45 minutes-1 hour, were associated with dizziness, anxiety and precordial chest pain. Physical examination was normal with a normal body weight and body mass index. Vital signs were normal. Cardiac examination was normal. Initial electrocardiography showed sinus rhythm, normal QRS axis and



Figure 1. Cardiac angiography. a. Compression of the mid-left anterior descending artery during systole (arrow). b. Same segment during diastole (arrow).

no evidence of left ventricular hypertrophy. Blood tests did not reveal anemia, electrolyte or lipid abnormalities. On admission, troponin, CK-MB and NT-pro BNP levels were normal. Echocardiography showed normal cardiac and coronary anatomy. Exercise stress test (Bruce protocol) was stopped after 5 minutes due to inverted T wave in inferior and anterior leads (DII, DIII, aVF, V3, V4, V5). Dual source computed tomographic coronary angiography did not reveal any abnormality of coronary arteries. Exercise Tc-99 MIBI myocardial perfusion scanning showed minimal reversible perfusion defect in anterolateral wall consistent with ischemia. Based on these findings, we decided to perform coronary angiography, which revealed a coronary myocardial bridging and systolic compression with a length of 15 mm in the mid segment of the LAD (Figure 1). Patient was restricted from playing football, and calcium channel blocker was prescribed and he was discharged without any problems. After 3-month follow-up, he had no cardiac symptoms.

Conclusion: Myocardial bridging should also be considered in children without associated hypertrophic cardiomyopathy and left ventricular hypertrophy who presents with clinical findings that are consistent with myocardial ischemia.

P-72

The role of mitral propagation velocity, tricuspid annular plane systolic excursion and biventricular volumes in the assessment of children with repaired tetralogy of Fallot

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Introduction: We aimed to investigate mitral flow propagation velocity (Vp), TAPSE, biventricular volumes, BNP and exercise test, to explore the association between these parameters and clinical status.

Methods: Patients with repaired TOF (mean age, 11.6 ± 2.7 years, 14 boys, 11 girls) were selected from clinical database. All patients were classified as NYHA functional class I. Forty percent of patients had palliative shunts before corrective surgery (Blalock-Taussig shunt in 10 patients). We obtained Vp and TAPSE. Duration of exercise, blood pressure, maximum heart rate and finger pulse oximetry were recorded continuously in exercise testing. Venous blood samples for the determination of BNP values were obtained from all subjects before and after the exercise test.

Results: There was no significant differences between the levels of brain natriuretic peptide before and after exercise in the study group ($p=0.320$, $p=0.321$ respectively). Comparison of brain natriuretic levels after exercise in the study group revealed that patients who had modified BT shunt before corrective surgery had significantly higher levels compared to patients who had not history of modified BT shunt ($p=0.013$). Right ventricular end diastolic and end systolic volume indexed to body surface area was significantly higher in patients than controls ($p=0.001$). Patient group had higher left ventricle eccentricity index ($p=0.001$). The mean TAPSE values were significantly lower in study group when compared with healthy subjects ($p=0.001$). Vp were higher in

patient group, however there were no statistical significance ($p=0.655$). Duration of follow-up after surgery was negative correlated with BNP level before and after exercise and positive correlated with right ventricular end diastolic and end systolic volume in patient group ($r=0.507$, $r=0.648$, $r=0.587$, $r=0.598$ respectively).

Conclusions: According to the mid-term results of children with repaired TOF, we did not show any statistical difference in mitral flow propagation velocity (V_p). Up to our knowledge, there were no study regarding V_p . TAPSE along with BNP can be used in the follow-up of patient with repaired TOF.

P-73

Syncope in Children; Is Rhythm Holter Monitoring Necessary?

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Objectives: Holter monitoring (HM) is usually used in patients with syncope when etiology cannot be explained with history, physical examination and electrocardiography (ECG). Our objective was to evaluate the diagnostic value of Holter monitoring in children with syncope.

Patients and Methods: Databases were collected retrospectively by analyzing the HM results of 3122 pediatric patients between 2010–2014. Gender, age at initial syncope, detailed clinical history, physical examination, 12-lead electrocardiographic and echocardiographic results were noted using standardized form.

Results: The study included 323 patients with syncope with a mean age of 13.21 ± 3.67 . There were 199 female and 124 male patients. Among all patients 284 (87.9%) had normal HM results while there were 11 (3.4%) abnormal holter studies considered to explain syncope. Three of these patients with abnormal HM results explaining syncope had already been diagnosed with previous ECG. Hence, overall diagnostic value of the Holter examination was low as 2.4%. In contrast, diagnostic value of HM results in patients with positive family history was found as 16.6%. In our study, 7 patients considered as long QT syndrome according to HM findings although the ECG revealed normal QT measurements.

Conclusions: Detailed history have a great value in children with syncope. Holter monitoring was determined to be unnecessary in patients without high risk and its diagnostic value was considered as low among these patients while it could be used for concealed long QT syndrome especially if the syncope was associated with positive family history and exercise related syncope even with normal basal ECG findings.

P-74

Right ventricular affection assessed by tissue Doppler in pediatric dilated cardiomyopathy patients

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Dilated cardiomyopathy is not purely a disease of the left ventricle. Tissue Doppler imaging plays an essential role in evaluation of left and right ventricular functions. Aim of the work: we aimed to assess right ventricular systolic and diastolic functions in patients with dilated cardiomyopathy and to detect how much the right ventricular function impairment is correlated with the left ventricular function using tissue Doppler echocardiography.

Methods: Cross sectional analytical study was conducted on 30 cases with dilated cardiomyopathy their ages ranging from 2 months to 12 years with median age of 2.2 years in addition to 30 age and sex matched controls. Both cases and controls were subjected to tissue Doppler echocardiography. LV dimensions were measured from M-mode and LV systolic function was calculated. Right ventricular function was assessed by Doppler tissue S', E' and A' waves; RVMPI; TAPSE and RVFAC.

Results: Right ventricular systolic and diastolic functions in dilated cardiomyopathy were significantly impaired. Tricuspid S', E' and TAPSE were significantly reduced ($p < 0.05$). Tricuspid S' and E' waves were decreased significantly with decreasing LV FS ($r=0.518$, $r=0.481$) respectively.

Conclusion: Right ventricular function is definitely impaired in patients with dilated cardiomyopathy and this function impairment is correlated to the severity of left ventricular dysfunction.

P-75

Antithrombotic therapy in Kawasaki disease patients with giant coronary aneurysm

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Objective: The antithrombotic therapy in Kawasaki disease patients with giant coronary aneurysm (GA) has not been established. The aim of this retrospective study was to determine the outcome of patients with GA treated with antiplatelet therapy and anticoagulant therapy.

Method: Subjects of this study were 16 patients with GA from 1999 to 2013 in Chiba university hospital and Tokyo Women's Medical University Yachiyo medical center. We compared patients in whom cardiovascular events such as myocardial infarction (MI), angina (AP), and silent myocardial ischemia (SMI) occurred (E group, $n=7$), and patients in whom such events did not occur (non-E group, $n=9$). We started the antiplatelet therapy and anticoagulant therapy, using intravenous heparin administration, when the coronary aneurysm grew up to 4 mm. When patients got afebrile, C-reactive protein got negative and extension of GA was settled, we changed heparin into oral warfarin (Wa) administration. We took caseation oral Wa and using only antiplatelet therapy into consideration, when cardiovascular events did not occur in 2–3 years after the onset of GA.

Result: The mean observation period was 8.6 years (0.75 to 13.8). Target PT-INR was 2.0–2.5 or target TT was 15–30%. There was no difference in the day of starting initial treatment, onset of coronary aneurysms development, duration of fever, and observation periods between these groups. Total events in E group was 8. Five patients of these had MI and one patient with MI died. No patient had AP. Three patients had SMI. Three events occurred during heparin administration in acute phase; three events occurred during antiplatelet therapy with Wa administration, two events occurred during antiplatelet therapy without Wa administration. All five patients with GA in single-vessel disease had no cardiovascular event. Four of eight patients with GA in double-vessel disease had cardiovascular events. On the other hand, all three patients with GA in triple-vessel disease had cardiovascular events.

Conclusion: The prognosis of patients in this study is unsatisfactory, especially in patients with GAs in triple-vessel disease. It might be necessary to undergo antithrombotic therapy longer for patients with GAs in triple-vessel disease.

P-76

Postoperative Complications after Mechanical Valve Replacement in Children

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Objectives: The purpose of this study is to elucidate the postoperative complications after mechanical valve replacement in children.

Patients and Methods: This study enrolled 38 patients who underwent aortic valve replacement (AVR) or systemic atrioventricular valve replacement (AVVR). 16 were diagnosed as functional single ventricle, and the other 22 patients had two ventricles. The mechanical valve replacements were performed from January 1993 to December 2014. 43 replacements were done in 38 patients; 10 AVRs and 33 AVVRs. A retrospective chart review for each patient was performed.

Results: 11 complications had occurred in 43 mechanical valve replacements (25.6%). 6 prosthetic valve thromboses, 3 cerebral infarctions, 1 intracranial hemorrhage, and 1 ovarian hemorrhage. There is no coexisting or recurrent case. Table presents the patients' characteristics and the details of complications. 83% of the valve thromboses occurred within 6 months after the valve replacement. In the patients who were free from the complications, the average value of PT-INR was 2.54 ± 1.05 during 450 days after operation. On the other hand, the average value of PT-INR was 1.99 ± 0.59 during 45 days prior to the thrombotic complications (valve thrombosis and cerebral infarction), and it is significantly lower than that in patients without the complications ($p = 0.036$).

Conclusions: Valve thrombosis was the most frequent postoperative complication after mechanical valve replacement in children. 83% of the valve thromboses occurred within 6 months after valve

replacement. In the patients who were free from complications, the average value of PT-INR was 2.54 ± 1.05 .

P-77

Towards a proposal for a universal diagnostic definition of protein-losing enteropathy in Fontan patients

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Objectives: The definition of protein-losing enteropathy (PLE) in Fontan patients is variable and lacks standardization. A universal definition of PLE would significantly contribute to better comparison of treatment options and outcome, thus supporting essential clinical research in this critical area. The present study sought 1) to determine whether a clear definition of PLE in Fontan patients is routinely used, and 2) to identify useful diagnostic building blocks for composing a uniform PLE definition.

Methods: A systematic search of Medline (PubMed) was performed. Two reviewers independently screened titles and abstracts, and then evaluated full-text versions of all articles deemed potentially relevant. Clinical studies, written in English and comprising 4 or more Fontan with PLE were eligible for inclusion. PLE definitions were quantitatively analysed using the so-called 'building block approach', in which definitions were fractionated in constituent pieces of diagnostic information.

Results: We identified 363 papers. In the final analysis, data from 55 published articles were extracted. A definition of PLE was used in only 24/55 (43.6%) of the studies. PLE definitions were very heterogeneous. We identified 6 different diagnostic building blocks: (1) hypoalbuminemia ($n = 22$ studies, 91.7%), (2) hypoproteinemia ($n = 9$, 37.5%), (3) clinical presentation ($n = 17$, 70.8%), (4) documentation of enteric protein-loss ($n = 14$, 58.3%), (5) exclusion of other causes of hypoproteinemia ($n = 15$, 62.5%), and (6) hypoinmunoglobulinemia ($n = 1$, 4.2%). Most studies used 3 diagnostic building blocks (range 1 – 5) to compose a PLE definition ($n = 13/24$, 54.2%). Cut-off values for laboratory parameters (serum albumin, protein, or fecal alpha-1-antitrypsin) were frequently incorporated in the PLE definition ($n = 15$, 62.5%).

Conclusions: Our study emphasizes the need for a uniform and consequent use of a PLE definition in clinical studies concerning Fontan patients. Establishment of a universal diagnostic PLE definition is urgently needed, and the proposed building blocks may help constitute such a clinically useful definition.

P-78

Evaluation of the erythrocyte mechanical properties in children with bicuspid aortic valve

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Objective: Bicuspid aortic valve (BAV) is the most common congenital cardiac malformation, associated with significant aortic pathology and accounting for considerable morbidity and mortality. Aortic wall abnormalities associated with bicuspid aortic valve are due to cystic medial necrosis. Extensive loss of elastic fibers in the tunica media caused by increased metalloproteinase

Table.

age at operation	sex	valve type	complications	days after surgery	minimum PT-INR during 45 days prior to the thrombotic complication	PT-INR at the day of hemorrhagic complication
2Y4M	F	AVV	valve thrombosis	6	(-)* ¹	
8M	M	AVV	valve thrombosis	9	1.23	
2Y1M	M	AVV	valve thrombosis	38	6.99 ^{*2}	
12M	M	AVV	valve thrombosis	162	1.60	
2Y6M	M	AVV	valve thrombosis	171	1.38	
10Y7M	M	AV	valve thrombosis	2304	(-)* ³	
11Y2M	M	AV	cerebral infarction	237	(-)* ³	
3Y9M	F	AVV	cerebral infarction	408	1.53	
3Y0M	F	AVV	cerebral infarction	2920	1.72	
3Y6M	M	AVV	intracranial hemorrhage	449		(-)* ⁴
3Y2M	F	AVV	ovarian hemorrhage	4245		2.77

AV aortic valve, AVV systemic atrioventricular valve

AV aortic valve, AVV systemic atrioventricular valve

*1Heparin instead of warfarin was administered because of just after surgery.

*2Severe dehydration due to gastroenteritis was considered to be the cause of valve thrombosis and high PT-INR.

*3No anticoagulant agent was administered. The patient was prescribed only an antiplatelet agent.

*4Heparin instead of warfarin was administered due to severe systemic complications not associated with a prosthetic valve.

activity and cystic medial necrosis is thought to play a role in the pathogenesis of associated aortic wall abnormalities despite its clinical relevance; the pathogenesis of BAV is not clearly defined. The aim of this study was investigate the alterations in red blood cell (RBC) deformability in this disease and possible relationship between RBC deformability and BAV.

Methods: In this cross-sectional study, we evaluated 30 children with normally functioning or mildly regurgitate BAV and 27 healthy children as controls. RBC deformability was measured by laser diffraction analysis using an ektacytometer.

Results: RBC deformability was determined in 0.3 Pa, 0.53 Pa, 0.95 Pa, 1.69 Pa, 3 Pa, 5.33 Pa, 9.49 Pa, 16.87 Pa and 30 Pa. It was reduced in all shear stresses except 0.3 Pa at the BAV patients compared to healthy controls, the differences were statistically significant for all shear stresses except 0.3 Pa, 0.53 Pa and 30 Pa ($p < 0.05$).

Conclusion: RBC deformability plays an important role in blood circulation; facilitates flow of 8 μm -diameter erythrocyte through 2-3 μm -diameter capillaries. Although accumulating data in literature, shows alterations of RBC deformability in diseases such as vasculitis, hypertension, peripheral and coronary artery diseases, no study has yet evaluated possible changes of erythrocyte deformability in BAV disease. RBC deformability was decreased at the BAV patients compared to control group in our study. Our results emphasize the association between RBC deformability and BAV disease, and to our knowledge; this is the first study in the literature. Further pathophysiological studies included the peripheral vessels an addition the aortic wall are warranted to better clarify this issue in BAV disease.

P-79

The features of the toxic metals content in malformation locus of the heart tissues of children with CHD

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Objectives: Comparative analysis of toxic metals content in normal heart tissue and in malformation's locus of children with congenital heart diseases (CHD).

Methods: We had determined content of toxic in 107 biosubstrates of heart and great vessels of children with CHD (n = 55), 82 sample (76.6%) presented by intraoperative biopsates. We have selected 38 children, which had biopsies of the affected (locus malformation) and unaffected areas of the heart. For comparative analysis was used Wilcoxon T-test. All patients were examined by the spectral analysis of Al, Cd, Pb, Hg, Be, Ba, Tl, Bi, As, Ni, Sb, Sn, Sr, Ti, W, Zr, Ag, Li, B, Co, Si, V by methods of the atomic emission spectrometry in the inductively coupled plasma and atomic absorption spectrometry with electrothermal atomization.

Results: We revealed the presence of a wide range of toxic metals, a total of 10 in different areas of the heart and great vessels tissues, including malformation locus in children with CHD. The average concentration of toxic barium, lithium, nickel and arsenic in both investigated hearts areas had exceeded standard rates. The concentration level of toxic metals aluminum ($p = 0,011$), nickel ($p < 0,001$), barium, strontium, lead, arsenic and titanium was higher in malformation locus rate than in other parts of the heart.

Conclusions: The findings indicate about feature of the content of toxic metals in a locus malformations of the heart with a congenital defect and suggest about possible role of aluminum, nickel, barium, strontium, lead, arsenic and titanium in cardiogenesis violation in humans.

P-80

Evaluation of the clinical courses of patients who were diagnosed with acute rheumatic fever in children

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Introduction: Rheumatic heart disease (RHD) is an important public health issue, particularly in the developing country, but its true progress is unknown in our region. The purpose of this study is to evaluate the clinical progress of patients who were diagnosed with acute rheumatic fever (ARF) and RHD.

Methods: Medical records of patients who were diagnosed as having ARF in our clinic were evaluated retrospectively and patients who had at least 2-year follow-up after the diagnosis were included into the study. Diagnosis of ARF was done by using revised Jones criteria. Acute and chronic phase valve involvements are compared.

Results: A total of 163 children formed the study group. The follow up period ranged from 2-11 years. During the initial attack 88.3% of patients had carditis: mild in 86.8%, moderate to severe in 13.8%. Among patients with moderate-severe carditis, moderate to severe valve insufficiency was detected in at least one valve. Most common valve involvement was mitral insufficiency (n = 135, 93.8%).

At last visit rheumatic heart disease (RHD) was present in 135 valves of 102 cases; 92 mitral, 42 aortic. No valvular regurgitation was detected in 55.9% of patients with silent carditis, and in 20.9% with clinical carditis. In patients with mild, moderate and severe valvular regurgitation full recovery rates at last visit were 56.7%, 20.6% and 5.5%, respectively. Recurrence rate was 2.1% among patients who were well adapted to secondary prophylaxis, whereas the rate was 94.4% who were not well adopted.

Conclusions: In ARF patients, recovery rate of valve insufficiency is high if it is mild. The increased risk of progressing to severe chronic valvar disease was associated with moderate or severe carditis and recurrences of ARF. Regular and appropriate prophylaxis is important to prevent the recurrence of the disease.

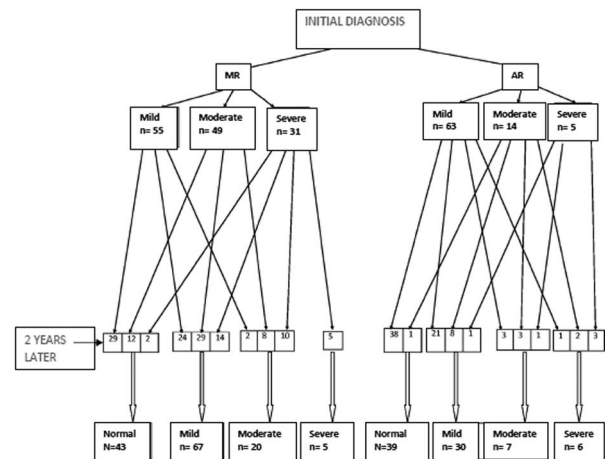


Figure.

P-81

Evaluation of cardiac function with tissue Doppler echocardiography in normotansive offspring of hypertensive parents

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Introduction: Early alterations in the cardiovascular system have been described in the normotensive children of hypertensive parents (NCHP). We aimed to identify left ventricular structural and functional changes in NCHP for predicting hypertension that may be developed in the future.

Methods: Ninety-two children who had history of parental hypertension and 90 children whose parents were normotensive were included to the study. The left ventricular structure and function were evaluated by tissue Doppler imaging and conventional echocardiography methods such as M-mode and pulsed wave Doppler for this purpose in two groups.

Results: There were no statistical difference between gender, age, weight, height and body mass index in two groups ($p > 0.05$). The mean systolic and diastolic blood pressure values were higher in the NCHP group ($p = 0.003$, $p = 0.001$, $p < 0.001$ respectively). Interventricular septum and left ventricular posterior wall thickness and relative wall thickness (RWT) were higher in NCHP group ($p = 0.029$, $p = 0.016$, $p = 0.041$, respectively). Septal and lateral mitral annular isovolumetric relaxation time (IRT) and myocardial performances index (MPI) with TDI were higher ($p < 0.001$, $p < 0.001$, $p = 0.001$, $p = 0.001$, respectively), ejection time was shorter ($p = 0.015$, $p < 0.02$), septal isovolumetric contraction time (IVCT) determined longer in NCHP group ($p < 0.002$). There was positive correlation between lateral and septal MPI with systolic blood pressure and a negative correlation with ejection fraction ($p = 0.042$, $p = 0.025$, $p = 0.032$, $p = 0.044$ respectively). There was no difference between septal and lateral MPI measures for detecting ventricular dysfunction in normotensive children of hypertensive parents ($p > 0.05$).

Conclusions: Systolic and diastolic dysfunction that accompanied by morphological changes in left ventricle were shown by TDI in normotensive children of hypertensive parents, even if there are not signs of clinical hypertension.

P-82

Anomalous origin of the left coronary artery from the pulmonary artery - retrospective study of 23 years of experience

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Background: Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) causes severe myocardial ischemia, global left ventricular dysfunction, and varying degrees of mitral regurgitation. Surgical strategy to construct a two-coronary system for a patient with ALCAPA has evolved with time but many questions remain unanswered.

Methods: We designed a retrospective, longitudinal, descriptive study that included patients with ALCAPA. We describe perioperative details such as clinical manifestations, variations in the surgical technique and the postoperative morbidity and mortality.

Results: Eighteen children underwent surgical reconstruction of a two-coronary system because of ALCAPA between 1991 and July 2014. Two patients were lost to follow-up, one early, one in recent years. Heart failure was the principal cause of hospitalization in 14/18 of our patients. Left ventricular (LV) dysfunction was present in 14/18 and 12/14 had moderate or severe mitral regurgitation (MR). Surgery was performed with direct coronary reimplantation in 14/17 patients and intrapulmonary tunnel (Takeuchi repair) in 3/17. The most common immediate postoperative complications were: low cardiac output (10/13), pleural effusion (3/13). There was one early postoperative death (30 days) due to heart failure and ventricular arrhythmia. Mean follow-up (16 patients) was 12 years and 4 months (5 months–23 years). There

was 1 late death at the age of 21 caused by VF. LV function and MR significantly improved during follow-up in all surviving patients. Global LV function by echocardiography was 66% (55–81%, Teichholz). Six patients had minor regional hypokinesis which was related to the presence of myocardial scar confirmed by perfusion scintigraphy and/or magnetic resonance imaging. Moderate MR was present in one patient, severe in none.

Conclusions: Long-term prognosis after surgical repair of ALCAPA is unclear. Standard echocardiography may underestimate LV scars and perfusion deficits. Lifelong surveillance of these patients, including magnetic resonance imaging, is recommended.

P-83

Pulmonary artery sling: associated anomalies, surgical management and follow-up. A single center experience

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Introduction: Left Pulmonary Artery Sling (LPAS) can be associated with other cardiac abnormalities and tracheal compression and/or stenosis. We describe our experience in the management of this condition.

Methods: In our centre between April 1996 and September 2014, 11 infants (5 male, 6 female) underwent surgical repair of LPAS. Mean age at the time of surgery was 14 months (range 22 days to 5 years and 9 m). In 5 patients LPAS coexisted with other cardiac abnormalities, 3 cases underwent an initial cardiac operation, without sling correction, in one case for critical neonatal COA, in the other two cases for misdiagnosis. The presence of LPAS was suspected by clinical presentation and features on chest X-ray and echocardiography. Confirmation of LPAS was established using angiography (6 patients) or CT (5 patients). Tracheal compression was confirmed by bronchoscopy, in 7 patients we found a tracheal stenosis with complete rings in 5 of them, tracheomalacia in 9 patients. Five patients required surgical LPA reimplantation with slide tracheoplasty (ST) (45%), four patients isolated LPA reimplantation (36%), 2 patients LPA reimplantation with repair of intracardiac anomalies (18%).

Results: Two deaths occurred in the first 30 days after surgery, one of them after ST. Another patient requiring ECMO after ST, is still followed-up under ventilator after two tracheal balloon dilations. In the 8 long-term survivors, LPA patency was assessed using echocardiography, angiography, CT or MR; only 1 patient required a balloon angioplasty (PTBA). Bronchoscopy was performed in all patients who underwent a ST, and 1 \geq balloon tracheal dilation was necessary in 3 patients.

Conclusions: LPAS has a low operative mortality and excellent long-term patency of LPA. The simultaneous presence of other cardiac abnormalities and/or tracheal stenosis complicates the short- and long-term prognosis. In our center we adopt a multidisciplinary approach. The role of imaging is crucial: in the diagnostic phase the gold standard is Bronchoscopy and CT. A Bronchoscopy is performed at 1, 3, 6 and 12 months post-surgical correction, within cardiology follow up. If tracheal stenosis or pulmonary artery obstruction are suspected CT or MR are performed. Angiography is performed in presence of severe pulmonary artery obstruction.

P-84

Cardiac Output Measurement using the CO₂-rebreathing Technique during Maximal Exercise Test in Children with Fontan Circulation

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Introduction: Exercise performance is decreased in children with univentricular heart (UVH) after Fontan completion. The mechanism of exercise intolerance after Fontan is complex. Only few studies investigate the mechanism of decreased exercise, no studies highlight the pump function at maximal exercise. Cardiac output (CO) and stroke volume (SV) can be measured using the CO₂-rebreathing (CO₂-R) technique during exercise test (CPET).

Methods: Patients with univentricular heart in NYHA class I who were referred for CPET between 2010–2012 underwent CO measurement at rest (CO_{rest}) and within 10 seconds after peak exercise (CO_{max}). Data were compared with children who performed a normal exercise test. Stroke volume at rest and at maximal exercise was calculated. SV_{diff} and CO_{diff} was defined as SV_{max} and CO_{max} minus SV_{rest} and CO_{rest}. Data were expressed as mean ± standard deviation.

Results: 19 children with UVH (11,7 ± 3,3 yrs, 38,9 ± 9,9 kg) were compared with 38 controls (11,24 yr ± 2,2, 39,6 k ± 10 kg). HR max (157,2 ± 29,4 vs 190 ± 11,4 bpm), VO₂ max (32,1 ± 7,8 vs 44,5 ± 9,7 ml/min), test duration (9,9 ± 3,6 vs 13,2 ± 4,8 min) and load (76,5 ± 35,7 vs 105 ± 41,8 Watt) were statistically different. Respiratory exchange ratio reached 1 in both groups. 12 patients successfully underwent CO measurement. The reason for CO-R failure was technical (4) or non-cooperation (3). CO_{rest} (3,6 ± 1,0 vs 5,82 ± 2,4 l/min), CO_{max} (8,8 ± 2,7 vs 13,1 ± 5,1 l/min) (P < 0,001) and stroke volume (SV) at rest (43,7 ± 13,7 vs 62,8 ± 29,2 ml/beat) and max (71,9 ± 18,6 vs 96,5 ± 28,7 ml/beat) (P < 0,05) were different. SV_{diff} (28,2 ± 14,9 ml/beat vs 34,0 ± 14,1) and CO_{diff} (4,3 ± 2,3 l/min vs 5,0 l/min) were not significantly different.

Conclusion: Children with UVH have lower maximal exercise performance due to a combination of chronotropic incompetence and decreased stroke volume, leading to lower cardiac output. Stroke volume and cardiac output are decreased at rest (resp 30% and 40% lower) and during maximal exercise (resp 25% and 33% lower) compared to the control group. Although there also is a tendency towards less increase in SV_{diff} and CO_{diff}, the difference of CO_{rest} and SV_{rest} and the chronotropic incompetence are more important to declare decreased CO_{max} and SV_{max}. These findings demonstrate that even at rest patients with Fontan circulation have an important restricted pump function, worsening more at maximal exercise.

P-85

Severe congenital heart disease: mortality and impact of prenatal diagnosis

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Objectives: Describe mortality rate of patients diagnosed with severe congenital heart disease (CHD) in the prenatal and postnatal setting, compare prenatal and postnatal overall mortality and evaluate the impact of associated pathology.

Methods: Retrospective database research of 542 patients with severe CHD, diagnosed and/or treated with CHD at the University Hospital of Ghent (pre- and postnatal) or Antwerp (prenatal) between 01/01/2006 and 31/08/2014. CHD was divided in 11 pathologies: coarctation (CoAo, N124), tetralogy of Fallot (TOF, N118), univentricular heart defect (UVHD, N105), transposition of the great arteries (TGV, N74), atrioventricular defect (AVSD, N69), isomerism (N15), truncus arteriosus (N11), pulmonary atresia with intact interventricular septum (N10), double outlet with transposition (N9), Ebstein anomaly (N4) and double discordance (N3).

Results: Overall, CoAo was most frequent (23%), followed by TOF (22%) and UVHD (19%).

In 162 of 542 patients (30%), diagnosis was made prenatally. In the prenatally diagnosed group, UVHD accounted for 45%, TOF for 16% and TGA for 10%. Termination of pregnancy (TOP) was carried out in 43% of prenatal diagnoses, 67% being for UVHD. In the UVHD group, 11% of the fetuses had an associated genetic or structural abnormality, in contrast to 100% of fetuses with AVSD (6% of TOP).

Of all patients born alive (N = 477), 87 had a prenatal diagnosis. The overall survival was 81% with higher survival rates for patients with TGA (71/74), CoAo (111/119) and AVSD (57/64). The overall mortality of patients born alive was 19%. Compassionate care was given in 25 patients (29% of postnatal deaths), of which 11 had TOF with significant additional pathology. Spontaneous pre-operative death was noted in 10 newborns. 432 patients underwent interventional treatment with an overall survival of 88%.

Conclusion: The overall survival after diagnosis of a severe CHD is 70%, with important differences between pathology groups. Despite organised prenatal screening, only 1/3 diagnoses is made prenatally. Prenatal diagnosis is associated with a high mortality due to the high incidence of TOP in our centre. Postnatal mortality is 19% with a high attribution of postnatal compassionate care, spontaneous pre-operative demise and late death related to non-cardiac causes.

P-86

Heart rate variability is related to disease severity in children with pulmonary hypertension

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Background: Pulmonary hypertension (PH) is characterized by progressive pulmonary vascular remodeling and consecutive elevation of pulmonary vascular resistance and pressure that may result in chronic right heart failure, which is associated with an increase in sympathetic tone. This may adversely affect cardiac autonomic control. We aimed to assess whether parameters of heart rate variability (HRV) are related to disease severity in children with pulmonary hypertension.

Methods: Parameters of HRV [SDNN = standard deviation of normal-to-normal intervals, SDANN = standard deviation of mean values for normal-to-normal intervals over 5 min, rMSSD = square root of the mean square differences of successive RR intervals, and pNN50 = proportion of the number of pairs of successive normal-to-normal intervals that differ by more than 50 ms divided by total number of normal-to-normal interval] were determined from Holter electrocardiograms of 17 patients with PH (10 female, mean age 12.8 ± 8.7 years). 13 of the 17 patients had idiopathic PAH, 3 patients had associated chronic lung disease, one had recurrent pulmonary vein stenosis. An additional group of 5 adolescents with Eisenmenger syndrome (EMS) was included.

Results: Patients were allocated to 2 groups according to their disease severity: (1) Patients with severe PH (ratio of PAP/SAP > 0.75) (n = 6), (2) patients with mild PH (PAP/SAP ratio < 0.75) (n = 11). Children of group 1 had significantly lower values of HRV [SDNN (73.8 ± 21.1 vs. 164.9 ± 38.1), SDANN (62.2 ± 19.0 vs. 139.5 ± 33.3), rMSSD (31.0 ± 8.7 vs. 73.6 ± 22.7), and pNN50 (5.8 ± 3.4 vs. 28.0 ± 8.8)] compared to group 2 (p < 0.0001 for all). SDNN inversely correlated with ratio of PAP/SAP (r = -0.838; p < 0.001). EMS patients showed no significant

difference of HRV [SDNN 157.6 ± 43.2 , SDANN 141.2 ± 45.3 , rMSSD 66.8 ± 16 , and pNN50 18 ± 11.6] compared to patients of group 2 ($p > 0.05$ for all).

Conclusions: According to our results, children with severe PH may have alterations in HRV. Since HRV appears to be related to disease severity, it may therefore serve as an additional diagnostic marker of PH. Remarkably, although EMS patients have suprasystemic pulmonary arterial pressures, they seem to have preserved HRV, which might reflect a more favourable autonomic adaptation.

P-87

Long term follow up of bidirectional cavopulmonary anastomosis patients: Multi-institutional study

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Background: The bidirectional cavopulmonary shunt is a step towards the Fontan operation in palliation of patients with single-ventricle heart. The Fontan procedure is performed with children between three and five years of age. In Egypt, there is a delayed age of children undergoing Fontan procedure due to long waiting lists and budget restraints on pediatric cardiac surgeries.

Aim of the work: To assess long term follow up of patients with bidirectional cavopulmonary shunt and to determine the clinical and hemodynamic effects of delayed Fontan procedure.

Methods: A total of 125 patients with bidirectional cavopulmonary shunts from three centers: Cairo university, Ain Shams university and Egypt kids hospital were followed up from January 2012 till July 2014. At follow up, mean age was 7.5 (1.7-16 years) and weight was 23.05 (9-58 kg). Males were 61.5%, 64% had single ventricle morphology, 76% had additional pulmonary flow and 38.5% had prior palliative cardiac surgery.

Results: The mean age at Cavopulmonary shunt was 2.98 years (0.4 – 12 years). NYHA was class I in 82% and II in 15%, mean oxygen saturation was 81% (60-95). Cardiac catheterization was done in 45%, mean Glenn pressure was 15.5 (6-28 mmHg) and mean Nakata index was 287 (108-910). Catheter interventions were done in 15%; seven patients had procedures to minimize the extrapulmonary flow: closure of the shunts, collaterals, patent ductus arteriosus and/or device occlusion of the forward flow, while eight patients had balloon and/or stent of pulmonary arteries, superior vena cava (SVC) or pulmonary veins. Three patients had occlusion of venoatrial collaterals and/or left SVC to coronary sinus. The only significant difference was intensive care unit (ICU) stay which was shorter in patients with additional pulmonary flow (P value 0.016), and mean Glenn pressure which was lower in patients with single ventricle (P value less than 0.001). Mean survival was 11.33 years with 4% late mortality.

Conclusion: Cavopulmonary shunt can be a long term palliative procedure for patients with single ventricle morphology in developing countries when Fontan operation cannot be done on time due to economic restraints.

P-88

Functional outcomes after the Ross procedure

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Introduction: Compared to mechanical and biological aortic valve replacement, Ross patients do not require anticoagulation and the pulmonary autograft has potential for growth in the aortic position. Despite these advantages, the Ross procedure remains controversial mainly due the potential risk of inducing bivalvar disease. Little is known about functional outcome in children and adults after Ross procedure.

Methods: Between January 1999 and December 2010, our single-center results for the Ross procedure with analysis of morbidity, mortality, health-related quality of life and actual exercise capacity at midterm follow-up were performed.

Results: Thirty-five patients (70% male) underwent the Ross procedure at a mean age of 13.5 years (0.1-49.2 years, 70% of patients were under 18 years). Indication for surgery was combined aortic stenosis and regurgitation in 53% of patients. Three patients died during follow-up and 5 patients required re-operation during the midterm follow-up of 6.0 ± 3.2 years. Objective exercise capacity measured by peak oxygen uptake (VO₂ max) was reduced to 32.7 ± 8.3 ($z = -1.3 \pm 1.1$) in patients operated during childhood and adolescence, and to 28.6 ± 8 ml/kg/min ($z = -2.4 \pm 0.8$) in patients operated in adulthood. Quality of life in children measured by KINDL-R questionnaire was equal compared to healthy standard population (total score: 76.8 ± 13 vs 76.3 ± 10.1 ; $p = 0.90$). In adults patients after ROSS procedure health-related quality of life measured by SF-36 was equal or even better than in healthy standard population (physical health score 54.7 ± 4.1 vs 48.4 ± 9.4 ; $p = 0.005$, mental health score 48.4 ± 15.1 vs 50.9 ± 8.8 ; ns).

Conclusion: Despite reduced exercise capacity at mid-term after a Ross operation, quality of life remains good when the procedure is performed in child- or young adulthood.

P-89

Clinicopathological investigation on children with dysrhythmias using endomyocardial biopsy-with special reference to histopathology between tachycardia induced cardiomyopathy and bradyarrhythmia related cardiomyopathy-

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Purpose: Tachycardia-induced cardiomyopathy (TIC) has gained increasing awareness and attention, however recently not only TIC but also bradyarrhythmia related cardiomyopathy has been recognized. WPW syndrome with dilated cardiomyopathy (DCM) in the absence of tachyarrhythmias is also recognized. The incidence of latent DCM state among dysrhythmia patients is unclear. Accurate diagnosis of TIC and bradyarrhythmia related cardiomyopathy is clinically important as these cardiomyopathy can develop severe ventricular dysfunction or DCM. This study examines the demographic, clinical and histopathological features of dysrhythmia related cardiomyopathy in childhood with special reference to its subtype of dysrhythmias. Clinical symptoms were compared with baseline ECG, 2DE, EPS, MRI and endomyocardial biopsy (EMB) with light microscope as well as ultrastructural characteristics.

Patients and Method: Between 1990 and 2012, a total of 77 patients with dysrhythmias were enrolled, including 56 tachyarrhythmias; 35 SVT, 2 AFL, 19 VT and 21 patients with bradyarrhythmias; 7 idiopathic congenital complete heart block (CCHB),

4 myocarditis, 1 hypertrophic cardiomyopathy (HCM). Histopathology was evaluated with semiquantitative morphometry.

Results: DCM was found 9 in tachyarrhythmias and 3 in bradyarrhythmias. Ablation was performed 29 for SVT and 5 for VT, 3 pacemaker implantation for CCHB and 1 ICD for HCM. Histopathology on EMB showed abnormalities of inflammatory cell infiltration, disarray of myocytes, vacuolar degeneration, lysis of myofibrils and higher % fibrosis in long-term dysrhythmia patients. There were no significant differences in histopathological abnormalities between both dysrhythmias, although severe myocardial damages was noticed in long-standing dysrhythmias and LBBB morphology bradyarrhythmias.

Discussion and Conclusions: Although the incidence of TIC and bradyarrhythmia related cardiomyopathy has been reported not so frequently, some cases were present among dysrhythmia patients in our study. Histopathology showed various abnormalities but not specific. Persistent or recurrent tachyarrhythmias, myocarditis and dyssynchronous ventricular contraction could be the cause of left ventricular dysfunction, leading to dysrhythmia related cardiomyopathy. Abnormal accessory pathway also may include LV dyssynchrony, leading to adverse remodeling and ventricular dysfunction. The clinical spectrum of this complex pathology is highly varied. EMB may still be helpful to determine etiology in undiagnosed cardiomyopathy. Early diagnosis of dysrhythmia related cardiomyopathy enables the start of effective treatment with the purpose of better outcomes in this population.

P-90

Evaluation of 2283 patients with isolated ventricular septal defect by echocardiography up to 26 years from a single center

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Spontaneous closure of ventricular septal defects has been reported. While ventricular septal aneurysm is an important mechanism of closure, it was observed that aneurysmal transformation may be particularly likely to develop left ventricular to right atrial shunt and subaortic ridge. Here we present the medical records of 2283 patients with ventricular septal defect to assess the rate of spontaneous closure, aneurysmal transformation, left ventricular to right atrial shunt, subaortic ridge, aortic valve prolapsed and aortic regurgitation.

Materials and Methods: The study population consisted of 2283 patients with isolated ventricular septal defect who had been studied at our institution from 1988 to 2014 with transthoracic echocardiography. Ventricular septal defects were classified according to their location and relation to the tricuspid annulus and semilunar valves. Defect size was expressed in terms of the size of the aortic root. Defects approximating the size of the aorta were classified as large, defects one-third to two-thirds the diameter of the aortic root were classified as moderate and defects less than one third the diameter of the aortic root were classified as small. SPSS17 was used for statistical analysis.

Results: Ventricular septal defects were perimembranous in 67.7% (1530), trabecular muscular in 24% (548), muscular outlet in 4.9% (112), muscular inlet 3.1% (69) and doubly committed subarterial in 0.8% (19) and Swiss cheese in 0.2% (4). Defect size was classified in 65.7% (1499) as small, in 17.8 (407) as moderate and 16.5 (377) as large. The percent of spontaneous closure in perimembranous ventricular septal defects was 13.1% and 30.9% in muscular ventricular septal defects. While aneurysmal transformation was detected 44.2% in the patients with perimembranous ventricular septal defect, aortic valve prolapse was the most common complication (14.1%) observed during the follow ups. Left ventricular

to right atrial shunt, subaortic ridge and aortic regurgitation were detected with the percentage of 12.3%, 3.4% and 9.2% respectively in patients with perimembranous defects.

Discussion: The rate of spontaneous closure in muscular defects was higher than the rate of spontaneous closure in perimembranous defects.

P-91

Exercise Blood Pressure Response In Children After Aortic Coarctation Repair

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Objective: Despite successful repair of aortic coarctation, systemic hypertension can persist in a significant percentage of patient. The aim of this study was to assess blood pressure in children operated for aortic coarctation, both at rest and after exercise.

Material-Method: Nineteen children were operated upon for aortic coarctation. The patients data were compared with those obtained from 19 healthy controls of the same age. Demographic and clinical data including age at intervention, blood pressure at rest and on exercise, transthoracic echocardiography and treadmill exercise test results were evaluated. Results are compared statistically.

Results: Of the operated aortic coarctation patients 7(47%) were female, 12 (63%) were male, of the control group 7 (47%) were female, 12 (63%) were male. Mean age of patients were $12,2 \pm 4,6$ years and control group was $12,0 \pm 2,0$ years. Both groups were compared for weight, height, arterial tension, heart rate and echocardiographic measurements, 24 hours blood pressure and on exercise pressure. Nineteen patients performed a treadmill exercise test. (Bruce protocol). The mean duration of exercise was $8,1 \pm 2,3$ minutes, mean peak heart rate was 154 ± 22 beats per minute and mean systolic pressure 140 ± 26 . Exercise times were found statistically shorter in patient group than control group ($p < 0,05$). Fourteen (73%) patients had a hypertensive response during test, among whom only five (26%) had uncontrolled blood pressure at rest. Age at surgery and type of aortic coarctation repair were not associated with a hypertensive response on exercise ($p < 0,05$).

Conclusion: Operated aortic coarctation patient have an alarming prevalence of hypertension. Due to abnormal blood pressure homeostasis, hypertension should be aggressively pursued by ambulatory blood pressure measure assessment and exercise stress testing in this population. In this study we found a significant prevalence of exercise induced hypertension in patients after successful aortic coarctation repair despite adequate blood pressure control at rest. Exercise induced hypertension was significantly related to higher peak gradient in the descending aorta and treatment with angiotensin receptor inhibitor. These results highlight the complexity of the aortic coarctation population and show that, even after a good surgical result, several patients remain at high cardiovascular risk and require long term follow up.

Key words: aortic coarctation, exercise induce hypertension, children

P-92

Determining Arterial Functions In Children Who Got Repaired Aortic Coarctation

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Objective: Despite the apparently successful surgical repair of aortic coarctation, subsequent cardiovascular complications have

sometimes been encountered. In our study, we investigate arterial functions by measuring carotis intima-media thickness, brachial artery dilatation by blood stream and distensibility of the abdominal aorta.

Material-Method: 18 patient who repaired aortic coarctation and 14 healthy children take part in our study. Patient group is chosen retrospectively. Physical examination, telecardiography, electrocardiography, echocardiography examinations. Left ventricular systolic functions, diameters of aortic anulus, asendan aorta, arcus aorta, isthmus and abdominal aorta in systole and diastole by M mode echocardiography. Aortic strain, aortic distensibility and aortic stiffness is calculated by using diameters of aortic aorta in systole and diastole. Diameter of carotis intima media and dilatation of brachial artery's stream dependent dilatation by using linear probe. Results are compared statistically.

Results: Of the operated aortic coarctation patients, 6 (33%) were female, 12 (66%) were male, of the control group 6 (42%) were female, 8 (58%) were male. Mean age of patients were $12,2 \pm 4,6$ years and control groups were $13,0 \pm 2,0$ years. Both groups were compared for weight, height, arterial tension, heart rate and echocardiographic measurements. Aortic strain and aortic distensibility were lower in patient group than control group but they are not meaningful statistically ($p > 0,05$). Aortic stiffness measurements were found statistically higher in patient group than control group ($p = 0,035$). Carotis intima-media diameters were higher in patient group but it is not meaningful statistically ($p > 0,05$). Stream mediated brachial artery dilatation in 1rd minute was statistically lower in patient group than control group ($p = 0,047$).

Conclusion: By means of parameters measured in operated aortic coarctation group, being higher aortic stiffness and lower stream mediated brachial artery dilatation, are cautionary for close follow-up for long time and taking preventive measures for hypertension.

Key words: aortic coarctation, aortic stiffness, brachial artery dilatation

P-93

Surgical vs Percutaneous closure of PDA: How is the condition in preterms < 2 kg

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Background and Aim: There is no doubt about that symptomatic PDA should be treated as soon as possible but treatment method in preterms is a highly controversial topic. For long years it was used to be known as surgical ligation is the definitive treatment. As new devices come into the market, percutaneous techniques improve and interventionalists become more experienced; percutaneous closure gets more common in preterms.

In this study we aimed to compare efficacy and safety of PDA closure surgically versus transcatheter method in preterms less than 2 kg. Best of our knowledge this study is the first one that compares outcomes of surgery and percutaneous PDA closure in preterms.

Material and Method: Between the dates July 1997 to October 2014 in our center PDA of 18 patients less than 2 kg were closed percutaneously and 29 patients less than 2 kg operated. A comparison was made between the data of the patients whose PDA were closed percutaneously (GroupA) and surgically (groupB).

Results: The median patient age was 32days, in groupA and 31days in groupB. The median weight of the patients in groupA was

1603 gr and 1288 gr in groupB. The mean PDA diameter in percutaneous group was 2.8 ± 0.91 mm and 2.95 ± 0.45 mm in surgery group.

There was no statistically significance between 2 groups in terms of age, defect size and additional heart defects. Only weight of patients in percutaneous PDA closure group was significantly more than the surgery group ($p:0.004$). Mean gestational age of the patients in groupA was 30 ± 1.8 weeks, in groupB was $28,6 \pm 3.5$ weeks. In groupA; all cases were closed successfully except one. having large window type PDA which was sent to surgery. There were no major complications reported. Left pulmonary arterial stenosis was detected in 4 patients which were all resolved in 6 months duration.

In groupB 2 major complications like: pneumomediastinum and chylothorax but no minor complications were reported. There was no statistically significance between complication and success rates between two groups.

Conclusion: Percutaneous PDA closure is the candidate for taking the place of surgery in preterms. However, not applied routinely; can only be done in fully equipped large centers by experienced interventionalists.

P-94

Prevalence of non-anterior early repolarization pattern in young teen competitive athletes in the different sports and relation with left ventricular remodeling

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Background and Objectives: Early repolarization (ERP) on the electrocardiogram (ECG) is more common among young athletes than the general population. It has been considered a benign finding. In some publications, especially non-anterior ERP has been associated with increased risk of sudden cardiac arrest. The objectives of this study were to evaluate the prevalence of ERP in the different sports, to describe whether there is any association between ERP and echocardiographic measures of left ventricle (LV) remodeling or not, and to evaluate the effect of physical training.

Methods: ERP was assessed in 140 athletes with a mean age of 13 years (range 10 to 18) from clubs for five different sports (basketball, swimming, football, wrestling, and tennis) who had practiced regularly at least 3 h per week for at least 2 years. ERP was defined as J-point elevation ≥ 0.1 mV in at least 2 leads within a non-anterior territory (inferior [II, III, aVF] or the lateral territory [I, aVL, V4-V6]).

Results: The overall prevalence of ERP in our study population was 12.1% in the inferior leads, 6.4% in the lateral leads and 5.7% both leads. Non-anterior ERP was found more common in swimmer (15.6%) and basketball player (24%) than football, tennis, and wrestling. Although the type of exercise was not associated with ERP, it was more common in the combined exercise that have dynamic and static components. Weekly training hours were not statistically different between athletes with ERP and without ERP (9.5 ± 4.6 v.s 11.1 ± 6.0 hours respectively). There were no associations between ERP and echocardiographic measures of LV remodeling and LV geometric pattern.

Conclusions: Our study confirms that the frequency of inferior ERP is very high in young athletes. Albeit the incidence is variable from one sports to the other. Further studies are required to understand these results better.

P-95

Congenital left atrial appendage aneurysm interfering with preexisting pericardial effusion in an infant with myopericarditis

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Left atrial appendage aneurysms (LAAAs) are usually congenital and are very infrequent anomalies of the heart. Differential diagnosis on echocardiography involves acquired left atrial aneurysms, extra pericardial herniation of portions of the heart, solid or cystic para-cardiac tumors and rarely, pericardial or extra-cardiac fluid collections. Here, we report a pediatric case of LAAA with concomitant myopericarditis who had a fatal course.

Case: A previously healthy, 7-month-old girl was admitted to a state hospital with complaints of cough and respiratory distress for several days. Because of cardiomegaly was noted on chest x-ray, echocardiography was performed and referred to our clinic for drainage of large pericardial effusion. At admission, she had tachypnea, tachycardia, and poor peripheral perfusion. Immediate echocardiography showed a large left para-cardiac chamber communicating with a normal-sized left atrial cavity via a wide neck, with evidence of spontaneous echo contrast (Figure). Thus, a giant LAAA was diagnosed. Pericardial effusion (12 mm) was also noticed around the left ventricle (LV). Cardiac markers were increased (BNP > 35.000 pg/ml, Troponin I: 0.79 ng/ml, Troponin T: 0.35 ng/ml) and LV function was mild-to-moderately impaired with an ejection fraction of 45%. So, she was also diagnosed as myopericarditis, and appropriate management was given. To better define the anatomical relationship of the aneurysm, a computerized tomography was performed. The aneurysm size was 39 × 29 mm and extended laterally toward the cardiac apex. Surgical resection of the aneurysm was considered. However, she had cardiac arrest after an episode of “torsades de pointes” that was unresponsive to resuscitation on the fifth day of admission.

Conclusion: Early recognition of an LAAA is important prognostically since the complications associated with this abnormality

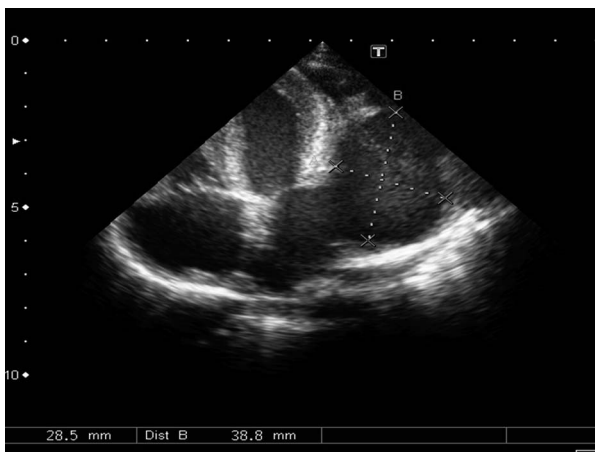


Figure. Echocardiographic view showing a giant para-cardiac chamber communicating with a normal-sized left atrial cavity via a wide neck with evidence of spontaneous echo contrast.

can be devastating. Compression of LV by LAAA may adversely affect LV systolic function. Presence of this anomaly is considered to be a predisposing factor for fulminant course of myopericarditis in our case.

P-96

Quality of life, posttraumatic stress disorder and other psychiatric problems in children with tachyarrhythmias

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Objective: In this study, the aim was to search the quality of life, posttraumatic stress disorder (PTSD) symptoms and other mental health problems in children with tachyarrhythmia; and to evaluate the variables that can affect these factors.

Methods: Thirty pediatric patients with tachyarrhythmia between the ages of 8-18 years that had been followed up at pediatric cardiology outpatient clinic were included in the study. Control group consisted of age-and-sex matched 32 healthy individuals. Socio-demographic information form, pediatric quality of life inventory (PedsLQ), state-trait anxiety inventory (STAI-TX), STAI-TX for children under 14 years of age, strength and difficulties questionnaire (SDQ), general health questionnaire (GHQ) were administered to children and the mothers in both the study and control groups. To evaluate PTSD in the study group, the children filled children's PTSD-reaction index (CPTS-RI), and the mothers filled traumatic stress symptom checklist (TSSC).

Results: The quality of life in children with tachyarrhythmia was not different from the control group. It was determined that the prosocial behavior scores of patients with tachyarrhythmia were lower than the control group, and the mothers of the patients were more anxious than the mothers in the control group. 40% of patients reported PTSD symptoms, in whom 2 (6.6%) had severe symptoms of PTSD. PTSD symptoms were correlated with age at the diagnosis, number of emergency referral, quality of life reported by children and occupational concern for child and mother. We found that 56% of the mothers of patients had symptoms accordant with possible PTSD, and PTSD symptoms were correlated with PedsLQ scores reported by children and the mothers, SDQ scores and mental health problems of mothers. PTSD symptoms of the patients and the mothers were not correlated with each other.

Conclusions: In our study, it was determined that the mothers of children with tachyarrhythmia were anxious and anxiety level of the mother were correlated with the mental health and quality of life of the patients. Also, PTSD symptoms can be seen in both children with tachyarrhythmia and their mothers. However, as tachyarrhythmia is a paroxysmal disorder, long-term follow-up studies are needed in these children.

P-97

Heart-Type Fatty Acid Binding Protein (HFABP), Creatine Kinase Myocardial band (CKMB) and Cardiac Troponin I (cTnI) Levels Before and After Pediatric Cardiac Catheterization

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Introduction: The aim of the present study was to evaluate the presence of cardiac injury using biomarkers during cardiac catheterization in pediatric cases.

Methods: The study was conducted on patients who underwent cardiac catheterization at our clinic for diagnostic purposes. The first serum samples were obtained shortly before the procedure, with second serum samples obtained 4–6 hours after the procedure and third serum samples obtained 24 hours after the procedure. Quantitative tests were utilized to measure cardiac troponin I (cTnI), creatine kinase myocardial band (CKMB) and heart-type fatty acid binding protein (HFABP) levels in the serum samples.

Results: The study analyzed a total of 36 patients underwent cardiac catheterization for diagnostic purposes (16 girl, 20 boy) with a median age of 20 months (range 4–192 months) and a median weight of 7.2 kg (range 4.3–44 kg). The laboratory findings were as follows: cTnI, 0.19 ± 1.91 ng/ml (before procedure), 1.90 ± 2.1 ng/ml (4 hours after procedure) and 0.57 ± 0.66 ng/ml (at 24 hours); and CKMB 3.62 ± 1.93 ng/ml (before procedure), 9.03 ± 8.64 ng/ml (4 hours after procedure), 3.87 ± 4.00 ng/ml (at 24 hours) and HFABP, 3.98 ± 2.09 ng/mL (before procedure), 3.93 ± 1.89 ng/mL (4 hours after procedure) and 3.25 ± 1.34 ng/mL (at 24 hours). Substantial increases were observed in the levels of cTnI and CKMB after angiocardiography procedure, while HFABP levels remained unchanged. The increases were even more remarkable in patients aged below 1 year and weighing below 10 kg.

Conclusions: Myocardial injuries can occur during diagnostic cardiac catheterization in the pediatric age group. Special attention must be paid in cases younger than 1 year old and in those weighing below 10 kg.

P-98

Platelet volume parameters in both cyanotic and acyanotic congenital heart diseases

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Introduction: The mean platelet volume and platelet distribution width are parameters that indicate platelet activation and function. The increase in mean platelet volume and decrease in platelet distribution width demonstrates an increase in platelet reactivity and a tendency to thrombosis. Interestingly, it is known that individuals with cyanotic congenital heart disease are predisposed to thrombosis. We aimed to determine the relationship between platelet count, mean platelet volume, and platelet distribution width in patients with cyanotic and acyanotic congenital heart disease.

Methods: This study included 40 cyanotic patients (group1) that were followed in the pediatric cardiology clinic, 40 acyanotic (group2) patients that applied to the outpatient clinic with heart disease, and 40 control patients (group3) that did not have any heart diseases. Groups were compared in terms of platelet count, mean platelet volume, and platelet distribution width.

Results: In group1 there were 14 female and 26 male patients with a mean age of 1.5 years (1 day–13.5 years), in group2 there were 24 female and 16 male patients with mean age of 3.6 (1 month–17

years), and in group3 there were 15 female and 25 male patients with mean age of 4.7 years (5 days–17 years). There was no significant difference between the groups in terms of the platelet count. Mean platelet volume in group1 was 9.6 (8.9–10.7) femtoliters (fl), in group 2 was 6.8 (5.9–7.9)fl, while in group3 it was 6.9 (6.5–8.8)fl. The post-hoc analysis showed that there was a statistically significant difference between group1 and groups 2 and 3 ($p=0.001$), but no significant difference between groups 2 and 3. Mean platelet distribution width in group 1 was 11.6 (10.4–14.6)fl, group2 was 16.9 (16.1–17.4)fl, and group3 was 16.5 (11.8–16.8)fl. The post-hoc analysis showed that there was a significant difference between group1 and groups2 and 3 ($p=0.001$), however there was no significant difference detected between groups 2 and 3.

Conclusions: Mean platelet volume was elevated in pediatric patients with cyanotic congenital heart disease compared to patients with acyanotic heart diseases or patients without any disease. There is a need for further studies to investigate whether the mean platelet volume can be used as a marker for predisposition to thrombosis and platelet activation, and may serve as an indicator for cardiovascular complications.

P-99

Growth differentiation factor-15 levels of before and after treatment in children with acute rheumatic fever

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Introduction: We aimed to investigate the role of growth differentiation factor-15 in acute rheumatic carditis by evaluating correlations with acute-phase reactants and echocardiographic values.

Methods: The 32 patients were in the age group of 6–16 years (mean 11.00 ± 2.6 years). We chose 32 healthy children, matched for sex and age, as the control group: 20boys and 12 girls, aged from 7 to 16 years (mean 11.9 ± 2.3 years). All patients were examined by paediatric cardiologists. After routine cardiovascular examination, chest roentgenogram and electrocardiogram were obtained for all patients. All of patients were treated with oral prednisolone. The reaction is the plasma GDF-15 level was measured of healthy controls and also at the time of diagnosis and 6–8 weeks of the treatment for children with acute rheumatic carditis. Cardiac diagnosis was confirmed by echocardiographic investigation. Blood samples were drawn for analysing plasma GDF-15 and acute-phase reactants, ESR, CRP levels at baseline on Days 1, and at 8, weeks.

Results: There was no statistically significant difference between baseline age, gender, body mass index. On day 0 (before the treatment) ASO titers, ESR and CRP levels were significantly higher in the patient group than the control group. The plasma GDF-15 level was higher in patient group before therapy than in controls but there were no significant differences ($p:0.28$). Following the antiinflammatory therapy, we found a progressive decrease in plasma GDF-15 levels. There was not a significant difference between the control group and the patient group at the end of therapy in point of plasma prohormone levels ($p:0.874$). There were no patients with a positive throat culture for group A

β -haemolytic streptococcus but all patients had supportive evidence of a preceding streptococcal infection which was documented by high antistreptolysin-O titers. However, no patients had significantly cardiac failure and no cardiomegaly was noted on chest radiograms.

Conclusion: Plasma GDF-15 levels appears to be regulated uniquely in the setting of a cardiac inflammatory process and plasma levels increased to different extents in the acute stage of illness. Thus, in order to determine the diagnosis and activation of disease, GDF-15 may be used as a biomarker of cardiac involvement.

P-100

Incomplete Kawasaki Disease With Coronary Artery Lesion After \leq 5 Days Of Fever Responded To Antibiotics

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Objectives: Kawasaki disease (KD) is the most common vasculitis in the developed countries and the diagnosis of KD is made by more than 5 of 6 diagnostic criteria including \geq 5 days of fever. However, the Japanese annual survey of KD showed certain number of patients with incomplete KD with coronary artery lesion (CAL). The aim of this study was to identify and characterize patients with incomplete KD who do not fulfill the diagnostic criteria and responded to standard antibiotics treatment but left with coronary artery lesion (CAL).

Methods: We searched our KD database to identify patients with incomplete KD with \leq 5 days of fever and responded to standard antibiotics treatment but left with CAL. Based on the retrospective medical chart review, we characterize patients' demographics, number of signs, treatment, location and the timing of identifying CAL, and the prognosis.

Results: Of 561, we found 5 patients (0.9%) that met with the inclusion criterion. The patient's age ranged from 2 months to 2.8 years with a mean of 14.8 ± 15 months old. Numbers of presented diagnostic signs of KD were 2 or 3 except for fever. Three patients were treated with intravenous antibiotics, 1 with oral antibiotics, and the remaining 1 did not receive any antibiotics. CALs were located in both coronary arteries except for 1 in right coronary artery and were noted from 7 to 41 days after the onset. The maximum size of CAL ranged from 3.1 to 8.0 mm with a mean of 4.7 ± 1.4 mm. All patients were placed on oral aspirin and 1 was added warfarin. Of 5, 3 patients showed regression but 2 were left with CAL at 1 year after the onset, confirmed by coronary angiography.

Conclusions: Though it is rare, there are certain numbers of patients who do not fulfill diagnostic criteria of KD and respond to standard antibiotics treatment with \leq 5 days of fever but left with CAL. Pediatric cardiologists must be aware of this fact and schedule timely echocardiography in these patients.

P-101

Experimental Immunohistochemical Study on Persistent Vascular Remodeling related to Development of Arteriosclerosis or Atherosclerosis in Kawasaki Disease

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Introduction: Atherosclerotic coronary heart disease has recently emerged as a clinical issue among young individuals with a history

of Kawasaki disease (KD), which is a systemic vasculitis unique to children. However, whether or not and how KD promotes atherosclerosis remains unclear. We hypothesized that, analogous to the pathogenesis of arteriosclerosis or atherosclerosis, endothelial injury and the resultant intimal thickening are induced in coronary arteries after attenuation of vasculitis.

Methods: We used a rabbit model of KD developed by Onouchi et al. and performed histopathological analysis of the coronary arteries at acute (1, 3, 5, and 7 days) and chronic (3 months) phases of the disease.

Results: In these rabbit models, a pan-arteritis with significant intimal cellular hypertrophy was histologically detected in the acute phase, and arterial intimal thickening was observed during the chronic phase. Immunohistochemical analysis of the coronary arteries revealed that the thickened intimal lesions observed during the chronic phase comprised abundant α -smooth muscle actin (α -SMA)-positive cells, most of which concomitantly expressed vascular cell adhesion molecule-1 and nuclear factor- κ B. Although macrophages positive for RAM11 were barely detected, macrophage colony stimulating factor was strongly expressed in migrating smooth muscle cells in the intimal layer. In addition, the accumulation of proteoglycan as extracellular matrix was distinctly visible in the thickened intima, indicating progressive accumulation of lipids and proliferation of smooth muscle cells within the lesion.

Conclusions: These findings suggest that, in KD-associated vasculitis, the migration of α -SMA-positive cells into the thickened intima might induce continuous vascular inflammation and remodeling, which might progress to coronary arteriosclerosis or atherosclerosis.

P-102

Risk Factors in Developing Coronary Artery Abnormalities Among Children With Kawasaki Disease

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Introduction and Objective: Kawasaki disease is an acute febrile illness seen in childhood, with 80% of the patients less than 5 years old. It affects the medium sized arteries with predilection of the coronary arteries, causing coronary artery abnormalities. The main objective of this study is to determine the risk factors for developing coronary artery abnormalities based on sociodemographic factors (age and sex), laboratory examinations (hemoglobin count, white blood cell count, platelet count, erythrocyte sedimentation rate, C reactive protein), and the timing of treatment with intravenous immunoglobulin.

Method: This is a case control retrospective study. All children up to 5 years of age, who have met the inclusion criteria, admitted in our institution from 2000 to 2012 with Kawasaki disease were included in the study and divided into two groups according to age. Patients 1 year old and below belonged to group A, and patients beyond 1 year old belonged to group B. Their risk factors were identified, analyzed, and compared using Chi Square test, conditional maximum likelihood estimate, and logistic regression analysis.

Results: Two hundred six records were reviewed and analysed. Twenty six out of 83 patients in group A and 34 out of 123 patients in group B developed coronary artery abnormalities. There were more males who developed coronary artery abnormalities in group A but there were more females in group B. Lower hemoglobin levels (less than 105 milligram per decilitre) in patients with coronary artery abnormalities were mostly seen in group A as compared to group B. Leukocytosis (more than 12,000 cells per cubic millimetre), thrombocytosis (more than 400,000 cells per cubic millimetre), elevated C reactive protein (more than

6 milligram per litre), and elevated erythrocyte sedimentation rate (more than 10 for males and 20 for females) were evident in both groups. Timing of treatment with intravenous immunoglobulin was at an average of 8.67 days for group A and 8 days for group B. Only thrombocytosis, with a *p* value of 0.02, was statistically significant.

Conclusion: Thrombocytosis is a risk factor in developing coronary artery abnormalities among patients up to 5 years of age with Kawasaki disease.

P-103

Safety and tolerability of Bisoprolol treatment in children

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Introduction: Bisoprolol is a widely used beta-blocker in adults due to its pharmacokinetics and cardioselective properties. However, its safety and efficacy is unknown in children. The aim of this retrospective study was to determine the safety and tolerability of bisoprolol in a large paediatric population.

Methods: The indications, the dosage and outcome of 184 consecutive children commenced on Bisoprolol treatment between October 2003 and May 2014 were retrospectively reviewed.

Results: Indications for Bisoprolol were long QT syndrome (*n* = 59), hypertrophic cardiomyopathy (*n* = 34), ventricular arrhythmias (*n* = 22), supraventricular tachycardia (*n* = 19), atrial arrhythmias (*n* = 17), aortic root dilation (*n* = 11), cardiac dysautonomia (*n* = 10), hypertension (*n* = 6), repolarization abnormalities with a family history of SADS (*n* = 2), ARVC (*n* = 2), LV diastolic dysfunction (*n* = 1) and CPVT (*n* = 1).

Bisoprolol treatment was started at a median age of 12.8 years (range 0.4–18.8 years) in 176 patients. Median starting dose was 1.25 mg (range 0.625 mg–10 mg) in 174 patients. The initial dose was higher when converted from another beta blocker. The dose was up-titrated as tolerated and clinically-indicated. 150 patients were on Bisoprolol at their last clinical appointment with a median dose of 2.5 mg (range 1.25 mg–10 mg).

Side effects were documented in 39 patients (21.2%): lethargy and/or tiredness (*n* = 22); dizziness/lightheadedness (*n* = 4); headaches (*n* = 2); sleep disturbance (*n* = 5); epigastric pain and dizziness (*n* = 2); blurred vision (*n* = 1); worsening eczema (*n* = 1); decreased exercise tolerance (*n* = 1), unknown (*n* = 1).

Bisoprolol was discontinued in 54 patients (at a median 4.45 months (range 2 days – 5.04 years): due to side effects in 23 (12.9%); no longer required in 20 (successful RFA *n* = 8, scheduled RFA *n* = 2, no arrhythmia/symptoms recurred *n* = 4, deceased *n* = 2, on Berlin Heart *n* = 2, heart transplantation *n* = 2); it was not effective in 7; patient's non-compliance in 3; changed to another medication once prescription run out in 1.

Conclusions: Bisoprolol is safe and relatively well tolerated in children. Further prospective studies are required to establish its efficacy in specific paediatric indications.

P-104

Postoperative outcome in infants with aortic coarctation of the aorta

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This study was to assess prognostic factors of outcome, in neonates and infants with coarctation of aorta (COA).

Methods: Single-center analysis of clinical and echocardiographic data of neonates and infants <1 year of age with COA.

Results: 275 cases were included, diagnosis was suspected prenatally in 95 cases. Heart failure (HF) was the main symptom, absent in patients prenatally diagnosed (*p* < 0.0001). Diameter of isthmus (AOI), transverse arch (TrAO) and AAO were lower in antenatal cases. Systolic fraction was decreased in patients diagnosed after birth. Pre-operative HF was associated with narrowest AOI, AAO, TrAO and decreased LVSF. PGE was administered in 53% preoperatively and mechanical ventilation in 39%. Among 275 confirmed COA, 272 underwent Crafoord operation, at mean age 29 days (median 12 days) and mean time after admission 3.4 days. 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 <1 month age at surgery impact on the risk of restenosis.

P-105

Long term outcome and survival of neonates with total anomalous pulmonary venous return

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The aim of this study was to assess long term survival of children with total anomalous pulmonary venous return (TAPVR).

Methods: Clinical, echocardiographic, and long term complications and reinterventions were assessed in all cases diagnosed with TAPVR from 1973 to 2014.

Results: 180 patients were diagnosed with TAPVR: 78 supracardiac 43%, 48 intracardiac 27%, 35 infracardiac 19% and 19 mixed 11%. Mean FU was 11 years. One hundred and forty two patients survived in-hospital stay (79%). The incidence of late complications was 19.6% (28 cases): 8 pulmonary veins stenosis, 5 residual ASD or partial APVR, 3 vena cava stenosis, 3 right to left atrial shunt, 3 neurological sequelae and 6 miscellaneous.

Duration of bypass, mixed pattern TAPVR and preoperative mechanical ventilatory support were significant risk factors for late complication respectively *p* = 0.01, *p* = 0.0023 and *p* = 0.09). Eight patients suffered from late pulmonary venous stenosis, of whom 7 died (6 in the early course after redo surgery and 1 before reoperation) and 6 were operated on before 2000.

Sixteen patients were reoperated on for pulmonary venous stenosis (7), residual ASD or partial PVR (5) and vena cava stenosis (4). Twelve late deaths occurred (6.7%), 5 from non cardiac causes and 7 due to pulmonary veins stenosis.

Survival was 80%, 75%, 70% and 65% at respectively 1 year, 10 years, 20 years and 40 years of FU. Survival was significantly lower in infracardiac type (*p* = 0.0017). Overall 131 patients survived of whom 84.7% are in NYHA class I with none ongoing cardiac medication. Pulmonary pressure levels range within normal value in 109 cases (83%), grade I PHT persists in 9 (7%).

Conclusion: Overall late outcomes of patients with TAPVR is favourable, and the infracardiac type is associated with the lowest survival rates. Pulmonary veins stenosis is a rare but lifethreatening late complication.

P-106**Pulmonary Vasodilators Treatment in Failing Fontan: Data from the Spanish Registry for Pediatric Pulmonary Hypertension (REHIPED)**

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Introduction: Proteing losing enteropathy (PLE) and Plastic Bronchitis (PB) are a growing cause of morbidity, as survivors of the Fontan operation continue to age. Both disorders can lead to death, describing a poor prognosis, with a reported 50% mortality at 5-years after diagnosis. Limited data suggest the successful use of pulmonary vasodilators (PV) as a treatment strategy. Our purpose was to study the clinical/hemodynamic findings and survival of patients who had received PV for PLE/PB.

Methods: From 2009 to 2014, 26 Fontan patients receiving PV following diagnosis PLE/PB (84% PLE) were identified from clinical database at the Spanish Pediatric Pulmonary Hypertension Registry. Data were collected retrospectively.

Results: Mean age at PLE diagnosis was 10.4 (5.1–20) years. Fontan operation was performed at 5.9 (2.4–8.7) years of age. Fontan operation to PLE/BP diagnosis was 4.5 (0.2–13.2) years. Diagnosis of PLE/BP to PV started was 8.4 months. Transplant-free-survival or death was 88.5% at 5-years. Treatment achieved complete resolution of clinical symptoms and laboratory values in 6 patients (23%). Symptomatic and functional class improvement was achieved in 16 patients (61%) at 6-months follow-up, but this symptomatic improvement lasted only in 7 patients (27%) at 12-months follow-up, and 6 patients (23%) at 24-months follow-up. Patients who died or received a heart transplantation (HT), had lower cardiac index (2.36 ± 0.3 vs. 3.47 ± 1.1 l/min/m²; $p = 0.002$), lower mixed venous saturation (SvO₂) (67.5% vs. 73%; $p = 0.005$), and NYHA functional class >2 at diagnosis, and later they began therapy with PV (2.8 vs. 0.6 years post-diagnosis, $p = 0.042$). In 12 patients, in which clinical outcome was not satisfactory, an hemodynamic assessment was repeated. Only significant change was observed in the SvO₂ (Pre-treatment: 67% vs. Post-treatment: 72%; $p = 0.03$). However, a trend towards decreased central venous pressure and decreased cardiac index, as a result of intensive management after PLE/PB diagnosis, was observed.

Conclusions: PV have potential to improve symptoms and delay need for HT in this patient population. Although, the success rate of PV (associated to conventional therapy) was not superior to the previous reported series. As resolution always happened in the first 6–12 months after treatment, HT should be considered in any PLE/PB when there hasn't been response after that period.

P-107**Evaluating markers of positive response on therapy and survival of children with pulmonary arterial hypertension in Russia**

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Objective: to evaluate an association between clinical, functional and haemodynamic features, current treatment options and outcomes in children with pulmonary arterial hypertension (PAH).

Methods: 50 consecutive patients with PAH were included and followed up through Jan 2011 – Dec 2014; mean age at 9.4 ± 5.4 and 51% were males. Tests included routine cardiological examination; echocardiography, CT angiography and scintigraphy; six-minute walk test (6MWT) in children aged 7+; right heart catheterization (RHC) and acute pulmonary vasodilator testing in 63% of pts. Kaplan Meier Method and Cox proportional hazard model were employed for testing differences in survival.

Results: PAH was associated with congenital heart defects in 42 pts (84%), with portal hypertension in 2 pts (4%). In 21 pts (42%) more than one associated conditions were found. 34% of children were with Down's syndrome. Associated conditions were not identified in 12% patients. They were classified as having idiopathic PAH. Late diagnostics of PAH in children was observed in 71% of pts. At first examination 29% of pts had functional class (FC) II, 65% of pts had FC III; and 6% - FC IV. Mean pulmonary artery pressure varied from 30 to 105 mmHg, and pulmonary vascular resistance (PVR) varied from 8.5 to 29.3 UW. Median cardiac index was 1.9 ± 0.5 L/min/m² and median arterial oxygen saturation was $90 \pm 2.7\%$. Patients were treated with sildenafil ($n = 4$), bosentan ($n = 25$), combination therapy by bosentan and sildenafil ($n = 16$), bosentan, sildenafil and Inhaled iloprost ($n = 5$). The atrioseptostomy was performed in 4 pts. Median duration of follow-up was 42 months. Reduction of dyspnea and improvement of RV function were observed in 22% of pts. 6MWT distance increased by 116.2 ± 12.7 m, PVR decreased by 8.9 ± 3.4 UW. Five IV class pts have died. Reduction of dyspnea, improvement of RV function and increased cardiac index are significantly associated with positive response to therapy.

Conclusions: Pediatric PAH frequently presents with associated conditions and syndromal abnormalities. FC and hemodynamic parameters are the strongest predictors of survival in children with PAH. There are no statistically significant sex differences in incidence, age at onset, disease severity.

P-108**Analysis of different methods of evaluation of myocardial hypertrophy in children with arterial hypertension**

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Basics: To analyse indexes of evaluation myocardial mass (MM) and myocardial hypertrophy (MH) in teenagers with arterial hypertension.

Methods: 106 boys of $15 \pm 1,6$ years old. We excluded teens with secondary hypertension and those, who received hypotensive therapy during ≥ 3 weeks. All patients were divided into 2 groups: 1st – with ambulatory hypertension I ($n = 22$) with Systolic blood pressure (SBP) load 25–50%, 2d – with severe ambulatory hypertension ($n = 84$) and SBP load over 50%. From all the group 35 (33%) boys had normal body mass, 71 (67%) had increased body mass index (BMI), including 43 (40,6%) with obesity. 11 patients from 1st group (50%) and 60 (71,4%) from the 2nd group had increased BMI. In every patient we performed transthoracic echocardiogram (TTE), estimation of myocardial mass, ambulatory blood pressure monitoring with calculation indexes of SBP load. We focused on 6 indexes to estimate MH: 1) MM/Height^{2,7} (>45 g/m^{2,7}), 2) MM/Height^{2,7} (>48 g/m^{2,7}), 3) MM/Height^{2,7} (>51 g/m^{2,7}), 4) zMM(F) ($>1,65$), 5) MM/BSA (>115 g/m²), 6) MM/Body Weight ($>3,0$ g/kg).

Results: frequency of MH varied from 11.3% to 35.9%, depending on the chosen index. MM correlated with SBP and SBP load. Hypertrophy was frequent in patients with severe ambulatory hypertension (28,6%, $p < 0,05$) and with obesity ($p < 0,05$). The highest frequency of MH ($n = 46$ (43.4%)) was received using the 1st index: $MM/Height^{2,7} (>45 \text{ g/m}^2,7)$. While using 1-4 indexes, that did not include body weight percentage of MH remained high in obesity teens: 34% ($n = 24$), 27% ($n = 19$), 18% ($n = 13$), 21% ($n = 15$) corresponding with the index list. Analyzing average indexes ($MM/Height^{2,7}$ и $zMM(F)$) the number of patients with MH was reliably higher in those with severe ambulatory hypertension, than in ambulatory hypertension I: 24 (28,6%) and 1 (4,5%), $p < 0,05$. Last two indexes ($MM/BSA (>115 \text{ g/m}^2)$, $MM/Body \text{ Weight} (>3,0 \text{ g/kg})$) could not detect MH in children with obesity, while other indexes confirmed hypertrophy.

Conclusion: severe ambulatory hypertension is more often in obesity children: $OR = 3,48$ ($CI 1,07-11,28$). Exceeding 95th %tile of SBP is a main reason of appearance of MH. Most adequate indexes to detect MH in teens are $MM/Height^{2,7} (>48 \text{ g/m}^2,7)$ и $zMM(F) (>1,65)$. Indexes MM/BSA , $MM/Body \text{ Weight}$ are not recommended to use in obesity teenagers.

P-109

Can progression of pulmonary vein stenosis be stopped by lobectomy of affected lung?

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Introduction: Primary pulmonary vein stenosis (PVS) is considered a progressive disease caused by neoproliferation of myofibroblasts induced by local or peripheral signaling. However advances in surgical and interventional treatment, there is still long-term morbidity and mortality. We present radical surgery as a treatment option.

Method: A seven year follow up of three patients with PVS who were treated with lobectomy or pneumonectomy between 2007- 2008.

Results: Patient one: Term female with a birth weight of 2980 g, diagnosed at 3 months of age with a stenosed upper right pulmonary vein. She had complicating interstitial pulmonary disease. Mean pulmonary pressure was 29 mmHg at diagnosis. She was initially stented, but 3 months later treated with bilobectomy of the upper and middle right lobe because of severe re-stenosis. Mean pulmonary pressure was 60 mmHg at surgery. Follow up catheterization demonstrated reduced pressure. However, a progressive narrowing of the left lower pulmonary vein was identified and the patient died at 2 years of age from a respiratory tract infection.

Patient two: Preterm female born at 28 weeks gestational age with a birth weight of 868 g, diagnosed at two months of age with two atretic right sided pulmonary veins. She was treated with right-sided pneumonectomy. At 7 year follow-up she is doing well with height and weight for age just below the 3rd percentile.

Patient three: Preterm female born at 25 weeks gestational age with a birth weight of 525 g, diagnosed at 7 months of age with an atretic left upper vein and a narrowed left lower vein. Mean pulmonary pressure at diagnosis was 51 mmHg. She underwent lobectomy of the left upper lobe. Her mean pulmonary pressure was reduced to 26 mmHg after three weeks and 22 mmHg after one year. Today no signs of re-stenosis are seen. She is healthy with weight for age on the 10th percentile and length for age on the 25th percentile.

Conclusion: PVS still carries a high rate of morbidity and mortality. Radical treatment with lobectomy or pneumonectomy of tissue containing severely stenosed veins might slow or halt disease progression.

P-110

Automated Electronic Diagnosis of Pathologic and Innocent Murmurs

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Introduction: Approximately 50% of children have heart murmurs, but only <1% are clinically relevant pathological murmurs. The majority of elective referrals made to paediatric cardiologists result from the inability of general practitioners/paediatricians to distinguish between innocent and pathologic murmurs. We improved and validated a computational algorithm to automatically detect pathologic and innocent murmurs from recorded phonocardiograms (PCGs).

Methods: Patients from general paediatric cardiology clinics in CHEO were recruited (November-December 2013). The cardiologist recorded PCGs using an electronic stethoscope. Two PCGs per patient were recorded at the loudest location for 15-30 seconds; one PCG free-breathing, one PCG with breathhold. If the patient could not breathhold then both PCGs were recorded free breathing. The PCGs were exported as standard .wav audio files to be used in the further testing of the algorithm.

Results: $N = 106$ patients (2 days-18 years, mean age 8 years). 57% (60/106) males. 85% (90/106) of the patients had echo confirmation of the murmur diagnosis. 39% (41/90) had pathologic murmurs, 15% (16/90) had innocent murmurs and 31% (33/90) had no audible murmur. Of the patients referred for assessment of a murmur 86% (12/14) were not pathologic. 10/12 murmurs were diagnosed as innocent by the physician of which 1/10 was confirmed by echo. 2/12 patients had no audible murmurs at the time of auscultation.

An analysis of the algorithm's performance to distinguish pathologic from innocent murmurs yielded a sensitivity and specificity of 95% and 93%, respectively. This was performed on 66 randomly selected PCGs that were used in the training of the algorithm (all free breathing, echo confirmed diagnosis in 54/66, pediatric cardiologist confirmed in 12/66). The performance on 30 randomly selected PCGs that were NOT used in developing the algorithm resulted in a sensitivity and specificity of 83% and 100%, respectively (all breath-held, echo confirmed diagnosis in 24/30, pediatric cardiologist confirmed in 6/30).

Conclusions: We successfully show that it is possible to utilize a computational algorithm to diagnose electronically recorded PCGs accurately. With echocardiogram as a gold standard, despite breathing and noise, this algorithm can detect murmurs with high sensitivity and specificity.

P-111

Outcomes of coronary artery lesions after neonatal arterial switch operation

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Background: Coronary artery lesions (CL) are the main cause of morbi-mortality after arterial switch operation (ASO) for

transposition of the great arteries (TGA). Outcome and treatment of CL after ASO has been scarcely reported.

Objective: To study the long-term outcomes of CL after ASO.

Methods: We identified 75 patients with CL after ASO over a period of 30 years. CL were either ostial or proximal, and involved the left main stem in 34 patients, the left anterior descending artery in 19, the circumflex artery in 10, and the right coronary artery in 12 patients. 35% of patients were symptomatic and diagnosed at time of an ischemic event. The remaining 65% were asymptomatic and diagnosed during a systematic screening. In this group, myocardial ischemia (MI) was demonstrated in 45% of patients.

Results: First intention treatment was coronary revascularization in 32 patients (43%) (surgical angioplasty in 25, graft by-pass in 3, percutaneous balloon dilatation in 5), medical treatment in 15 (20%), and surveillance in 25 (33%). Three patients died before any treatment. Mean follow up was 10.6 ± 7.9 years. Survival was 90% at 20 years. A second intention treatment was needed because of a new anatomical lesion or new onset MI in 27% of patients who received medical treatment as first line therapy, in 20% of patients who were not treated, and in 12.5% patients who underwent revascularization. Overall, revascularization was performed in 73% of symptomatic patients, 72% of asymptomatic patients with MI, and in 22% of asymptomatic with no MI at diagnosis. At last follow-up, one patient has a residual MI.

Conclusion: Coronary lesions after ASO are not uncommon. In patients with MI, revascularization seems to be the treatment of choice. In non-ischemic patients at diagnosis, early revascularization needs to be considered in light of the severity of the lesion and as MI can appear during follow-up.

P-112

Myocardial fibrosis detected by cardiac magnetic resonance imaging in children with hypertrophic cardiomyopathy

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Background: Contrast-enhanced cardiovascular magnetic resonance with late gadolinium enhancement (LGE) has emerged as an in vivo marker of myocardial fibrosis, although its role in stratifying sudden death risk in pediatric hypertrophic cardiomyopathy (HCM) remains incompletely understood.

Here, we sought to determine the prevalence of LGE in children with HCM with different causes and its relation with left ventricular mass and geometry.

Methods: CMR protocol included T2 weighted sequence in short axis view, TRIPLE IR FSE sequence, cine SSFP in short axis, two-chamber, three and four chamber view without contrast and perfusion analysis and late enhancement after injection of contrast agent. If left ventricular wall thickness was asymmetric, the size and location of relatively thickened segments were noted.

Results: A total of 74 children were included. Age at diagnosis was 3 years (range 1 day to 16 years). Mean age at CMR was 11 years (range 1-18 years). CMR was successfully performed in all patients, revealing a better performance in comparison to echocardiography to define precisely the anatomy of LV hypertrophy. Mean LV mass was estimated at 94 ± 41 gr/m². LV hypertrophy was concentric in 35 patients, asymmetric in 39 patients, with evidence of LV non-compaction aspect in 10 patients. Right ventricular hypertrophy was observed in 7 cases. LGE was detected in 6 patients within the septum and in 1 patient in apical wall.

Perfusion defects were present in 5 patients in papillary muscles. LV ejection fraction was < 55% in 7 patients. We found no correlation between LV mass, LV outflow tract obstruction or geometry of the hypertrophy and the presence of LGE. Children with LGE did not have more cardiac events than those without.

Conclusion: In our series, the prevalence and extent of LGE in children with HCM was rare compared to adults with HCM. Furthermore, LGE was not associated to adverse events, such as sudden death that also occurred in patients without LGE.

P-113

Cardiac biomarkers in newborns with congenital heart defects

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Introduction: BNP is released from the ventricular myocardium in response to stretching of the ventricular wall. Troponin I, myoglobin and CK-MB are biomarkers of cardiomyocyte injury widely used in the management of adult patients. The role of these biomarkers in neonates is still not established. The purpose of this study was to evaluate the diagnostic and prognostic value of cardiac biomarkers in newborns with congenital heart disease (CHD).

Materials and Methods: Cohort consecutive study of 54 newborns, 34 with prenatal diagnosis of CHD admitted at the Neonatal Intensive Care Unit and a control group (n=20) of healthy newborns delivered in the same tertiary hospital. Plasma levels of cardiac biomarkers (BNP, troponin I, myoglobin and CK-MB) were evaluated and echocardiogram performed within the first 24 hours of life. Patients were followed during the first 28 days of life (neonatal period) and accordingly with the outcome categorized as surgical or conservative-treatment group.

Results and Discussion: Median BNP was higher in patients with CHD than in controls (43.3 vs. 19.8 pg/mL; p = 0.001). From the 24 patients with CHD with a surgical indication, 10 underwent cardiac surgery during neonatal period. BNP was higher in patients that had cardiac surgery during the neonatal period (73.7 pg/mL) than in those discharged home without surgery (25.6 pg/mL; p = 0.016). A BNP cut-off point of 35.85 pg/mL predicted neonatal surgery (sensitivity 90.0% and specificity 64.3%). Median (P25-75) levels of CK-MB were higher in patients that had cardiac surgery in the neonatal period [7.35 (4.90-13.40) ng/mL] than in patients who were discharged home without surgery [4.2 (2.60-5.90) ng/mL; p = 0.032]. Troponin I and myoglobin levels were not significantly different between conservative treatment and surgical group. BNP and CK-MB levels correlated with the tissue Doppler image (TDI) peak early diastolic velocity of the mitral annulus/late diastolic velocity of the mitral annulus ratio ($\rho = -0.543$, p = 0.007; $\rho = -0.480$, p = 0.018, respectively).

Conclusions: Newborns with CHD presented higher BNP levels than healthy newborns. BNP and CK-MB levels in the first hours of life have prognostic value for neonatal cardiac surgery and may be indicators of diastolic cardiac function.

P-114

Exercise tolerance after Senning operation for transposition of great arteries and its limiting factors

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Objectives: Senning operation is associated with increased long-term risk of cardiovascular mortality and morbidity. Decreased exercise capacity carries a substantially higher risk of death. The purpose of the study was to assess the exercise capacity in patients with Senning operation and to determine its limiting factors.

Methods: 87 out of a total of 152 pts (men 64/87) who underwent the Senning operation between 1984–1997 could be recruited for a cross-sectional evaluation at median 22.5 (range 15.2–29.4) years after surgery consisting of a questionnaire, clinical examination, echocardiography, magnetic resonance imaging and cardiopulmonary exercise test. Maximum oxygen uptake (VO₂max) was considered to be an objective exercise performance parameter in the study. Pathologic limits were defined as those exceeding 2 Z-values.

Results: 73% of pts were NYHA class I and 27% NYHA II. Involvement in some kind of sport activity was declared by 83.9%. Maximum exercise level could be achieved in 67/87 pts. The median Z-values of both VO₂max and of the maximum achieved heart rate (-1.971 and -1.795, resp.) were decreased with significant decrease in maximum exercise capacity in 49% of pts. 22% pts had increased blood pressure at rest and/or during exercise. In a multivariate analysis decreased VO₂max was predicted by the VE/VCO₂ slope at exercise (OR = 1.271, CI 1.069–1.511, P = 0.007) and BMI (OR = 1.193, CI 1.027–1.386, P = 0.030) but not by the right ventricular ejection fraction, degree of tricuspid regurgitation, heart rate at maximum exercise or NYHA class.

Conclusions: The Senning operation is associated with decreased exercise tolerance in 50% of the pts. Decrease in exercise capacity is predicted by increased BMI and by ventilation inefficiency, but not by objective hemodynamic parameters. A significant proportion of pts has an arterial hypertension. Obesity, hypertension control and respiratory rehabilitation should thus be an essential part of the long-term management.

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P-115

Left Ventricular Mass Indexation in Infants, Children and Adolescents: a Simplified Approach for the Identification of Left Ventricular Hypertrophy in Clinical Practice

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Introduction: LVH in children is a marker of cardiovascular risk in both congenital and acquired diseases. However the complex relationship between heart growth and body growth in children has made indexing difficult for younger ages. Thus, a number of approaches have been suggested, mostly needing time-consuming computation of specific centiles, resulting in low applicability in clinical practice. Our purpose was to determine a simplified method to identify presence of LVH in pediatric populations.

Methods: Four hundred healthy children (N = 400, 52% boys, 0–18 years) from two different European hospitals were enrolled in the study. Participants were children evaluated for innocent murmurs or chest pain who were then determined by echocardiography to have normal hearts. Exams were performed on commercially available machines and DICOM files were reviewed off line on digital review stations to obtain measurements of ventricular

diameters and thickness. Left ventricular mass was calculated according to the Devereux formula.

Results: There was a strong non-linear correlation between height and LV mass. Left ventricular mass was related to height to a power of 2.16 with a correction factor of 0.09. Similar results were obtained when data were separated by gender or by age group. As compared to formula currently used in clinical practice (i.e. LVM/height^{2.7}) in which scatter of residuals increased at lower height range, analysis of residuals for LVM/[(height^{2.16}) + 0.09] did not change with increasing height, also when dichotomizing the population by sex, suggesting an homoscedastic distribution in both genders throughout the whole height range. A partition value of 45 g/m^{2.16} was defined as the upper limit of normality for LV mass index.

Conclusions: We have derived a simplified formula to index LV mass from a population of 400 comprising both genders and equally distributed among all pediatric age range. Our data support the possibility to have a single partition value to identify LV hypertrophy, without the time-consuming need of computing specific percentiles for height and gender. Indexation (height^{2.16}) + 0.09 with a partition value of 45 g/m^{2.16} may be used in pediatric patients of both genders of any height, to identify LVH easily in newborns, children and adolescents.

P-116

Cardiac involvement in Acute Myeloid Leukemia: as rare as important

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Introduction: Acute myeloid leukemia (AML) is rare in children (incidence of 8 cases/million inhabitants/year). Extramedullary myocardial AML infiltration is an exceptional presentation defined as myeloid sarcoma (MS).

Objective: To describe the first case in a 2 year old child.

Methods: A 2 year-old-child with a 2 week history of Bell paralysis plus eyes and lips edema was admitted for study. In physical examination no lymphadenopathy, organomegaly or skin pathology were found. There was testis inflammation and tachycardia. Blood count and cerebrospinal fluid (CSF) tests and cultures were taken, also bone marrow aspirate (BMA) and genetic tests. Extension study was performed.

Results: Peripheral blood was abnormal: there was neutrophilia with circulating myelocytes, 24% of blast cells in blood count, normal platelets and LDH 980 and no anemia (hemoglobin 15 mg/dl). X-ray revealed cardiomegaly, abdominal echography showed discrete hepatomegaly and mild renal hydronephrosis. Scrotal echography suggested tumoral infiltration. BMA diagnosed AML M4/M5 (40% blasts). CSF proved infiltration with hypercellularity and 97% blasts. Echocardiography revealed a diffuse and heterogeneous cardiac infiltration, involving pericardium and myocardium associated with a moderate circumferential pericardial effusion without hemodynamic compromise and preserved cardiac function. 3 days after induction chemotherapy treatment hemodynamic instability started with increased global infiltration and pericarditis. Iv furosemide and milrinone were used with good results. *Control echocardiography 3 weeks after (patient in first complete hematological remission) showed a significant global decrease in the cardiac infiltration with a minimal residual pericardial effusion, but showed a persistent decrease in left ventricular ejection fraction (LVEF = 40%).* The pretransplant echocardiographic control, after two cycles of chemotherapy treatment, showed recovery of systolic and diastolic biventricular function, without cardiac infiltration or pericardial effusion.



Figure.

Conclusions: The difficulty diagnosing MS requires a high degree of clinical suspicion in the setting of unusual leukemic presentations. AML is an exceptional cause of myocardial infiltration. Prompt recognition of the underlying leukemia and initiation of appropriate therapy are key to reducing overall morbidity and mortality. Echocardiographic aided both diagnosis and assessment of response to treatment of the *cardiac infiltration*.

P-117

Preoperative treatment of pulmonary hypertension in congenital heart disease is associated with improvement in hemodynamics, oxygen saturation and circulating interleukin-6

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Objectives: Perioperative treatment of pulmonary arterial hypertension (PAH) has been considered for selected patients with congenital heart disease (PAH-CHD) in order to minimize the risk of complications following repair of cardiac shunts. Despite the lack of evidence to support such routine, PAH drugs have been used worldwide in pediatric patients undergoing cardiac surgery. We wished to investigate hemodynamic, oximetric and biochemical (anti-inflammatory) effects of short-term pre-operative PAH therapy (sildenafil) in children with PAH-CHD with moderately elevated pulmonary artery pressure (PAP) and vascular resistance (PVR).

Methods: Fifteen patients were enrolled, with age of 15 [9-30] months (median and interquartile range), with a mean PAP of 55 ± 13 mmHg (mean \pm SD) and a PVR index of 5.2 [4.2-8.9] Wood units \cdot m². Oral sildenafil was started at the dose of 1.0 mg/Kg/day, and increased until development of pulmonary congestion (desired effect) or the observation of a >10% decrease in systemic pressure (safety limit). The maximal dose of sildenafil per patient ranged from 1.5 to 5.0 mg/Kg/day, and treatment duration was 8 to 60 days. Efficacy was further evaluated by measuring the pulmonary-to-systemic blood flow ratio (Qp/Qs,

Doppler-echocardiography), systemic oxygen saturation and the circulating level of interleukin-6 (IL-6, chemiluminescence assay). All measurements were performed before surgery.

Results: In the whole patient group, sildenafil treatment resulted in a significant increase in Qp/Qs ratio (1.95 [1.30-2.60] at baseline to 2.25 [1.70-2.90] at maximal dose, $p=0.021$) and peripheral oxygen saturation ($92 \pm 4\%$ to $95 \pm 3\%$, $p=0.005$). These changes were associated with a decrease in serum concentration of IL-6 (pixel density of 286 [72-510] and 134 [47-262], respectively at baseline and maximal sildenafil dose, $p=0.041$). Three patients did not develop any features suggestive of pulmonary over-circulation while on sildenafil treatment, and were considered as non-responders. The remaining ones were subjected to surgical repair of the anomalies, with one immediate postoperative death.

Conclusions: These preliminary observations suggest that most young PAH-CHD patients with moderately elevated PVR respond favorably to PAH therapy. Hemodynamic improvement seems to be associated with changes in proinflammatory profile, with potential benefits. Extended observations in larger patient populations are required for a better understanding of the impact on outcomes.

P-118

Arterial hypertension in children – continuous monitoring of blood pressure

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Introduction: Arterial hypertension in children is increase of systolic blood pressure and/or diastolic blood pressure above the 95th percentile for age, gender and height. The prevalence of hypertension in children is about 3%. In diagnosis, treatment, and prognosis of hypertension in children, important place is given to non-invasive continuous 24-hour automatic blood pressure measurement.

Aim: The aim was to display results of continuous 24-hour blood pressure measurement in clinical environment.

Methods: The study had retrospective-prospective character, and included the period March 1996.-January 2015. ($n=615$). Data were collected from “The Register of Holter blood pressure” of Pediatric Clinic, CCUS.

Results: Patients were aged from 6 months to 18 years. On 370 males (60,1%), 245 females (39,9%), continuous monitoring of tension was performed in order to diagnose, threat or adjust the therapy of hypertension. Most of them were children of school age- 333 (54,1%), there were 246 adolescents (40%), and 36 (5,9%) were preschool children. First monitoring was in 421 (68,4%) cases, a control monitoring in 194 (31,6%), mainly related to the control of treatment efficacy. Of 421 patients, hypertension was proven in 169 (40,1%). Primary (essential, idiopathic) hypertension was in 74 (43,7%) patients, and secondary in 95 (56,3%). Secondary hypertension was related to endocrine diseases with or without metabolic syndrome in 30 (31,5%) patients, kidney disease (parenchymal and vascular) in 25 (26,3%), cardiac diseases in 14 (14,7%) patients, neurological diseases in 7 (7,3%), systemic diseases in 7 (7,3%), other causes of hypertension in 12 (12,9%) patients. 73 patients (17,3% of all monitored, 43,1% of verified) were treated with antihypertensive therapy, only with antihypertensive agents (ACE inhibitors, beta blockers) 52 (71,2%) of them, with antihypertensive and other therapy 21 (28,8%) of them.

Conclusion: 24-hour continuous pressure measurement is necessary clinical method in everyday diagnostics, especially in early diagnosis and treatment of hypertension in children.

Key words: arterial hypertension, children, monitoring

P-119

Coronary Artery Involvement by Chronic Graft-Versus-Host Disease Presenting as Sudden Cardiac Arrest

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Introduction: We present a case with chronic graft-versus-host disease (GVHD) who experienced sudden cardiac arrest 2½ years after allogeneic peripheral blood stem cell transplantation (allo-PBSCT). Sudden cardiac arrest was resulted from myocardial ischemia caused by coronary obstruction.

Case: A 44 month old boy was admitted to the intensive care unit due to sudden collapse at the emergency department (ED) visiting with fever. He was diagnosed at 6 months with chronic granulomatous disease by CYBB gene mutation. Since 3 month of age, he has suffered from multiple abscesses at body and internal organ such as liver and kidney. Allo-PBSCT using HLA matched unrelated donor was performed at 13 months. His post-transplantation course was complicated by CMV infection and acute gut and cutaneous GVHD. His chronic GVHD manifested as elevated hepatic enzymes and sclerosis of skin with multiple joint contractures. For a week before visiting ER, he had shown upper respiratory infection-like symptoms with poor oral intake. At ED, he suddenly collapsed and was successfully resuscitated from ventricular fibrillation. On admission, echocardiography showed no abnormality, however, ECG showed abnormal q-wave and ST changes in the inferior leads, suggesting coronary event. Cardiac magnetic resonance imaging revealed myocardial thinning and hypokinesia of left ventricle with a weak contrast uptake consistent with subendocardial infarct. Coronary artery imaging with multidetector computed tomography showed obliterative narrowing of left anterior descending artery (LAD) and luminal irregularity at the right coronary artery (RCA) and left circumflex artery (LCX). Coronary angiography revealed complete obstruction of proximal LAD, subtotal obstruction of mid LCX and mild narrowing at the RCA. Distal LAD was filled by the small collateral arteries from RCA and LCX. His poor general condition didn't permit further intervention and after 6 months of follow-up, he died suddenly at home.

Conclusions: The coronary artery disease is only rarely occurring cardiac event in children and cardiac complications associated with GVHD are uncommon. Coronary artery involvement, albeit rare, should be recognized as one of the important manifestation of chronic GVHD in children.

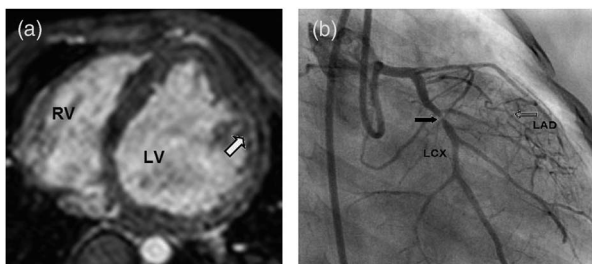


Figure.

P-120

Outcomes of transposition of great arteries in a country without possibility of surgical correction

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Background: Transposition of great arteries (TGA) is a congenital heart disease with very good surgical result in developed countries. The situation is very different in developing countries such as Tunisia. In Fact, neonatal cardiac surgery has poor outcome and especially arterial switch surgery which is not available. Management of babies with TGA is a real challenge for physicians and very expensive for our government. The aim of this study was to assess the outcomes of TGA in Tunisia according to these difficulties.

Methods: TGA patients, who were followed at the Hédi Chaker University Hospital in Sfax from January 2002 to December 2014, were retrospectively evaluated for patient characteristics, clinical manifestation, preoperative management, intraoperative findings, postoperative progress and follow-up status.

Results: Sixty-one patients were included with a sex ratio of 2.39 (43 boys and 18 girls). Of all cases, only one was prenatally diagnosed. The mean age at presentation was 24 (±12.6) days and 82% of TGA were discovered in neonates. The most common symptoms were cyanosis (93.4%) and polypnea (36.1%).

Diagnosis was reached through Doppler echocardiography in all cases. Among our patients, 45 had simple TGA and 16 had TGA with a large ventricular septal defect (VSD).

Forty-five patients underwent Rashkind balloon atrial septostomy (73,7%) at a mean age of 30.7 (±16.9) days, and 19.7% received prostaglandin E1 infusion for an average of 5.5 (±1.1) days. Twenty-four preoperative deaths (39.3%) occurred on average 14.7 (±3.8) days after the diagnosis. All survivals had travelled abroad to be operated (France, Italy and Switzerland). Surgical correction was performed on 37 patients (mean age was 82.7 ± 28.1 days, and mean time from diagnosis to surgery was 48.4 ± 9.8 days. Repair was by one-stage arterial switch operation (ASO) (n = 33), two-stage ASO (n = 3) and Senning (n = 1).

The government paid for these babies an average of 25 000 euros per baby.

Follow-up ranged from one to 126 months (mean 35.8 ± 6.6 months) and it was free of reoperations and late deaths. The 36 survivors have no cardiovascular symptoms. They have normal left ventricular function, have no ischemic problems, and receive no medication.

Conclusions: Mortality rate in TGA is high in spite of early diagnosis. Rashkind atrial septostomy and prostaglandin E1 allowed us to delay the surgical correction which is not yet available in Tunisia. This fact hampers us to reach developed countries' results. Moreover, the cost of corrective surgery is heavy in a country where is mortality of some simple congenital heart defect is still high and unacceptable.

P-121

The prevalence of malnutrition in a regional cardiac service – a cross sectional survey

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Introduction: Growth faltering has both short and long term implications. The WHO defines moderate malnutrition as a weight for height [WHZ] and height for age [HAZ] of <-2 Z score. Growth faltering is commonly seen in children with CHD, particularly those with cyanotic disease and univentricular physiology.

Methods: To investigate the prevalence of malnutrition amongst children attending a regional CHD service anthropometry (weight, length/height) were recorded in all children on the day of their clinic appointment (October 2013-August 2014) using standard operating procedures.

Results: n = 1781 children's anthropometrical measures were recorded. Children with CHD were significantly more stunted

Table 1. Prevalence of malnutrition in outpatient attenders at Paediatric Cardiology UHS October 2013 – August 2014

Z score*	UK prevalence	0-12 months (n=256) [95% CI; Mean, SD]	Z score	UK prevalence – [3, 4]	12-60 months (n=451) [95% CI; Mean, SD]	5-19 years old (n=1074) [95% CI; Mean, SD]
WAZ<-2	0.6%	29.1% (14.5%, 24.2%) -0.85 ±1.61	WAZ <-2	0.6%	2% (2.8%, 6.9%) 0.03 ±1.16	5.4% (2.7%, 7.2%) 0.03, ±1.08
WHZ<-2	0.6%	10.5% (0.3%, 2.8%) -0.21 ±1.3	HAZ <-2	0.6%	17% (10.6%, 17.2%) 0.5 ±1.3	9.5% (5.9%, 9.2%) -0.12 ±1.26
HAZ<-2	2.3%	28.4% (14.2%, 24.3%) -0.76 ±1.57	BMI <-2	2.3%	2.5% (0%, 3.4%) 0.34 ±1.03	4% (2%, 4.2%) 0.26 ±1.24
			BMI > 2	9 - 14%	4.9% (1.4%, 7.5%) 0.34 ±1.03	10.2% (7%, 10.5%) 0.26 ±1.24

*Abbreviations: WAZ=weight-for-age z score; WHZ = weight-for-height-z-score; HAZ=height -for-age-z-score; BMI=body-mass-index

compared to the UK prevalence ($p < 0.0001$), particularly apparent in infants < 12 months (29.1%) < -2 z scores vs. 2.3%. As children got older although the number of stunted children decreased, those who were overweight/obesity increased in line with the national average at 10.2% (5-19 year-olds).

Conclusion: Stunting or poor linear growth has the greatest influence impact on neurodevelopment and poor functional outcomes. As children get older the incidence of stunting remains higher than the UK average, but the trend towards wasting is reversed as the incidence of overweight/obesity rises, increasing the risk of subsequent co-morbidities, during early adulthood particularly coronary artery disease.

P-122

Infective Endocarditis in paediatric and congenital heart disease patients: thirty years experience in a single centre

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Introduction: Infective Endocarditis (IE) is a serious complication in both operated and unoperated patients with congenital heart disease (CHD). In 2008, new guidelines were introduced in the UK, contraindicating the use routine antibiotic prophylaxis for potentially bacteraemic procedures in patients with CHD. The current study explores our experience of IE in paediatric and CHD patients in a single centre, and analyses this experience in relation to prophylaxis guidelines.

Methods: Patients with CHD and children with structurally normal hearts, diagnosed with IE from January 1985 to February 2013 were identified from our database and their case records reviewed. The incidences of IE before and after 2008 were compared.

Groups were analysed using descriptive statistics (frequencies, crosstabs) and chi square test where applicable.

Results: Sixty cases with IE were identified; 54 with CHD (36 adults, 18 children) and 6 children with normal hearts. At the time of IE diagnosis, 27 (50%) patients (17 adults and 10 children with CHD) had previously undergone corrective or palliative cardiac surgery. Thirty three patients (55%) (18 adults and 10 children with CHD) and 5 children with normal hearts had undergone a previous documented invasive, non-cardiac procedure one to twenty weeks before the diagnosis of IE. Forty two (70%) cases (27 adults and 13 children with CHD and 2 children with normal hearts) underwent surgery for IE. Six patients (14%) (4 adults and 2 children, that had surgery for IE) presented with IE relapse, 1 (17%) of the patients within the first year after the operation. Two (4.7%) adults that had undergone IE surgery died following prolonged hospitalisation.

The frequency of IE did not differ in patients with CHD that had a non cardiac invasive procedure documented before IE diagnosis compared to those who did not (33.3% vs 58.9%, $p = 0.131$, respectively) before and after the implementation of new national guidelines.

Conclusions: Infective endocarditis is a significant burden in patients with congenital heart disease, carrying important morbidity and mortality. Changes in antibiotic prophylaxis regime did not result in increased incidence of SBE.

P-123

Target organs lesion in adolescents with essential arterial hypertension

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Aim: To assess the state of target organs in adolescents with essential arterial hypertension (EAH) at different stages of its formation.

Methods and Materials: 299 adolescents with EAH at the age from 12 to 18 years old (average age 14.9 ± 2.0) were examined: 215 youths and 84 girls. According to 24-hour blood pressure monitoring adolescents with EAH were divided into three study groups: first group – 98 adolescents with "white coat hypertension" (WCH), second group – 108 patients with labile AH (LAH), third group – 93 adolescents with stable AH (st AH). 27 persons were included into the control group. Left ventricle (LV) function were assessed according to Echo data. Brain MRI was performed using MRI scanner "Magnetom-OPEN". Endothelial dysfunction was assessed by means of plasma level of Von Willebrand factor (VF).

Results: LV geometry changes were disclosed in 23,1% of examine adolescents with EAH. Concentric hypertrophy was discovered in 2%, concentric remodeling – 6%, eccentric hypertrophy – 15%. Patients with st. AH had the possibility of eccentric hypertrophy formation higher in 3,13 times than in WCH group and in 2,7 times than in LAH group ($p = 0,0049$ and $p = 0,0076$, correspondingly). Hypertensive encephalopathy (HE) manifestations were discovered in 74% of adolescents with EAH. The difference of average sizes of arachnoid cavity (AC) of postcranial fossa in all study groups in relation to control with adjusted sex and age was clinically significant. Increase of average BP by 1 mm Hg per day and increase of time index of systolic BP at night by 1 unit (%) led to increase of AC postcranial fossa by 0,14 mm (CI 0,01:0,46), $p = 0,032$ and 0,09 mm (CI 0,01:0,16), $p = 0,028$,

correspondingly. Significant increase of WF concentration was marked in groups of liable and stable AH in relation to control group ($p = 0,0056$ and $p = 0,0018$, correspondingly). Higher differences of average WF and ILVMM activity were marked in patients with EAH and HE than in the group of patients without structural brain disturbances ($p = 0,025$ and $p = 0,01$, correspondingly).

Conclusion: Target organs lesion starts to form at earliest stages of EAH formation.

Key words: essential arterial hypertension, adolescents, target organs.

P-124

Experience of Harvard Step Test using for exercise tolerance assessment in children with single ventricle

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Aim: To present the assessment of exercise tolerance (ET) in children with single ventricle (SV) after total cavopulmonary shunt using modified Harvard Step Test (MHST).

Study Methods: MHST was performed to 37 patients at the age from 6 to 10 years old (average age $6 \pm 0,4$). The testee was proposed to climb a step during three minutes. Taking into account the patient's age it was considered to standardize the height of the step till 20 cm for all the groups. After the test was stopped the heart rate was assessed at the second, third and fourth minute during 30 seconds. MHST index is rated by formula: $tx100 / (f1 + f2 + f3) \times 2$, where t - climbing duration in seconds, $f1$, $f2$, $f3$ - heart rate at the second, third and fourth minute during 30 seconds of restoration, correspondingly. Condensed formula can be used in screening programs: $MHST = tx100 / fx5,5$, where t - climbing duration in seconds, f - heart rate taking into account the time of test carried out by the patients with heart failure (HF) signs.

Results: NYHA II (74%) predominated in most patients with SV. Initial heart rate corresponded to age norms. Heart rate increase was from 5 to 10% in 82% of pts with SV. Adverse response to exercise stress by way of heart rate decrease in 5-10% was marked in the rest of the children. According to study protocol the study was carried out in full in 67% of pts with SV. Test duration was lesser than necessary 3 minutes due to low ET. The reasons for stopping the test were muscle weakness and dyspnea in this group of patients. MHST varied from 24.1 to 75 in examined patients. The maximum test values were marked only in patients with test time 3 minutes.

Conclusion: Modified Harvard Step Test can be performed to preschool children and children of primary school age with SV after total cavopulmonary shunt to assess ET if spiroergometry is not possible.

Key words: single ventricle, exercise tolerance, modified Harvard Step Test.

P-125

Double outlet right atrium in AVSD: atrioventricular malalignment with different ventricular morphology

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Double outlet right atrium (DORA) is a rare form of atrioventricular malalignment with dislocation of the left atrium (LA) and

the atrial septum to the left and an absent connection of the LA to one of the ventricles.

We report about 2 neonates with DORA and AVSD. Common feature was an LA with regularly connected pulmonary veins, but absent connection to the left ventricle, whereby the LA drained exclusively via an ASD II into the right atrium (RA). Patient 1 showed an unbalanced ventricular morphology with a dominant left ventricle, pulmonary stenosis and left superior vena cava draining into the coronary sinus. Patient 2 presented with two balanced ventricles. Patient 1 was treated by univentricular palliation with BT-Shunt and closure of the pulmonary artery at the age of 8 weeks followed by a bidirectional Glenn-anastomosis at the age of six months. Patient 2 underwent a biventricular repair at the age of 2 months. This was accomplished by complete excision of the interatrial septum, patch repair of the AVSD and separation of the atria by an oblique patch, connecting the left atrium with the left ventricle.

Since DORA is the result of atrioventricular malalignment, patients with this malformation present either with two atrioventricular valves or as in our patients a common atrioventricular valve with a right and left portion. This is in contrast to mitral valve atresia and other forms of a absent left atrioventricular connection. The diagnosis is obtained by echocardiography. Since in patients with DORA the left atrium is able to drain its blood exclusively via an ASD, the issue of left atrial hypertension should be of major concern. Despite an absent left atrioventricular connection a biventricular repair is possible in selected patients with DORA. In our second case, who presented with two adequate size ventricles, corrective surgery was accomplished by oblique septation of the atria connecting the displaced left atrium with the left ventricle.

P-126

Evaluation of Cardiac Functions with Conventional and Tissue Doppler Imaging in Patients with Henöch-Schönlein Purpura

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Introduction: Henöch-Schönlein Purpura is a self-limiting systemic vasculitis. Cardiac involvement is very uncommon with documented involvement in HSP being limited to only a handful of case reports which are predominantly related with severe morbidity and mortality. The aim of this study is to investigate the cardiac functions of patients with HSP using tissue Doppler imaging.

Patients and Methods: Study was performed between December 2013 and January 2014 in Yuzuncu Yil University Faculty of Medicine in Turkey. 43 patients diagnosed as HSP and 32 healthy patients for control group were included. M-mode, two dimensional, Doppler and tissue Doppler echocardiography were performed for all subjects.

Results: Tissue Doppler imaging parameters showed left and right ventricular diastolic dysfunction in patients with HSP while there were not statistically significant differences in M-mode and two dimensional echocardiographic parameters between two groups (Table 1).

Discussion: Cardiac involvement is a rare but life threatening complication of HSP. For this reason cardiac scanning must be done early. Subclinical cardiac involvement in HSP may be more

Table 1. Mitral (m) and tricuspid (t) valve lateral annulus PW tissue doppler measurements

	Patient Group (n:43)	Control Group (n:32)	P
Em (cm/sn)	11,27 ± 1,86	12,62 ± 1,72	0,002
Am (cm/sn)	8,11 ± 1,62	7,28 ± 0,95	0,019
Em/Am	1,40 ± 0,14	1,72 ± 0,16	< 0,001
IVRTm (m/sn)	56,30 ± 7,15	50,90 ± 6,20	0,001
IVCTm (m/sn)	62,41 ± 7,07	59,81 ± 6,70	0,111
MPIm	0,37 ± 0,045	0,34 ± 0,042	0,005
Et (cm/sn)	10,58 ± 1,90	12,40 ± 1,43	< 0,001
At (cm/sn)	8,27 ± 1,24	7,40 ± 1,34	0,005
Et/At	1,46 ± 0,22	1,62 ± 0,22	0,004
IVRTt (m/sn)	57,27 ± 7,60	52,59 ± 6,11	0,005
IVCTt (m/sn)	66,04 ± 8,27	67,06 ± 7,61	0,588
MPIt	0,39 ± 0,037	0,36 ± 0,040	0,001

often than known. Tissue Doppler imaging may be a good diagnostic tool for detecting cardiac involvement of the disease especially when the cardiac symptoms are not clinically evident.

P-127

Abnormal laboratory findings prior to the diagnosis of protein-losing enteropathy after single ventricle strategy palliation

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Introduction: Protein-losing enteropathy (PLE) occurs in up to 15% of patients palliated surgically with single ventricle strategy and is associated with a 5-year survival rate of only 49–88%. Protein loss is often accompanied by lymphopenia, which may lead to development of serious infections that contribute to the overall mortality of PLE patients. Early and adequate diagnosis leads to earlier treatment implementation, which may prevent complications.

Aim of the study: Aim of the study was to assess if and to what extent abnormal laboratory findings may precede clinical diagnosis of protein-losing enteropathy.

Material and Methods: Between 1993 and 2013 13 patients with PLE following Fontan palliation were diagnosed in Children's Memorial Health Institute. The results of laboratory tests prior to the development of symptomatic PLE were available in 8 patients (50% male) with single ventricle (left in 4, right in 4). Data including serum protein level and lymphocyte count at the time of clinical diagnosis and during asymptomatic period was collected retrospectively. Progress of the disease was assessed using matched-pair analysis.

Results: Last procedure of surgical palliation was performed at the mean age of 5,1 years. Mean age at the time of PLE diagnosis was 8,9 years. In all patients abnormal laboratory findings suggesting a possible onset of PLE were found prior to the clinical symptoms – an average of 10,1 months earlier. The laboratory results were compared between both periods – prePLE and PLE: mean serum protein level decreased to 44,9 g/L and 40,11 g/L (NS), mean lymphocyte count was 1,5 G/L vs 1,05 G/L (p = 0,045), mean lymphocytes percentage in WBC differential was 26,9% vs 19,4% (p = 0,018). Lymphopenia (below normal for age) was found in 5 patients before and in 6 after PLE diagnosis.

Conclusions: Abnormal laboratory findings such as hypoproteinaemia and lymphopenia may precede PLE diagnosis and suggest

probable PLE onset. Collected data indicates increasing abnormalities during asymptomatic period, especially in the WBC differential. Therefore, in patients after Fontan palliation, we suggest considering performing common, low-cost blood tests – serum protein level and lymphocyte count – in order to seek possible onset of PLE and prevent complications.

P-128

Electrocardiographic changes as physiological adaptations in children and young adolescent athletes

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Introduction: Regular participation in intensive exercise is associated with specific electrical manifestations that reflect increased vagal tone and structural remodelling of the heart. Sport category is an important determinant of cardiac adaptation. Sports can be characterised as being high or low in dynamic and static components. Most of the literature concerns the effect of exercise in adult athletes.

This study aims to assess the influence of sports category on electrocardiographic findings in young athletes who participate in class IC (low static, high dynamic) and IIIA (high static, low dynamic) competitive sports.

Methods: Ninety-two young athletes (range 10 to 18 years; 91% male) were included: 50 soccer players (IC) and 42 competing in martial arts (IIIA): judo (24), karate (17) and taekwondo (1). Inclusion threshold was based on at least one year of training in competitive sports. They were investigated in outpatient sports clinic, by physician, licensed specialist in sports medicine. The 12-lead ECGs were analysed following recent guidelines on the interpretation of ECGs in athletes. For each sports group – class IC and class IIIA – prevalence of ECG changes was analysed; comparison between groups was assessed using two-way joining cluster analysis.

Results: Of examinees, standard 12-lead ECG with changes have had 58 athletes (58/92, 63%); in the group of soccer players 31/50 (62%), and in the martial arts group 27/42 (64.3%) (X² = 0.051, p = 0.8210). The most frequent ECG changes in both groups included sinus bradycardia, incomplete RBBB, and nonspecific intraventricular conduction disturbances. Other ECG changes were: sinus arrhythmia, atrial ectopic rhythm, first degree AV block, complete RBBB, isolated QRS voltage criteria for LVH, left atrial enlargement, early repolarization, repolarization abnormalities, long-QT interval, and supraventricular extrasystoles. With chi-square test there were no difference between the two groups for any of that changes (p ≥ 0.05). But, in soccer players group more athletes have had combination of changes (X² = 5.123, p = 0.0236).

Conclusions: In a study population of young athletes, ECG findings related to physiological cardiac adaptations are frequent. Different sport category didn't influence on specific type of ECG changes, but in a group of low static and high dynamic activity ECG changes were more noticeable.

P-129

Bicuspid aortic valve syndrome as a centre of congenital heart defects of left ventricle outflow tract

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Introduction: Although bicuspid aortic valve (BAV) is not included in epidemiological studies of congenital heart defects (CHD) in

children and is stated that the predictive factor for development of serious complications in BAV is the age over 30, our observations indicate that BAV is also a significant factor in CHD pathology in children. Here we tried to determine that significance.

Methods: All children examined in our institution during 11 years period (2000–2011) with diagnose of bicuspid aortic valve, isolated and/or associated with other congenital heart defects were included.

Results: We have found 229 BAV patients. The most common associated LVOT disorder was coarctation of aorta (32.75%). In our study 42.7% patients with an isolated coarctation had BAV. 29,7% of all BAV patients had aortic stenosis (AS), aortic insufficiency (AI), and/or ascending aortic dilatation (DAA). Percentage of BAV patients with isolated aortic stenosis or insufficiency was the same (7%), BAV with stenosis and insufficiency in 12.7%, AS and DAA in 9.17% patients. Of all the children with BAV, 62.44% had hemodynamic alterations on the aortic valve which manifested themselves as AS and/or AI. In our study progression of aortic stenosis was common finding during childhood while insufficiency mostly stayed mild. Ascending aortic dilatation showed progression with age in significant number of patients. Significant number of patient needed interventional and/or cardio-surgical treatment with increasing number of interventions with age, in accordance with the expected progression of pathological changes (AS, AI, DAA).

Conclusion: Bicuspid aortic valve is a congenital heart defect with a progressive development of hemodynamic changes in the left ventricular outflow tract which may become haemodynamically relevant in childhood age already. Therefore, BAV should be regularly controlled, and dilatation of the ascending aorta should be prevented with beta blockers or ACE-inhibitors. For all the mentioned reasons, we believe that bicuspid aortic valve should be included in the epidemiological studies of congenital heart defects.

P-130

Prognostic value of Troponin T/I in infants with hypoplastic left heart syndrome between stage I and II

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Introduction: The period between stage I and II procedure for treatment of hypoplastic left heart syndrome (HLHS) bears a high mortality and morbidity. Troponin T/I (Trop) is a well-recognized marker for myocardial damage. We sought to analyze the prognostic value of Trop for predicting outcome in infants with HLHS.

Methods: Retrospective analysis of 70 infants treated for HLHS at our institution between March 2001 and October 2014. Stage I procedure consisted of Norwood-operation in 35 (50%) and Hybrid-approach in 22 (31%) patients and was performed at a median age of 6 days (range 0–52). Palliative care was chosen for 13 (19%) patients. All available Trop values up to stage II were collected from clinical charts and were analyzed in relation to the overall outcome of the children.

Results: Trop was significantly higher after Norwood-operation in comparison to Hybrid-approach (median 7.1 µg/l (0.7–20.9), vs. 1.2 µg/l (0.3–17.9), p 0.0003).

Overall mortality of the treated patients was 39% (22 patients). Survival was 54% (19 patients) in the Norwood and 73% (16 patients)

in the Hybrid-approach group. Independently from the procedure used, maximal Trop and initial lactate levels were significantly higher in non-survivors than in survivors, with median Trop of 9 µg/l (0.6–18.8) vs. 3.4 µg/l (0.4–20.9), p 0.007, and median lactate of 3.7 mmol/L (1.6–25) vs. 2.9 mmol/L (0.3–14.6), p 0.03. Reinterventions between stage I and II were required in 17 (30%) patients, including 4 (11%) after Norwood and 13 (59%) after Hybrid procedure. No correlation was found between Trop levels and need for reinterventions.

Trop values were available during follow-up in the interstage period for 39 patients; Trop levels normalized in 5 (13%) patients, all survivors, while it remained elevated in all non-survivors.

Conclusions: In patients with HLHS Trop levels were significantly higher after Norwood procedure than after Hybrid-approach. Maximal Trop values were related to mortality.

During the interstage period normalization of Trop levels was infrequent, occurring in only 13% of the patients, all survivors. Trop levels did not correlate with the need for reinterventions.

P-131

Reduced pulmonary function in young adults operated for ventricular septal defect in early childhood: a long-term follow-up

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Introduction: Ventricular septal defects (VSDs) are normally closed in early childhood, and postsurgical the patients are generally considered healthy with normal physical capacity. Nevertheless, we have previously demonstrated reduced peak exercise capacity in this patient cohort, and therefore the aim of this study was to establish whether there are demonstrable abnormalities in pulmonary function during exercise.

Methods: We tested cardiopulmonary exercise capacity in 27 patients and 30 healthy control subjects on an ergometer cycle. Each test was preceded by a standard spirometry at rest, and the exercise test was performed as a maximal incremental test using an individually chosen test protocol. Pulmonary ventilation and gas exchange were simultaneously measured breath by breath with Jaeger MasterScreen CPX[®]. Our endpoints were minute ventilation and ventilatory equivalents (O₂ and CO₂) at peak exercise.

Results: In the VSD-group the median age at surgery was 1.9 (95% CI 1.1 – 2.8 years) and the age at the time of examination was 21.1 ± 3.1 years in the VSD-group vs. 21.2 ± 2.5 years in the control group. Minute ventilation at peak exercise was lower in the VSD-group compared with the controls: 1.4 ± 0.4 L/kg/min vs. 1.8 ± 0.4 L/kg/min, p < 0.01. Likewise oxygen uptake was reduced: 38.0 ± 8.2 ml/kg/min in the VSD-cohort vs. 47.9 ± 6.5 ml/kg/min among controls, p < 0.01. In terms of breath rate and ventilatory equivalents (O₂ and CO₂) there were no differences between the groups.

Conclusions: During exercise patients with a surgically closed VSD had substantially reduced minute ventilation despite uncompromised ventilatory efficiency compared to healthy controls.

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Evaluation of exercise capacity with cardiopulmonary exercise testing in adult patients with Fontan circulation

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Introduction: Reduction of functional capacity in patients that underwent Fontan's surgery is well known. Exercise intolerance in this group is directly related to increased morbimortality. In congenital heart disease patients a peak value of oxygen consumption (VO₂) <15 mL/kg/min or <50% of the predicted VO₂ has been recommended as a cut-off value for cardiac transplantation according to current ACC/AHA Guidelines.

Aims: To evaluate Fontan's patients' functional capacity using the cardiopulmonary exercise-test (CPET).

Methods/Results: Eleven patients (45% male, mean age 29 ± 5.9 years) that underwent CPET between 2010 and 2014 in a tertiary center dedicated to the follow-up of Adult Congenital Heart disease were included in our analysis. Statistical analysis was performed using R 3.1.0 software. Results were considered statistically significant if $p < 0.05$.

The most common type of surgery carried out was the cavopulmonary technique with an intracardiac conduit (55% patients). In the remaining, a classical Fontan operation or one of its variants was performed. The mean age at surgery was 8 ± 4.1 years. Two thirds of the population had Tricuspid Atresia (TA) with transposition of great vessels and one third, isolated TA or associated with pulmonary stenosis.

About 80% of the patients reported limitations to their physical activity, six were in NYHA class II and three were in NYHA Class III. The CPET in this population showed an average VO₂ of 20 ml/kg/min, with half of the patients in Class B Weber. Although approximately half of the patients (n = 5) were in Ventilatory Class I, 35% had a minute ventilation/carbon dioxide production (VE/VCO₂ slope) between 36 and 43 (ventilatory class III). The latter patients had a significantly higher incidence of thromboembolic events and arrhythmias. Heart rate (HR) reserve, was higher in patients who had undergone Fontan surgery at a younger age ($p < 0.05$; $R_2 = 0.4$).

Conclusions: The majority of patients had a moderate reduction in functional capacity. Patients with severe intolerance to exercise had a higher risk of major cardiac events. The completion of the Fontan surgery at a younger age is associated with higher HR reserve.

P-133

Abnormal aortic elasticity and aortic dilation progress with age and impact on LV mass in repaired tetralogy of Fallot
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Background: Aortic root dilatation and impaired aortic elasticity have been reported in children and adults with repaired tetralogy of Fallot (TOF). However, no study has described the changes of aortic elasticity with age and effects of abnormal aortic elasticity on left ventricular (LV) function.

Objectives: The aim of the study was to assess the hypothesis that elastic properties of aorta were altered with age and impaired aortic elasticity affected on LV function including LV mass in repaired TOF.

Methods: The subjects consisted of 46 repaired TOF (mean age: 9.4 ± 8.2 yrs) and 35 age-matched controls. Aortic dimension and aortic elastic properties such as the aortic strain (AoS), aortic distensibility (AoDis) and aortic stiffness index (AoSI) were obtained by cardiac catheterization. Aortic dimensions were expressed as z scores. LV mass was also obtained by LV angiogram and adjusted for height.

Results: Ascending aortic dimension was significantly increased in TOF compared with controls (Z score: TOF: 2.11 ± 1.07 vs. Control: 0.58 ± 1.21, $p < 0.01$, and increased with age in TOF. The AoS (%) and AoDis were significantly reduced in TOF (AoS: 12.9 ± 5.0%, AoDis: 0.008 ± 0.004 cm² dynes⁻¹ 10-6) than controls (AoS: 20.8 ± 5.0%, AoDis: 0.012 ± 0.004 cm² dynes⁻¹ 10-6, $p < 0.01$). The AoSI significantly increased in TOF (4.4 ± 2.2) than in controls (2.9 ± 1.4). Also, AoSI and AoDis weakly but significantly decreased with age ($r = 0.34$ in AoSI and $r = 0.24$ in AoDis). The LV mass index was increased in TOF (51.6 ± 14.6 g/BHx2.7) than that in control (35.7 ± 12.4 g/BHx2.7, $p < 0.01$). No significant correlation was found between parameters of aortic elastic properties and LV endo-diastolic volume and ejection fraction in TOF.

Conclusions: In repaired TOF, increased aortic root diameters progress and abnormal aortic elasticity changes with age. Increased aortic stiffness may produce the LV hypertrophy.

P-134

The risk of pregnancy in women with pulmonary hypertension Review of cohort studies in the advanced pulmonary hypertension therapy era

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Objectives: Mortality risk associated with pregnancy among women with pulmonary hypertension (PAH) was reported to be 17–33% in the era of advanced PAH therapy. Since most risk assessments were based on case reports and case series prone to bias, we aimed to review pregnancy associated risk in women with PAH who chose to go through pregnancy albeit contraindication in current cohorts.

Methods: Literature search included: English language Publications after 1996, addressing the risk of pregnancy in cohorts of women with PAH, with clear definition of PAH and outcome.

Results: Of 193 papers found, 6 (307 women) met inclusion criteria. Among 95 women, in 5 studies reporting mortality, 8 (8.4%) died and one required heart-lung transplantation. Mortality rates ranged between 3.3% and 17% between studies. The lowest mortality rate was reported in a centre with the largest average case number/year (5), and the highest mortality rate in centres with the lowest average cases number/year (0.24). Three women died during pregnancy, one at delivery and 4 within 6 weeks of delivery. Death was due to right heart failure in 4, hypotension in 2 and sepsis in one woman. PAH therapy was missing or suboptimal in all patients who died or required heart-lung transplantation. The average week of delivery was 33.4 to 36.1 with 50–100% by cesarean-section. No neonatal mortality was reported.

Two studies differentiated mild and severe PAH. There were 30 woman with mild and 42 with severe PAH. Women with severe PAH had higher risk of cardiac complications, NYHA class deterioration and increase in pulmonary pressure compared to women with mild PAH. Severe PAH was also associated with earlier delivery, higher risk of cesarean-section and lower birth weight.

Conclusions: According to limited current data available, women with PAH choosing to complete their pregnancy have < 10% maternal mortality on average. Women with mild PAH have better maternal and obstetric prognosis. Limited center experience and suboptimal therapy may contribute to adverse outcome.

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Right ventricular end-systolic volume as prognostic factor of right ventricular remodeling after pulmonary valve replacement

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Background: Optimal timing for pulmonary valve replacement (PVR) in pulmonary regurgitation (PR) in congenital heart disease (CHD) is a controversial topic, specially in asymptomatic patients. Most guidelines consider a RV end-diastolic volume (RVEDV) over 150 ml/m² as indication for PVR.

Methods: Retrospective analysis of clinical and cardiac MRI variables of asymptomatic CHD patients that underwent surgical PVR between September 2006 and February 2013. Only asymptomatic patients with both pre- and post-surgery MRI were included.

Results: Thirty-five patients (74,3% males) were included. Mean age at PVR was 25,8 years (SD=7,18) and mean weight was 64,5 kg (SD=12,03). Diagnosis: Tetralogy of Fallot (n=28), pulmonary atresia with VSD (n=2), primary PR (n=2) and PR after pulmonary stenosis treatment (n=2). Maximal RVEDV pre-PVR was 267 ml/m² and right ventricular end-systolic volume (RVESV) was 183 ml/m².

In the multivariate analysis we observed that RVEDV after PVR related with the width of the QRS complex and RVEDV pre-PVR (R²=0.335). We also observed a relationship between RVESV pre-PVR and the RVEF after PVR (R²=0.374, AUC=0.902). Our patients with a RVEDV up to 170 ml/m² combined with a RVESV less than 90 ml/m² achieved a good outcome, defined as RVEDV under 110 ml/m² (sensitivity 87.5%), RVESV under 55 ml/m² (sensitivity 100%) and RVEF over 50% (sensitivity 100%).

Conclusions: When deciding the optimal PVR timing in asymptomatic patients both RVEDV and RVESV should be considered. Larger prospective studies should be performed in order to establish the importance of REVSV in the timing for PVR and outcome of these patients.

Table 1. RV size and function were established by MRI

	Pre-PVR	Post-PVR	p
RVEDV (ml/m ²)	162 (SD=39.1)	94 (SD=23.6)	<0.001
RVESV (ml/m ²)	87 (SD=28.9)	44 (SD=15.7)	<0.001
RVEF	44.8% (SD=8.17)	52% (SD=9.9)	<0.001

RVEF = right ventricular ejection fraction.

Table 2.

	Post-PVR RVEDV (ml/m ²)	Post-PVR RVESV (ml/m ²)	Post-PVR RVEF
Pre-PVR RVEDV <170 ml/m ² and RVESV <90 ml/m ² (n=19)	84 (SD=15.8)	39 (SD=10.8)	54% (SD=8.2)
Pre-PVR RVEDV >170 ml/m ² and RVESV >90 ml/m ² (n=10)	105 (SD=23.9)	54 (SD=18.1)	48% (SD=10.4)

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The Influence of Aortic Dilatation and Aortic Valve Regurgitation for Plasma Brain Natriuretic Peptide Level in Patients with Fontan Procedure

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Objective: The Fontan procedure has improved the morbidity and mortality of patients with single ventricular morphology. Several reports showed that plasma brain natriuretic peptide (BNP) levels were elevated in late after Fontan procedure with systemic ventricular failure. However, mechanisms of elevation of plasma BNP levels in Fontan patients were still unknown. The objective of this study was to evaluate plasma BNP levels with long-term after Fontan procedure, and reveal relationships with cardiac magnetic resonance (CMR) parameters.

Methods: Ten Fontan patients (median age 18.0 years, range 12.7 - 39.3 years, 7 males, 3 females, median follow-up 13.8 years, range 10.8-24.8 years) underwent BNP analysis and CMR.

Results: Median level of plasma BNP was 14.7 pg/mL (range ≤0.2 - 418.4 pg/mL). In CMR parameters, mean end-diastolic volume index (EDVI) was 81.7 ± 21.3 mL/m², end-systolic volume index (ESVI) was 36.2 ± 12.4 mL/m², ejection fraction (EF) was 56.4 ± 6.9%, ascending aortic diameter (AOD) was 25.5 ± 4.7 mm, and aortic valve regurgitation (AR) was 11.7 ± 12.7%. Eight patients had New York Heart Association (NYHA) class I, and 2 patients had NYHA class II. Plasma BNP levels were positively correlated with AOD (r=0.685; P=0.014), AR (r=0.697; P=0.013) and NYHA functional class (r=0.609; P=0.031). Other CMR parameters (EDVI, ESVI, and EF) were not correlated with plasma BNP levels.

Conclusion: The plasma BNP level may be associated with aortic dilatation and the severity of AR long-term after Fontan procedure. To improve the prognosis of Fontan patients, we should pay more attentions to aortic dilatation and aortic valve regurgitation.

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Circulating blood volume positively correlates with cardiac output in Fontan patients

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Introduction: Patients with chronic heart failure (CHF) are known to have more amount of circulating blood volume (CBV) than patients with normal heart. Fontan patients are characterized by low cardiac output, high central venous pressure (CVP), and high neurohormonal activity (NHA), like in patients with CHF. There have been few reports about the CBV of Fontan patients. The aim of this study is to examine the usefulness of CBV in interpreting hemodynamic parameters and NHA in the Fontan patients.

Methods: Between June 2013 and December 2014 we evaluated CBV in twenty six Fontan patients aged 14.3 - 50.8 years (median 24.4 years). The patients after extracardiac rerouting type of Fontan operation are 54% of all patients, intraatrial grafting are 27%, atriopulmonary connection are 12% and intraatrial rerouting are 7%. The CBV was measured by pulse dye densitometry using indocyanine green during the cardiac catheterization. CBV indices (CBVI, CBV/body surface area) were compared with hemodynamic parameters (cardiac output (CO), systemic vascular resistance (Rs), and CVP) obtained during the cardiac catheterization

and plasma norepinephrine level (NE), plasma dopamine level (DA), logarithm of atrial and brain natriuretic peptides (log ANP and log BNP) as the NHA.

Results: The CBV of Fontan patient was 2.53–8.62 L (median 4.22 L) and CBVI was 1.86–4.76 L (median 2.39 L/m²). There are no relation between CBVI and type of Fontan operation. CBVI had significant negative correlation with log BNP ($p < 0.05$), NE ($p < 0.01$) and DA ($P < 0.05$). Moreover, CBV was found to have positive correlation with CO in our series, which might suggest the different mechanism from conventional CHF lies under the low output and high CVP state in Fontan circulation.

Conclusion: In our study, CBV of Fontan patient has negative correlation with NHAs and positive correlation with CO.

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Mechanism of desaturation caused by posture and exercise in Fontan patients

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Introduction: Mild desaturation is often observed and its degree may vary depending on the posture, exercise, and the location of right to left shunt in patients after Fontan procedure.

The purpose of this study was to evaluate the degree of oxygen saturation change with posture and exercise, and to investigate the effect of hemodynamic variables and location of right to left shunt.

Methods: Retrospective chart review was carried out on consecutive patients older than 16 years old with Fontan circulation and had cardiac catheterization between 2012 and 2013 ($n = 43$). Percutaneous oxygen saturation (SpO₂) measured at supine position and standing was compared, and patients were divided into two groups; $\Delta\text{SpO}_2 < 4\%$ and $\Delta\text{SpO}_2 \geq 4\%$. Patients were also divided according to the ΔSpO_2 during 6 minutes walk (6MW). Correlations between SpO₂, catheterization data, contrast echocardiogram finding, and location of right to left (RL) shunt were evaluated.

Results: Catheterization data, NYHA functional class upon 6MW, location of RL shunt, and contrast echocardiogram findings are shown in the Table 1. In general, supine SpO₂ ($94.0 \pm 4.0\%$) was significantly higher than standing SpO₂ ($91.7 \pm 4.2\%$). There was no significant difference in hemodynamic parameters and shunt locations between two groups. SpO₂ become significantly lower during 6MW (91.9 ± 3.9 vs. $88.5 \pm 5.8\%$). Patients with $\Delta\text{SpO}_2 \geq 4\%$ had lower SaO₂ at baseline, higher Rp, and higher incidence of RL shunt from innominate vein than those with $\text{SpO}_2 < 4\%$ during 6MW.

Conclusions: Underlying mechanism causing SpO₂ changes in Fontan patients by posture and exercise may be different. Desaturation during exercise is more likely to be related to pulmonary circulation and RL shunt.

Table 1.

	Postural change				6 MW			
		$\Delta\text{SpO}_2 < 4\%$	$\Delta\text{SpO}_2 \geq 4\%$	p	$\Delta\text{SpO}_2 < 4\%$	$\Delta\text{SpO}_2 \geq 4\%$	p	
Cath data	SaO ₂ (%)	93.6±3.4	93.5±4.7	N.S	94.4±3.8	92.3±3.6*	0.008	
	CVP (mmHg)	13±2	14±5	N.S	13±4	14±3	N.S	
	C.I (L/min/m ²)	2.3±0.6	2.5±0.6	N.S	2.3±0.4	2.3±0.7	0.89	
	Rp (wood)	2.2±1.3	2.2±1.3	N.S	1.8±0.9	2.8±1.7*	0.028	
NYHA	I (600m<)	0	1	χ ²	1	0	χ ²	
	IIs (450-600m)	18	10*	0.047	18	6	N.S	
	IIIm(300-450m)	9	1		4	10		
RL shunt	Innominate V	22/30	9/12	N.S	21/25*	8/15	0.032	
	Baffle	10/31	4/12	N.S	10/24	4/15	N.S	
	hepatic/IVC	9/30	2/12	N.S	6/24	5/15	N.S	
	injected at PA +	11/22	4/12	N.S	8/21	7/12	N.S	

CVP; central venous pressure, C.I; cardiac index, Rp; pulmonary resistance, IVC; inferior vena cava, PA; pulmonary artery

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Why do they survive? Native congenital heart disease in a grown-up African population without access to care

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Objectives: To describe 104 patients over 13 years old with congenital heart disease (CHD) and without access to corrective surgery or intervention.

Methods: Case notes of all patients seen at Windhoek Central Hospital between January 2009 to December 2013 were reviewed for congenital heart disease. Clinical features, ECG, chest x-ray, echocardiogram, cardiac catheterisation, operation reports and follow-up records were checked. Diagnosis was established by echocardiogram. Surgery was conducted at Windhoek Central Hospital or, the Christiaan Barnard Memorial Hospital in South Africa.

Results: One hundred seven (107) patients were identified. Three received surgery prior to presentation and were excluded. In 104 patients age ranged from 13 to 86 years (mean 25). 68 were female. Pathology included, VSD 30, secundum ASD 32, PDA 12, AVSD 10 (9 primum ASD), AS 4 and coarctation of aorta 5. Cyanotic disease included Tetralogy of Fallot 7 (TOF), DORV with TOF 4, PS 7, Ebstein's 2 and TGA/VSD/PS 2.

Seventy eight were deemed "operable". Thirty-one had cardiac catheterisation. There were 7 interventions for, coarctation (2), PDA (2), ASD (2) and valvular PS (1). Surgery was performed on 52; VSD 15, ASD 2 16, PDA 5, primum ASD 5, TOF 6 (3 with DORV). Three with complex disease were repaired. Eleven were inoperable (8 Eisenmenger syndrome).

Over a 5 year follow-up (mean 32 months), three patients have died. Two with inoperable secundum ASD and one after shunt surgery for TOF with pulmonary atresia. Fifteen patients have been lost to follow-up.

Conclusions: This cohort reflects a time when there were no diagnostic or curative services for CHD anywhere. A surprising number of patients with large but "simple" lesions (VSD, ASD and PDA) were still operable, albeit with advanced pulmonary hypertension and congestive cardiac failure. Secondly, there is a sub-set of patients with Tetralogy of Fallot who survive to adulthood where the anatomy and therefore the surgical challenge, is profoundly different from that in the infant. The high number "lost" reflects complex health systems challenges in a vast, low middle income country.

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Low-dose dual-source MDCT angiography imaging of partial anomalous pulmonary venous connection in pediatric patients

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Objective: To determine lobar distribution, drainage sites and associated cardiovascular anomalies of partial anomalous pulmonary venous connection (PAPVC) in pediatric patients using 256-detector multidetector computed tomographic (MDCT) angiography.

Material and Methods: 61 cases [27 female, mean age 4.7 years (13 days–15.4 years)] with PAPVC diagnosed by MDCT angiography were included in this study. In all patients, MDCT examinations were performed using a dual-source 256-MDCT scanner (Definition Flash, Siemens Healthcare) with high pitch protocol.

Results: In 61 patients, 73 anomalous pulmonary vein (APV)s were detected, 56 of them (77%) were right sided and 17 (23%) were left. 50 patients (82%) had only one APV, and 11 (18%) had two APVs. In 49 patients with 56 right sided APVs, 38 (68%) draining into superior vena cava, 8 (14%) into atrio-caval junction, 6 (11%) into inferior vena cava, 3 (5%) into right atrium, and 1 (2%) into levoatriocardinal vein. In the 12 patients with 17 left sided APVs, 16 (94%) draining into left innominate vein and 1 (6%) into coronary sinus. Only 7 (12%) patients had isolated PAPVC, whereas 54 (88%) patients had additional cardiovascular anomalies. The most common (n = 40, 66%) associated anomaly is an atrial septal defect (ASD). In patients with right APVs, 26 (53%) had superior sinus venosus type of ASD, 6 (12%) had secundum type ASD, and 3 (6%) had inferior sinus venosus type of ASD. The other associated cardiovascular anomalies are ventricular septal defect (n = 8), persistent left superior vena cava (n = 8), patent ductus arteriosus (n = 4), aortic coarctation (n = 3), hypoplastic left heart syndrome (n = 2), truncus arteriosus (n = 1), double outlet right ventricle (n = 1), cor triatrium sinister (n = 1), aberrant right subclavian artery (n = 1), and levoatriocardinal vein (n = 1). In 48 (79%) patients, PAPVC and associated cardiovascular anomalies were corrected surgically. The diagnostic agreements between MDCT and surgical results were 100%. The overall mean effective radiation dose was 1.12 mSv (range, 0.15–6.13 mSv), and it was 0.58 mSv (range, 0.15–0.73) in the patients younger than 1 year old.

Conclusion: The presence and course of the anomalous pulmonary veins and associated cardiovascular anomalies can be reliably detected by 256-slice MDCT angiography with lower radiation doses.

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Echocardiographic accuracy in detecting coronary pattern in transposition of the great arteries: a 10-year single center experience

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Introduction: Re-implantation of coronary arteries is a key factor in arterial switch operation. Preoperative assessment of coronary artery anatomy in transposition of the great arteries (TGA) is a crucial issue for planning surgical technique. Overall, patients (pts) with any variant coronary pattern have nearly doubled mortality. Echocardiography is the main diagnostic tool for the preoperative assessment of coronary anatomy in TGA.

Aim: Primary aim of our study was to retrospectively review our 10-years experience and to evaluate our echocardiographic preoperative accuracy in detecting coronary anatomy pattern comparing echocardiographic to surgical findings. Secondary aim was to determine association between post-surgical outcome and coronary pattern.

Methods and Materials: We retrospectively reviewed echocardiographic, operative and medical records of TGA cases referred to our institution for primary surgical repair between January 2004 and January 2014. Echocardiographic coronary anatomy descriptions were compared to surgical and/or autopsy findings to establish diagnostic accuracy.

Results: We identified 181 pts with TGA who underwent surgery from January 2004 to January 2014. Coronary pattern were described as “usual” for TGA in 111 cases (61%) in which surgical agreement was 95% (106/111pts); unusual coronary patterns were described in 70 pts (39%) and were confirmed at surgical time in

87% of cases (61/70pts). Overall 10 pts had intramural coronary artery, in whom 8 pts were identified by pre-operative echocardiography (80%).

Failure to detect correct coronary pattern by echocardiography was observed in 14 cases. Echocardiographic diagnostic accuracy in identifying intramural coronaries was 80% (sensitivity 88%, specificity 98%) but reached 100% in the last 6 years. Sensitivity and specificity for anomalous circumflex artery were 92% and 98%. Intraoperative mortality was 0%; early mortality at 60 days was 1%.

Conclusions: Echocardiography is a reliable tool in detecting coronary anomaly in TGA with an overall accuracy of 92%, including more complex TGA patterns and even in the presence of intramural course.

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Impaired myocardial deformation in asymptomatic patients with isolated left ventricular non-compaction

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Objective: The aim was to determine the early regional and global myocardial functional changes and whether the myocardial changes that cannot be detected by conventional echocardiography could be detected by tissue Doppler imaging (TDI) or two-dimensional speckle-tracking echocardiography (STE) in children with isolated left ventricular non-compaction (iLVNC) without symptoms.

Methods: Myocardial velocities and time intervals, longitudinal and circumferential strain (S) and strain rates (SR) determined by TDI and STE in twenty children with iLVNC aged 12.1 years was compared with those in twenty controls. All children underwent echocardiographic assessment using two-dimensional, M-mode, tissue Doppler and speckle-tracking echocardiographies. The myocardium was assessed at three segments; non-compacted (NC), neighbouring NC (NNC) and compacted (C), according to wall characteristics.

Results: According to TDI; isovolumic contraction time (100.5 vs. 69.5, 80.3 vs. 65.5 and 77.3 vs. 65.6 cm/s), isovolumic relaxation time (87.9 vs. 74, 76.7 vs. 66 and 71.9 vs. 63.5 cm/s) and myocardial performance index (0.95 vs. 0.51, 0.7 vs. 0.49 and 0.59 vs. 0.48) were significantly higher, while ejection time (198 vs. 275.8, 224.7 vs. 265.8 and 250.7 vs. 269 ms) were significantly lower in children with iLVNC, at all three segments. According to STE; longitudinal S (-14.4 vs. -23.2 and -18.1 vs. -24.9%) and SR (-0.38 vs. -0.61 and -0.43 vs. -0.83 s⁻¹) and also circumferential S (-19.1 vs. -27.8 and -24 vs. -29.3%) and SR (-0.37 vs. -0.87 and -0.47 vs. -0.85 s⁻¹) were significantly lower in children with iLVNC, in NC and NNC segments. The global longitudinal and circumferential S and SR at all three segments were significantly lower in children with iLVNC. Global longitudinal S were -18 vs. -23.8% and global longitudinal SR were -0.56 vs. -0.84 s⁻¹. Global circumferential S were -24.6 vs. -27.5% and global circumferential SR were -0.63 vs. -0.96 s⁻¹.

Conclusion: Both TDI and STE used to evaluate myocardial function and deformation could detect myocardial dysfunction and impaired deformation in children with iLVNC who are subclinic and whose left ventricular functions are normal by conventional methods.

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The evaluation of myocardial function by tissue Doppler imaging in neonatal sepsis

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Objective: The aim of this study was to determine the cardiac effects of sepsis by tissue Doppler echocardiography in neonates.

Methods: The myocardial velocities and time intervals at inter-ventricular septum (IVS) and left ventricular posterior wall (LVPW) before and after therapy were compared in 20 neonates with sepsis and 20 age and gender matched healthy neonates. The exclusion criteria were elevated cardiac enzymes and systolic dysfunction at conventional echocardiography, accompanying heart, lung or metabolic disease.

Results: The male/female ratio was 1.9 and mean age of patients was 21.7 days. The mean sepsis score used by Pennsylvania Hospital was 3.3 in patients. There were no significant differences for LVDd, EF and FS values of patients and controls before and after therapy. Myocardial velocities; Sm (3.7 vs. 5.5 cm/s) and Em (4.9 vs. 7.9 cm/s) at IVS and Sm (4.3 vs. 5.8 cm/s), Em (6.1 vs. 9.1 cm/s) and Am (5.1 vs. 6.9 cm/s) at LVPW were significantly lower ($p < 0.0001$) in patients before therapy. Time intervals; isovolumic contraction time (ICT) (59.4 vs. 43.7 ms), isovolumic relaxation time (IRT) (52.6 vs. 41.5 ms) and ejection time (ET) (147.6 vs. 182.4 ms) at IVS and ICT (59.4 vs. 43.1 ms), IRT (53.4 vs. 41.7 ms) and ET (143.8 vs. 187.1 ms) at LVPW were significantly longer ($p < 0.0001$) in patients before therapy. However, there were significant improvements ($p < 0.05$) for Sm (4.7 cm/s), Em (6.6 cm/s), ICT (48.6 ms), IRT (46.1 ms) and ET (161.3 ms) at IVS and Sm (4.8 cm/s), Em (7.4 cm/s), Am (5.6 cm/s), ICT (48.2 ms), IRT (48.9 ms) and ET (164.3 ms) at LVPW after therapy in patients. The myocardial performance index (MPI) at IVS (0.77, 0.59 vs. 0.47) and LVPW (0.79, 0.61 vs. 0.45) were significantly higher in patients before ($p < 0.001$) and also after ($p < 0.01$) therapy. However, there were no correlations between sepsis scores and MPI values at both IVS and LVPW.

Conclusions: The results of this study revealed that sepsis causes myocardial depression in neonates even though the left ventricular systolic functions are normal with conventional transthoracic echocardiography. This depression resolves with therapy in a considerable time period.

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A Novel Echocardiographic Quantification of Pulmonary Valve Regurgitation in Patients with Tetralogy of Fallot

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Introduction: The pulmonary regurgitation (PR) is the most important cause of the right ventricular failure and dilatation in patients long after the definitive surgery for tetralogy of Fallot (TOF), however its quantification of PR is difficult and not standardized by echocardiography but could be measured by cardiac magnetic resonance (CMR). We aim to quantify PR in TOF using a novel echocardiographic imaging modality, Vector-Flow Mapping (VFM) and to validate by CMR using phase-contrast analysis.

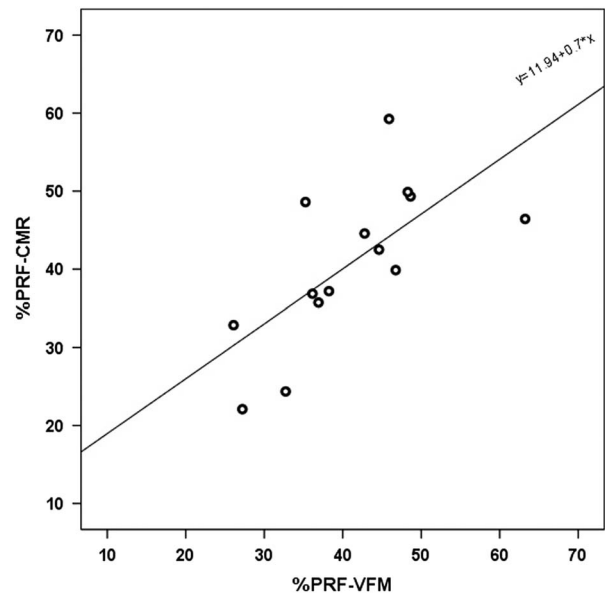


Figure.

Subjects and Methods: fourteen patients with repaired TOF, mean age 17 y (2.8–31 y), and mean interval from the definitive surgery 15 y (1.7–26 y). The color Doppler images of right ventricular outflow view including pulmonary artery (RVOT) was obtained by Prosound F75 (Hitachi-ALOKA Ltd) with VFM mode, and calculated the both-directional flow across the pulmonary valve as a sum of by integration of multiple sampling gate set on the line along the cardiac cycle by off line analysing program. The ratio of regurgitation to antegrade flow of VFM (PRF-VFM) was compared to those of CMRI using phase-contrast flow quantification (PRF-CMR). PRF-VFM was also evaluated by the previous reported indices such as the presence of diastolic reverse flow in peripheral PA branches, PR pressure half time < 100 ms, and PR index < 0.77 .

Results: PRF-VFM (40.9 \pm 9.8%) was well correlated with PRF-CMR (40.7 \pm 10.2%) ($r = 0.68$, $p < 0.01$) with good agreement between the two measurements. PRF-VFM $> 40\%$ had good sensitivity (85.7%) and specificity (85.7%), if PRF-CMR $> 40\%$ was considered as a severe PR, however other parameters showed higher sensitivity (71–100%) but poor specificity (14.3–33%).

To assess the severity of PR, the quantification of PR is mandatory with good sensitivity and specificity. Our new imaging modality of PRF-VFM demonstrates a good agreement with the results of PRF-CMR than any other parameters previously reported. The limitation of this technique is the quality of images of RVOT and color Doppler signal and the turbulent flow by the stenosis > 3 m/s.

Conclusions: PRF-VFM is feasible and handy diagnostic imaging with quantification for assessing the severity of PR in patients with TOF, which is clinical valuable.

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The impact of diastolic flow kinetic energy loss of the left ventricle on cardiac output (Q) in patients with tetralogy of Fallot

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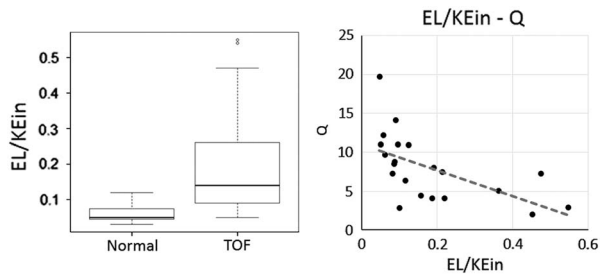


Figure.

Introduction (or Basis or Objectives): The impact of diastolic flow energetics on cardiac output remains unknown. We investigated the impact of intra-cardiac blood flow kinetic energy loss (EL) on cardiac output in patients after the definitive surgical repair of tetralogy of Fallot (TOF), by using a novel echocardiographic imaging modality of Vector Flow Mapping (VFM).

Methods: The subjects were 24 pts with TOF (age; 13.9 ± 7.4 y, interval from surgical repair; 143 ± 64 months) and 16 normal healthy children (8.9 ± 4.6 y).

The three chamber view of the left ventricle (LV) with color Doppler was recorded by Prosound F75 (Hitachi-ALOKA, Ltd) with VFM mode. Then we calculated EL (mW) from the reconstructed velocity vector components transformed into Cartesian coordinate system as previously reported by Itatani K. (Jpn J Appl. Phys 2013;52:07HF16). The EL data were indexed by measuring a ratio of EL to the inflow kinetic energy (KEin) through mitral valve in diastole. Cardiac output (Q) was calculated from velocity-time integral and cross-sectional area of LVOT. Results: EL/KEin was significantly higher in TOF (0.21 ± 0.16) than in N (0.06 ± 0.03). In TOF, EL/KEin had a negative correlation with Q ($R^2 = 0.54$, $p = 0.000231$).

Conclusions: The diastolic blood flow energy loss contributes to cardiac output (Q) in TOF.

P-146

Should we believe in peak velocities? Comparison of peak velocities determined by 2D phase-contrast MRI with those assessed by real-time phase-contrast MRI and pulse wave echocardiography?

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Introduction: Determination of peak velocities is an important parameter for estimation of the severity of stenosis, analysis of arterial stiffness and assessment of the pulmonary flow pattern. Magnetic resonance imaging (MRI) is a noninvasive tool for evaluation of aortic distensibility and PW velocity based on its high accuracy and reproducibility. MRI is known to underestimate peak velocities due to the typical long data acquisition times, echocardiography is considered as operator dependent. The aim of this study was to compare peak velocities (Vmax) assessed by two-dimensional (2D) phase-contrast velocity mapping (PC-MRI), real-time PC-MRI and pulse wave echocardiography regarding the degree of discrepancy.

Material and Methods: Quantitative through-plane PC-MRI flow/Vmax measurements were performed in the ascending aorta of 11 healthy subjects (mean age = 42.6 ± 13.7 years; 6 male) with a 3 T scanner. Using (a) retrospectively gated standard 2D PC-MRI,

(b) 2D PC-MRI (comparable to a) but with flow encoding in 3 spatial directions for 3D vector calculating); (c) real-time PC-MRI; (d) pulse wave echocardiography. Measurements were performed without repositioning of the subject. Normally distributed data were analyzed by paired Student-t-test regarding a p-value of 0.05 as statistically significant, Bland-Altman statistics and by calculating of regression coefficients.

Results and Discussion: Average Vmax (MR_3D) exceeded Vmax (MR_ref) by 4.4% (= mean, limits-of-agreement: +14.1% to -5.3%, $r \sim 0.96$, $p < 0.05$). This may be related to the occasionally non-perpendicular blood flow direction in a curved vessel underscoring the need for calculating of three-dimensional velocity vectors. Peak velocities were higher applying real-time PC-MRI compared to MR_ref (28.1%; +51.4% to +4.9%, $r \sim 0.64$, $p < 0.05$), which can be explained by the non-averaging character of this technique representing a snap-shot of the actual blood flow in the region-of-interest (Abb. 1). Accordingly, Vmax (echo) was increased by 25.2% in relation to Vmax (MR_ref) (+69.0% to -18.5%, $r < 0.5$, $p < 0.05$). Comparing real-time PC-MRI and pulse wave echocardiography high agreement was observed between both methods (2.7%; +44.5% to -39.2%, $r < 0.5$, $p < 0.05$) but with substantial scatters and a low correlation coefficient.

Conclusion: Peak velocities assessed by standard quantitative two-dimensional PC-MRI were underestimated by 25–30% in comparison to real-time PC-MRI and pulse wave echocardiography.

P-147

Analysis of the right ventricle by 4D-RV-Function 2 in healthy children and young adults – first results

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Introduction: Echocardiographic quantification of right ventricular (RV) function is limited if only a standard two dimensional approach is used. Realtime 3D-Echocardiography (RT3DE) is a promising method to get more valid data especially for end-diastolic (EDV), endsystolic (ESV), stroke (SV) volumes and ejection fraction (EF). Simultaneous assessment of deformation parameters as longitudinal Strain (LS) within the same dataset would be of additional value.

Methods: 20 healthy children and adults (6–40, median 16.4 ys, 13 female) were consecutively investigated by cardiac magnetic resonance imaging (CMR, 3 T Achieva, Philips, Cine SSFP, frame rate 20–24/s) and RT3DE (Vivid 7, IE33, 4-beat acquisition, frame rate 20–30/s). CMR data were analysed using the summation of disks method, RT3DE data were processed using a prototype of the 4D RV-Function 2 software (Tomtec, Germany) by two separate expert investigators blinded to each other. For interobserver variability a third investigator was introduced. Statistical analysis by Bland Altman defining differences in % and including limits of agreement (LOA), correlations by Pearson-Bravais.

Results: Feasibility of 90% (18 RT3DE datasets). Enddiastolic volumes (EDV) calculated by CMR ranged from 60–197 ml. RT3DE provided mild mean underestimation of EDV ($4.5 \pm 8.5\%$, LOA -12.5% to 21.5%, $r = 0.990$), ESV ($1.4 \pm 15.4\%$, LOA -29.5% to 32.3%, $r = 0.951$), SV ($6.5 \pm 12.4\%$, LOA -18.3% to 31.3%, $r = 0.974$) and EF ($2.0 \pm 8.9\%$, LOA -15.8% to 19.8%, $r = 0.969$). Intraobserver-variability for EDV ($-1.7 \pm 8.2\%$, LOA -18% to 14.6%, $r = 0.979$) and ESV ($2.9 \pm 13.2\%$, LOA -23.5% to 29.3%, $r = 0.928$) was low. Interobserver-variability for EDV ($0.8 \pm 15\%$,

LOA -29.1% to 30.8%, $r=0.928$) was better than for ESV ($2.0 \pm 24\%$, LOA -46% to 50%, $r=0.887$). Mean septal LS was $-21 \pm 5.9\%$, free wall LS revealed higher values of $-29 \pm 6.5\%$ ($p < 0.001$). Mean calculation time for RT3DE was 5 minutes per dataset.

Conclusions: RV-Function 2 provides good feasibility and promising accuracy within a wide range of volumes of healthy individuals. Reproducibility of ESV estimation was acceptable but less good than of EDV. Deformational parameters can also be assessed within a short period of calculation time from the same RT3DE dataset. Further validations have to be carried out in order to undermine that the method is an interesting tool for clinical use in patients.

P-148

Are organ size and heart rate advantageous measures for more precise clinical use of Velocity Time Integral in children?

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Introduction: Integration of doppler curves is an often used means of echocardiographic estimation of cardiac output. To compensate for maturational changes in children, physiological factors as heart rate, ventricular length and the Frank Starling mechanism have to be considered. Our aim was to create percentiles for aortic (VTIAo) and pulmonary velocity time integral (VTIPa) that are useful in clinical practice.

Methods: 377 echocardiographic datasets recorded in healthy children (0–20 years) were analysed. Measurement of left ventricular length, VTIAo (group I) respectively right ventricular length, VTIPa (group II) and heart rate (HR) was performed. To assess practicability 40 patients (0–6 years) with shunt lesions (group A: patent ductus arteriosus (PDA); group B: atrial septal defect (ASD)) were investigated and values integrated into the percentiles. Sensitivity of this method was compared to sensitivity of other reference values for VTIAo (C. Pees, *Pediatr Cardiol* (2013) 34: 1194–1200).

Results: Feasibility was 91% for group I (330 datasets: 166 male, 164 female) and 86% for group II (324 datasets: 164 male, 160 female). VTI/HR and ventricular length provided excellent correlations in children younger than 7 years (group I: $r=0.824$; group II: $r=0.772$). Adolescent individuals (7–20 years) showed lower correlation because of higher stroke volume variability (group I: male $r=0.462$, female $r=0.458$; group II: male $r=0.521$, female $r=0.456$). With the percentiles created more known aortic hyperperfusion (group A) could be detected (50% >2 standard

deviation (SD); 100% >1 SD) than with reference data of C (35% > 2 SD; 50% > 1 SD). Sensitivity of percentiles for VTIPa (group B) was also satisfactory (65% > 2 SD; 90% > 1 SD).

Conclusions: These reference values might facilitate the differentiation between normal and pathological VTI especially in young children.

P-149

Comparison of Basic Echocardiogram and Cardiac Magnetic Resonance of the Right Ventricle in Repaired Tetralogy of Fallot

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Background: Cardiac magnetic resonance (CMR) has become a standard tool to evaluate the right ventricle (RV). In repaired tetralogy of Fallot (rTOF), the right ventricular volume load is considered to be one of the key criterions for the pulmonary valve replacement. The objective of this study is to define whether the conventional echocardiographic measurement could be use as a parameter to define the right ventricular dilatation in comparison with the CMR measurement.

Methods: Patients with rTOF underwent CMR and echocardiogram. From the 4-chamber view, the RV dimension was measured in long axis and short axis during diastole and indexed by body surface area. The RV echocardiographic measurement was compared and correlated with the RV volume index obtained from CMR. The sensitivity and specificity of the echocardiographic threshold value predict the RV volume were determined.

Results: A total of 99 patients (16.2 ± 5.5 years, 69 male) were recruited. The echocardiographic measurement of RV end diastolic diameter (RVEDD) index and RV long- axis length index were correlated with the RV end diastolic volume index (RVEDVi) obtained by CMR ($r=0.47$, $p=0.01$ and $r=0.27$, $p=0.01$). The RVEDD index > 1.91 cm/m^2 had 75% sensitivity and 80% specificity to predict RVEDVi > 160 mL/m^2 with area under the curve of 0.81. While the RV long- axis length index > 5.10 cm/m^2 had 75% sensitivity and 50.8% specificity to predict RVEDVi > 160 mL/m^2 with area under the curve of 0.66.

Conclusion: The basic echocardiogram parameter of the RV dimension could be used to assess the right ventricle volume load in rTOF with reasonable CMR correlation. The RVEDDi has a better prediction of the RV volume in comparison with the RV long-axis length index.

P-150

Measuring Exercise Capacity in the Very Young Child

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Introduction: Exercise testing is an important cardiovascular assessment tool. Although there are well established protocols/equipment for older children there is limited knowledge for testing in small children. The aim of this project was to develop and test a bicycle ergometer as a means of testing exercise capacity in very young children.

Methods: Healthy children aged between 4–6 years were invited to participate. The cycle ergometer was a specifically adapted children's bicycle, connected to a load resistor and current sensor. The bike was also connected to a 'visual incentive' in the form of a

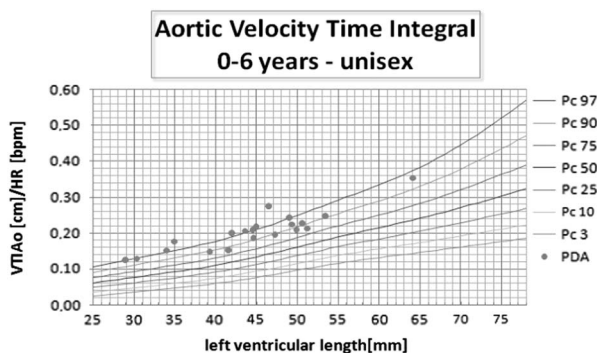


Figure.

Table 1. Patient Demographics and Exercise Stress Test Results Summary

	Mean (s.d.)	Interquartile Range
Age (years)	5.8 (0.66)	5.5 - 6.2
Weight (kg)	22 (3.6)	19.8 - 23.8
Height (cm)	113.6 (5.8)	109 - 118
Maximal Heart Rate (bpm)	165 (15)	159-173
% Max Predicted HR (%)	77 (6.9)	74-81
Exercise Duration (mins)	8.08 (1.1)	7.88-8.53
Average Power Over Test (Watts)	19.5 (6.7)	15.2 - 22.5
Maximal Power Attained (Watts)	104 (45.4)	83 - 109

LED light display. Each child was encouraged to exercise until exhaustion. The exercise protocol was a multistage, incremental protocol with increasing resistance every 3 minutes and two 30 second sprint sections. During exercise the child's heart rate was continually monitored.

Results: 48 children were recruited (29 male). Patient demographics and study results are summarised in Table 1. 18/48 (38%) participants achieved >80% maximal predicted heart rate with 42/48 (88%) reaching >70%. Boys achieved significantly higher maximum power output (Median [IQR]=103Watts [92-113]) compared with girls (Median [IQR]=87Watts [75-103]), $p=0.023$ (Mann-Whitney U-test) but there was no difference in test duration ($p=0.99$). Increased height was associated with higher maximum power output: Spearman's correlation coefficient = 0.37, $p < 0.01$.

Conclusions: Exercise testing in young children is technically more challenging. This bespoke exercise bicycle/software is reliable, produces replicable results, is enjoyable and easy to use for children. It is a useful tool in the screening and assessment of young children with inherited cardiac disease and congenital heart disease.

P-151

Determination of Reference Values for Tricuspid Annular Plane Systolic Excursion in Healthy Turkish Children

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Objectives: A tricuspid annular plane systolic excursion (TAPSE) is an echocardiographic measurement used in the evaluation of right ventricular systolic function. While established reference values of TAPSE exist for the adult population, only a limited number of studies have attempted to evaluate reference values for children. The aim of the present study was to determine reference values of TAPSE in healthy children in Turkey.

Methods: A total of 765 healthy children aged between 0 and 18 years, all of whom referred to our clinic with cardiac murmurs, were evaluated. Patients with no cardiac problems or other disorders were excluded from the study. The measurement of TAPSE was obtained using an M-mode echocardiography, and the relationship between age and surface area with TAPSE was investigated.

Results: The mean TAPSE value was found to be 19.56 ± 5.54 cm, and no significant difference was identified between the male and female children subjects. TAPSE values showed an increase with increasing age and surface area. The mean TAPSE was 9.09 ± 1.36

in newborns and 25.91 ± 3.60 cm in the 13-18 age group. A negative correlation was found between TAPSE and heart rate.

Conclusions: In the present study, reference values for TAPSE in healthy Turkish children were presented in percentile tables, and corresponding z-scores were determined. These reference values may be useful in daily practice for the evaluation of right ventricular systolic function in children.

P-152

Evaluation of pediatric patients after surgical repair of tetralogy of Fallot using tissue Doppler and tissue synchronization imaging

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Background: Tetralogy of fallot (TOF) is the most common cyanotic heart congenital disease. Although surgical repair is satisfactory, the operated heart is not anatomically normal. These patients develop many cardiac functional and electrical abnormalities. We aimed to detect left ventricular dyssynchrony and to evaluate systolic and diastolic function after successful Fallot repair in the pediatric age group.

Methods: This cross sectional study included 25 post-operative pediatric patients with surgically repaired TOF. Fifteen sex and age matched healthy children were included as control. All patients were subjected to clinical assessment, chest X-ray, electrocardiography (ECG) and conventional echocardiography in addition to tissue Doppler echocardiography and automated tissue synchronization imaging using (GE vivid 7) machine.

Results: The mean age of the studied cases was 9.04 ± 3.41 y. In our study there were systolic and diastolic RV dysfunction in patients in comparison to controls as manifested by statistically significant lower S', E', A' tissue velocities, longer relaxation time and increased MPI of RV in comparison with controls. There was a positive statistical significant correlation between age of patient at surgery and E' of RV ($P=0.02$) There was a positive statistical significant correlation between postoperative duration and IRT of LV ($p=0.05$). Using the automated TSI method, 8 patients (32%) had mild to moderate delay in all segment standard deviation but there was no statistically significant difference between cases and controls regarding the main six indices evaluated by the TSI algorithm including (septolateral delay, Basal septal delay, basal septal maximum difference, all segment maximum delay, all segment standard deviation, septal posterior delay).

Conclusion: There is evidence of early diastolic dysfunction and restrictive pattern in both ventricles. Automated TSI provides a simple, rapid, and comprehensive assessment of intra-ventricular dyssynchrony. The automated processing algorithm reduces the impact of operator skill and improves reproducibility. LV dyssynchrony is not detected in the pediatric age group after Fallot repair

P-153

Doppler parameters and aortic compliance in patients with aortic coarctation

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Introduction: Patients with aortic coarctation (AoCo) often show a Doppler flow pattern with diastolic flow in the descending aorta.

The effect of aortic compliance on flow pattern in AoCo was described in-vitro and with computer models. This study was performed to evaluate the relation between invasive gradients, Doppler flow patterns and arterial stiffness measured by tonometry, in AoCo patients.

Methods: Forty five invasive gradients and TTE studies were obtained in 28 patients (13 women, 24.7 ± 17.6 years of age). In 17 patients studies were performed before and after intervention. Systolic invasive gradients were measured (Sgrad), and ratio of coarctation to aortic diameter at the diaphragm (AoCo/DAo) calculated. Doppler parameters measured at the time of invasive evaluation, included Doppler corrected gradient (Dgrad), diastolic velocity at end of T wave (DVT), end diastolic velocity (DVQ), systolic and diastolic half pressure times (SHPT and DHPT) and velocity runoff (VR - time for velocity to decrease from maximum value (Vmax) to 33% Vmax). VR, SHPT and DHPT were corrected with Bazett's formula. Arterial stiffness was assessed by measuring pulsed wave velocity (PWV) between carotid and right radial arteries.

Results: Overall, including patients before and after intervention, mean Sgrad was 24 mmHg, mean AoCo/DAo 0.56, mean Dgrad 32 mmHg, mean DVT 1.0 m/s, mean DVQ 0.3 m/s, mean SHPT 98 ms, mean DHPT 65 ms and mean VR 372 ms. With simple regression models, Sgrad and AoCo/DAo showed correlation with Dgrad, DVT, DVQ, SHPT and DHPT ($p < 0.01$). Multiple regression models provided the formulas $Sgrad = -3.63 + 0.05 \cdot DHPT + 0.75 \cdot Dgrad$ ($R^2 = 0.76$) and $AoCo/DAo = 0.83 - 0.001 \cdot SHPT - 0.005 \cdot Dgrad$ ($R^2 = 0.51$). Continuous Doppler flow in the descending aorta was found in 93% of patients with Sgrad > 30 mmHg and AoCo/DAo < 0.46 . Diastolic tail with measurable DVT was found in patients with Sgrad as low as 7 mmHg (59% of patients with Sgrad between 7 and 26 mmHg). In this group of low to moderate Sgrad, a negative correlation was found between PWV and DVT ($p = 0.02$) suggesting that low aortic stiffness may contribute to persistent diastolic flow.

Conclusions: Doppler systolic and diastolic parameters correlated well with severity of AoCo. In patients with moderate AoCo, Doppler diastolic flow was associated with a lower compliance of the aorta.

P-154

Assessment of cardiac functions and aortic stiffness indexes with tissue Doppler ecocardiography in Familial Mediterranean Fever patients

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Familial Mediterranean Fever (FMF) is an autosomal recessive autoinflammatory disease, characterized with recurrent fever, abdominal pain, chest pain and joint involvement. Cardiovascular involvement can be seen secondary to chronic inflammation. This severe outcome is a major cause of morbidity and mortality in FMF patients. There are a few studies investigating cardiovascular involvement in FMF patients. This study is conducted in order to assess possible cardiac involvement in FMF patients using electrocardiogram, conventional and tissue Doppler echocardiography.

The study population included 75 patients with FMF and 50 healthy children. There are no significant differences between the two groups in age, height, sex, body mass index, arterial blood pressure parameters ($p > 0.05$). QT and QTc dispersion parameter, which shows ventricular repolarization homogeneity in the two groups were similar ($p > 0.05$). Left ventricle end-diastolic (LVd)

and end-systolic diameters (LVs), left ventricle posterior wall thickness (LVPWd, LVPWs) and interventricular septum thickness (IVSd, IVSs) and left ventricle mass (LVM), left ventricle mass index (LVMi) and relative wall thickness (RWT), LV ejection fraction and fractional shortening time, RV ejection fraction and fractional shortening time parameters assessed with conventional echocardiography in FMF patients were similar to the parameters in control group ($p > 0.05$). While mitral valve maximum E wave velocity and E/A ratio and tricuspid valve maximum E wave velocity and E/A ratio were found to be decreased, tricuspid valve maximum A wave velocity was found to be increased by standart Doppler echocardiography in FMF patients ($p < 0.05$). Left ventricle S_{dm} , E_{dm} , E_{dm}/A_{dm} were decreased and A_{dm} was increased, right ventricle S_{dt} , E_{dt} , E_{dt}/A_{dt} were decreased and A_{dt} , DT_{dt} , IR_{dt} were increased, interventricular septum E_{divs} , E_{divs}/A_{divs} were decreased and A_{divs} was increased by the measurements made by tissue Doppler echocardiography in FMF patients ($p < 0.05$). While aortic systolic and diastolic diameters were similar in both groups ($p > 0.05$), aortic strain and distensibility were decreased and Ep , Ep^* , stiffness parameters were increased in FMF patients ($p < 0.05$). Our findings show cardiac involvement can exist in FMF patients even during non-attack period.

P-155

Friedreich's Ataxia in pediatric patients: global and segmental echocardiographic assessment

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Introduction: Friedreich's Ataxia (FA) is an autosomal recessive neurodegenerative disorder resulting from deficiency of frataxin. Cardiac impairment presents hypertrophic and dilated cardiomyopathies, ventricular dysfunction and arrhythmias. Little is known in cardiac presentation in children. We hypothesized that strain echocardiography could be a potential marker for early left ventricular dysfunction despite normal left ventricular ejection fraction even in early stages in children and young adults.

Methods: Retrospective and prospective, 14 patients with FA were analyzed (ECG and echo - global LV systolic function using Simpson's method and for regional wall motion abnormalities using advanced CMQ (QLAB Philips (r)) for the longitudinal and circumferential strain analysis).

Results: 75% men, mean age when first seen in cardiology was 7 years (5-14) years.

Echocardiography: Hypertrophic cardiomyopathy was detected in 12/14 (85%), one of them hypertrophic obstructive cardiomyopathy with mild-to-moderate mitral regurgitation. All cases of hypertrophic cardiomyopathy had threshold levels or a decreased diastolic dysfunction. All of cases had a normal ejection fraction using Simpson's method (58%-79%). One with mid hypertrophy, a regional dysfunction was detected when the longitudinal strain was performed. In all cases of hypertrophy cardiomyopathy, longitudinal strain analysis showed a reduction with segmental variation that was not consistent to a particular region.

Arrhythmias: two patients with hypertrophic cardiomyopathy had asymptomatic atrial extrasystole registered in the 24 h-cardiac Holter. No other arrhythmias had been detected.

ECC: we detected T wave changes in all cases with hypertrophic cardiomyopathy, deep S wave and high R wave in leads V2-V4, and incomplete right branch block in two cases. PR and QTc intervals were normal.

Treatment: All cases were under treatment with idebenone or idebenone plus vitE/CoQ10/beta blocker. Betablocker therapy was started when an obstructive hypertrophy or diastolic dysfunction were detected.

Conclusions: Children with FA and hypertrophic cardiomyopathy had an asymptomatic diastolic dysfunction and segmental dysfunction detected with longitudinal strain with a normal ejection fraction. Thus, segmental echo should be performed to all patients with FA.

P-156

Determination of the right ventricular outflow tract velocity time integral in pediatric atrial septal defect patients – useful or useless?

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Introduction: Determination of the right ventricular outflow tract velocity time integral (RVOT VTI) is an important part of the non-invasive investigation of pulmonary blood flow in adults, however, pediatric reference data are lacking. We examined growth related changes of RVOT VTI values in healthy children and the predictive value of RVOT VTI values in identifying enhanced pulmonary blood flow in children with secundum type atrial septal defect (ASD).

Methods: A prospective study was conducted in a group of 570 healthy children and 52 children with a moderate-sized to large ASD. The effects of age, body length (BL), body weight (BW), and body surface area (BSA) on RVOT VTI values were determined. The predictive value of normal values stratified for age, BW, BL, and BSA was tested in our 52 ASD children.

Results: RVOT VTI normal values showed a positive correlation with age, BL, BSA, and BW. In our population RVOT VTI z-scores demonstrated a high specificity for detecting ASD patients (>97%) with sensitivity up to 71%.

Conclusions: We calculated z-scores of pediatric RVOT VTI values. We found elevated RVOT VTI values of our pediatric patients with moderate-sized to large ASDs when compared to age-related normal values. Therefore determination of the RVOT VTI might be an additional predictor in identifying significantly increased pulmonary blood flow in ASD patients.

P-157

Determination of right ventricular size parameters using 2D echocardiography in children with an atrial septal defect: ready for clinical use?

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Introduction: Determination of right ventricular (RV) size and function has gained more interest during recent years in both

adults and children, especially in patients with congenital heart disease. We investigated growth related changes of RV internal dimensions in a healthy pediatric cohort and the predictive value of RV parameters in identifying enlarged RVs in children with a secundum type atrial septal defect (ASD) to investigate for a possible use in clinical use.

Methods: A prospective study was conducted in a group of 576 healthy children and 37 children (age range: 1.4 – 17.7 years) with a moderate-sized to large ASD. We determined the effects of age, body length (BL), body weight (BW), and body surface area (BSA) on RV parameters: end-diastolic basal-diameter (EDb-d), end-diastolic mid-cavity diameter (EDm-d), end-diastolic length (EDL), end-systolic length (ESL), end-diastolic area (EDa), and end-systolic area (ESa). The predictive value of normal values stratified for age, BW, BL, and BSA was tested in our ASD children.

Results: RVEDb-d, RVEDm-d, RVEDL, RVESL, RVEDa, and RVESa showed a positive correlation with age, BL, BSA, and BW. RV z-scores showed a high specificity for detecting ASD patients with sensitivity up to 89%, especially in ASD children below eight years of age.

Conclusions: We could identify enlarged RV diameters of our ASD patients compared to age-related normal RV parameter z-scores, especially in children below eight years of age. This may be useful to guide decision making in ASD patients for timing interventional or surgical closure.

P-158

Pulmonary arterial wall thickness in Eisenmenger Syndrome: Prospective, cross-sectional, controlled clinical trial

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Background and Aim: The aim of current study is to investigate echocardiographic pulmonary artery wall thickness (PAWT's) association with angiocardiology, echocardiography, and biochemical findings and to demonstrate its predictive role in morbidity of disease.

Materials and Method: Nineteen patients with Eisenmenger Syndrome (ES) (13 females; a mean age of 12.0 ± 4.1 (min-max 4-17) years) and 24 (16 females; a mean age of 12.1 ± 4.3 (min-max 3-18) years) healthy subjects as a control group were included in this prospective, cross-sectional, controlled clinical study between December, 2012 and December 2013. Both groups underwent echocardiography including pulmonary artery wall thickness (PAWT), pulmonary artery diameter, right ventricle wall thickness (RVWT), TAPSE, right atrial diameter, vena cava inferior diameter in addition 2D echocardiography and plasma N Terminal pro brain natriuretic peptide (NTProBNP) measurement, six-minute walk distance (6MWD) test. Patient group performed right heart catheterization and functional class. PAWT were measured at the end of systole at distal of pulmonary valves at the parasternal short-axis.

Results: PAWT ($p=0.005$) was higher in the patient group together with pulmonary arterial diameter (Z-score, $p<0.001$), vena cava inferior diameter ($p=0.002$), and RVWT, while TAPSE was significantly lower ($p=0.002$). PAWT was strongly positively correlated to RVWT ($r=0.893$, $p<0.001$) and moderate negatively correlated to TAPSE ($r=0.597$; $p<0.011$).

Conclusion: PAWT can be used as an additional parameter with other echocardiographic parameters in the follow-up of Eisenmenger Syndrome in children.

Table.

Variables	PAWT	
	r	p
Age	0.147	0.328
BMI	0.126	0.630
SBP	0.302	0.904
SaO ₂	-0.301	0.241
6-MWT	0.161	0.536
Hemoglobin	0.363	0.153
Uric acid	0.090	0.732
NT-ProBNP	0.400	0.112
PA diameter	0.461	0.063
RA axial diam.	0.242	0.366
VCI diameter	0.114	0.664
TAPSE	-0.597	0.011
RVWT	0.893	< 0.001
Pp/Ps	0.197	0.448
PVR	0.124	0.636
PVR/SVR	0.105	0.690

BMI: body mass index, SBP: systemic blood pressure, SaO₂: arterial oxygen saturation, 6-MWT: six-minute walk test, BUN: blood urea nitrogen, NT-ProBNP: N-terminal of the prohormone brain natriuretic peptide, PA: pulmonary artery, RA: right atrium, VCI: vena cava inferior, TAPSE: tricuspid annular plane systolic excursion, RVWT: right ventricular wall thickness, Pp/Ps: Ratio of invasive Pulmonary peak pressure and Systemic pressure, PVR: pulmonary vascular resistance, SVR: systemic vascular resistance.

P-159

The importance of right ventricular myocardial performance index in children with Eisenmenger syndrome

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Background and Aim: The purpose of this study is to evaluate potential use of echocardiography with functional echocardiographic right ventricular variables besides right ventricle myocardial performance index (RVMPI) for assessing disease severity of Eisenmenger Syndrome (ES) and to determine new follow-up parameters for the disease.

Material-Methods: This study consisted of 24 normal controls (the control group) and 19 patients with ES. The echocardiographic practice consisting RVMPI, tricuspid annular plane systolic excursion (TAPSE), diameter of right atrium, vena cava inferior and pulmonary artery; right ventricle catheterization and six-minute walk distance (6MWD) test were performed to the patients with ES. NT-ProBNP was measured in both control and study groups. Study group was divided into two functional PHT stage according to WHO classification.

Results: In the study group TAPSE was found lower than the control group, besides the other echocardiographic variables were high. There was positive correlation between RVMPI and NT-ProBNP ($p = 0.001$; $r: 0.716$), between RVMPI and functional stage ($p = 0.001$; $r: 0.725$) and between RVMPI and pulmonary vascular resistance ($p = 0.045$; $r: 0.491$) in the study group. The patients with ES were divided into two stages as class 2 ($n: 9$) and class 3 ($n: 10$) according to WHO. RVMPI was high in stage 3 patients ($p = 0.001$). There was not statistically significant correlation between the functional stage and other echocardiographic parameters.

Conclusion: RVMPI which is one of the quantitative echocardiographic parameters can be used to evaluate the disease severity of ES in follow-up of outpatients.

Table.

Variables	RVMPI	
	r	p
Gender	0.079	0.755
BMI	-0.553	0.017
SBP	-0.120	0.636
SaO ₂	0.362	0.140
6-MWD	-0.400	0.100
Hemoglobin	-0.448	0.062
Uric acid	-0.389	0.110
NT-ProBNP	0.716	0.001
PA diameter	0.048	0.855
RA axial diam.	-0.419	0.094
VCI diameter	-0.164	0.524
TAPSE	0.050	0.845
RVWT	-0.165	0.487
Pp/Ps	-0.209	0.405
PVR	0.491	0.045
PVR/SVR	0.249	0.336

RVMPI: Right ventricle myocardial performance index, BMI: body mass index, SBP: systemic blood pressure, SaO₂: arterial oxygen saturation, 6-MWD: six-minute walk diameter, BUN: blood urea nitrogen, NT-ProBNP: N-terminal of the prohormone brain natriuretic peptide, PA: pulmonary artery, RA: right atrium, VCI: vena cava inferior, TAPSE: tricuspid annular plane systolic excursion, RVWT: right ventricular wall thickness, Pp/Ps: Ratio of invasive Pulmonary peak pressure and Systemic pressure, PVR: pulmonary vascular resistance, SVR: systemic vascular resistance.

P-160

A quantitative analysis of ventricular myocardial mass and volume in functional single ventricle using cardiac magnetic resonance imaging

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Introduction: Recently, the Fontan procedure has become feasible in most patients with functional single ventricle. However, post-operative gradual deterioration of ventricular function can occur. Its etiology still remains unclear.

Objectives: To analyze the ventricular volume and mass in patients with functional single ventricle quantitatively by cardiac magnetic resonance imaging (MRI), especially from the point of view of ventricular morphology.

Methods: We analyzed cardiac MRI data from 19 patients with functional single ventricle (RV morphology, 12 patients, LV morphology 7 patients).

Ventricular volume and mass were measured by manual tracing of endocardial and epicardial borders on each short-axis cine slice at end-diastole and end-systole. The Simpson method was applied to calculate end-diastolic volume, end-systolic volume, ventricular mass, and volume/ventricular ratio.

Results: Both indexed end-systolic volume (EDVI) and indexed end-systolic volume (ESVI) were significantly higher in the RV group when compared with the LV group (EDVI: 140.2 ± 62.1 mL/m² vs 89.3 ± 20.1 mL/m², $p < 0.01$, and ESVI: 91.9 ± 56.1 mL/m² vs 55.3 ± 16.5 mL/m², $p < 0.05$, respectively). However, indexed ventricular mass was similar between the two groups (54.5 ± 16.6 g/m² vs 56.3 ± 12.5 g/m², respectively, $P = 0.34$). Consequently, ventricular mass/volume ratio was significantly lower in the RV group when compared with LV group (0.410 ± 0.072 g/mL vs 0.642 ± 0.125 g/mL, $P < 0.0005$).

Conclusions: In patients with functional single ventricle, RV morphology has the potential risk for decompensation to pressure and/or volume overload in terms of ventricular mass/volume ratio.

P-161

Myocardial function long term after acute illness of Kawasaki disease: conventional echocardiographic analysis

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Objective: To study left ventricular myocardial function long term after Kawasaki disease (KD) using conventional echocardiography.
Patients and Methods: Observational case-control study on ninety-two KD subjects; 51% male, age 11.9 years (4.3–32.2), 8.3 years (1.0–30.7) from KD-diagnosis. Group I: with coronary abnormality (CAA +, N = 38) at 2 months after onset of KD or during follow-up. Group II: without (CAA-, N = 54). Group III: sex/age-matched controls (N = 51). Echocardiography studies were performed on GE Vivid 7. Myocardial function was assessed through conventional echocardiography. Fractional shortening (FS), 2D ejection fraction by Simpson's method (EF) were used for assessment of systolic function; whilst mitral inflow E/A ratio, pulsed wave tissue Doppler imaging (TDI) of mitral lateral annular velocities (M lat E') as well as E/E' (M lat E/E') were used to assess diastolic function.

Results: 136/143 studies were available for assessment with the results presented in the table below.

There was no statistically significant difference between the above groups (t-test $p > 0.05$), as well as the subgroup with current coronary abnormality (CAA + current) versus all the other groups. Mild systolic dysfunction did occur in one patient who required an earlier revascularization procedure.

Conclusion: Conventional systolic and diastolic function parameters long after KD remain without difference to normal age-matched controls, in patients with no significant coronary artery obstruction.

Table.

Parameter	Average			SD			Min			Max		
	CAA-	CAA+	Cont	CAA-	CAA+	Cont	CAA-	CAA+	Cont	CAA-	CAA+	Cont
FS(%)	37.06	37.05	36.52	4.61	4.68	4.98	30	24	28	48	45	54
EF(%)	62.20	62.31	61.33	3.70	5.52	5.94	55	42	50	70	70	75
E/A	1.93	1.99	2.04	0.46	0.50	0.56	1.05	0.95	1.00	3.20	3.07	3.45
M lat E'	18.8	17.97	18.98	3.31	3.68	3.83	11	11	11	27	29	26
M lat E/E'	5.64	5.85	5.87	1.15	1.45	1.69	2.93	3.50	1.86	8.08	9.83	9.73

P-162

Myocardial function long term after acute illness of Kawasaki disease: myocardial deformation imaging

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Objective: To study global myocardial function long term after Kawasaki disease (KD).

Patients and Methods: Observational case-control study on ninety-two KD subjects; 51% male, age 11.9 years (4.3–32.2), 8.3 years (1.0–30.7) from KD-diagnosis. Group I: with coronary abnormality (CAA +, N = 38) at first 2 months after onset of the disease or during follow-up. Group II: without (CAA-, N = 54). Group III: sex/age-matched controls (N = 51). Myocardial function assessed by conventional echocardiographic techniques and two-dimensional (2D) speckle tracking myocardial deformation.

Table.

Group	Mean strain %	SD + /-	Min %	Max%
Longitudinal				
CAA-	20.96	4.94	14.67	29.50
CAA +	21.76	4.49	16.00	29.00
Controls	21.45	5.86	16.17	29.50
Circumferential				
CAA-	20.36	6.74	13.67	28.83
CAA +	21.99	7.87	13.20	30.67
Controls	20.91	6.48	10.83	28.67

Results: A total of 132/143 patients/studies (50/54 CAA-, 34/38 CAA +, 48/51 Controls) were available for 2D Longitudinal and Circumferential Strain analysis.

There was no statistically significant difference between groups (t-test > 0.05 for comparison between the KD groups as well as between them and the controls). Further on the group of those who have current coronary abnormality 17/38 from the CAA + group had no statistically significant difference.

Conclusion: Global mean myocardial longitudinal and circumferential systolic strain in patients long after Kawasaki disease does not differentiate them from normal controls.

P-163

Impairment of left ventricular myocardial function in children with chronic renal failure in early stage by 2D-Speckle Tracking Echocardiography

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Introduction: The patients with chronic renal failure more frequently develop left ventricular (LV) structural and functional abnormalities. The aim of this study is to evaluate the LV regional function using 2D-speckle tracking echocardiography (2D-STE) in children with chronic renal failure.

Methods: In total 28 chronic renal failure patients between 6–18 ages and 20 healthy children were included. After standard echocardiographic measurement consisting left ventricle ejection fraction (EF), shortening fraction (SF), mitral E, mitral A as well as tricuspid annular plane systolic excursion (TAPSE) values were measured and also mitral E/A ratio was calculated. Using the 2D-STE method, the strain (S) and strain rate (SRs:systolic, SRe: early diastolic, SRa: late diastolic) values belonging to the longitudinal (L), circumferential (C) and radial (R) functions of the LV have been measured. SRe/a values were also calculated.

Results: Systolic and diastolic functions of LV in both groups were similar. TAPSE values were significantly decreased in children with chronic renal failure comparing to the control group, respectively (21.4 mm and 24.5 mm, $p < 0.05$). Longitudinal, circumferential and radial SRs values of the chronic renal failure patients were significantly decreased according to the control group (-1.3 ± 0.4 and -1.5 ± 0.3 , $p < 0.05$; -1.6 ± 0.4 and 1.8 ± 0.4 , $p < 0.05$; 2.1 ± 0.9 and 3.1 ± 1.3 , $p = 0.001$ respectively). Besides, on the basis of longitudinal function, the SRe and SRe/a values decreased significantly in children with chronic renal failure (2.1 ± 0.8 and 2.6 ± 0.7 , $p < 0.05$; 1.6 ± 0.7 and 2.4 ± 1.2 , $p < 0.05$ respectively). Although, global L,C and R strain (%) values were lower in patient group, no statistical significance was detected.

Conclusions: Although, systolic and diastolic functions of LV during standard examination were normal in children with chronic renal failure, regional LV functions using 2D-STE were impaired. 2D-STE

may contribute to early identification of impaired left ventricular myocardial function in children with chronic renal failure.

P-164

Correlation between cardio-pulmonary exercise test variables and health-related quality of life among children with congenital heart diseases

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Introduction: maximal oxygen uptake (VO₂max) correlates with health related quality of life (QoL) in adults with heart failure. Cardio-pulmonary exercise test (CPET) with VO₂max and VE/VCO₂ slope evaluation is recommended in the follow-up of adults with congenital heart diseases (CHD). Few data is available as regards correlation between CPET and QoL among children with CHD.

Method: 201 children with a CHD aged 8 to 18 performed in 2 tertiary care pediatric cardiology centers a maximal CPET including a spirometry. Children were separated into 2 groups depending on the ergometer (treadmill n = 96, cycle-ergometer n = 105). Common gas exchange measurement device was used. CHD severity was stratified into 4 groups (from Uzark et al.). On the same day, all children and their parents filled out separately the Kidscreen, a validated pediatric generic QoL questionnaire. Informed consent was obtained from all parents.

Results: VO₂max, anaerobic threshold (AT), flow-volume loop and ventilatory response during exercise with respiratory equivalent for CO₂ (VE/VCO₂ slope) and respiratory efficiency (VD/VTmax) were correlated to CHD severity (p < 0.05).

Physical well-being scores were correlated to VO₂max for parents reported QoL (r = 0.43, p < 0.0001) and children self reported QoL (r = 0.27, p < 0.0001). Parents reported QoL scores were correlated to AT (r = 0.33, p < 0.0001), VD/VTmax (r = -0.30, p < 0.0001) for physical well-being and to VO₂max (r = 0.21, p = 0.004) for psychological well-being. Self reported QoL scores were correlated to oxygen uptake efficiency slope (OUES: r = -0.26, p = 0.017), VE/VCO₂ slope (r = 0.22, p = 0.037) for school environment. Strongest correlations were observed in the treadmill group, especially between VO₂max and physical well being for parents (r = 0.57, p < 0.0001) and self (r = 0.36, p = 0.0005) reported QoL scores.

Conclusion: VO₂max and ventilatory response to exercise correlate with self and parents-reported QoL among CHD children. If QoL and CPET are clinical trials outcomes in CHD children, we suggest to use parents related QoL scores and treadmill.

P-165

Tissue Doppler Imaging - an important adjunct in Predicting the Outcome of Arterial Switch Surgery in Transposition of Great Arteries

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Background: Predicting the feasibility of ASO in patients with TGA is a contentious issue, especially in infants presenting beyond

3 weeks of age. We studied the usefulness of various echocardiographic parameters including tissue velocities, strain and strain rate imaging in predicting the outcome following arterial switch operation (ASO) for transposition of great arteries (TGA).

Methods: A total of 42 infants (aged 3 - 123 days) with dTGA and intact ventricular septum or a small ventricular septal defect (VSD) were included in the study. Standard echocardiography including Doppler studies was performed with 5 or 10 MHz probe (GE Vivid 7 Machine) prior to surgery. Intra and inter-observer variability of each parameter was assessed to be within acceptable limits. These echocardiographic variables were correlated with surgical outcomes. A composite end point of death, use of ECMO, and prolonged ICU stay (>10 days) was considered to indicate a complicated postoperative course.

Results: Out of the 42 patients with IVS, 12 patients had a regressed LV by conventional assessment. Left ventricular dimension in systole and posterior wall thickness were the only parameters that were significantly different in patients with a preserved left ventricle. In this cohort, 10 patients had a complicated postoperative course. On logistic regression analysis, LV mass index (OR 6.0; p value 0.04) and anteroseptal basal strain (OR 9.2; P value 0.03) were significant independent predictors of poor outcome. Age alone was not an independent predictor of poor outcome (OR 0.37; p value = 0.44). A cut off of LV mass index of 35 gm/m² and -18.5% peak systolic strain were good predictors of poor outcome. The area under the curve of ROC for peak systolic strain at -18.5% for differentiating good and poor outcome was 0.89.

Conclusions: Functional assessment of LV with tissue strain in addition to structural assessment with LV mass index are useful in predicting a complicated outcome in patients of TGA undergoing arterial switch operation (ASO). The utility of these measures have to be confirmed in larger studies.

P-166

Profile of Lung Ultrasound in Children With Congenital and Acquired Heart Disease - A Pilot Study

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Background: Lung ultrasound has been increasingly recognized as a necessary tool in the past few decades. However, lung ultrasound is yet to be applied in pediatric cardiology practice. This is the first study of lung ultrasound in children with heart failure in children.

Methods: We included 30 children (<12 years of age) after an informed parental consent. Patient's details including the cardiac diagnosis and presence or absence of heart failure were recorded. Lung ultrasound examination was performed bedside using a probe with a convex or linear tip placed at the level of an intercostal space, using an emission frequency ranging between 4 and 15 MHz. The lung ultrasound profile and B line scores were noted.

Results: The median age of the patients was 3.5 months (Range 4 days to 8.5 years) and 18 were male. Cardiac diagnosis included cyanotic conditions 18 (60%), and acyanotic conditions including shunt lesions 12 (40%). Overall 17 patients (56.7%) could be classified as having clinical heart failure.

Eleven cases (36.7%) were diagnosed as having a pneumonia based on either a C profile or an A/B profile with shred sign positive on lung ultrasound. Two patients had a collapse and in one of them the collapse was not evident on chest radiograph. The detection of consolidation by lung ultrasound was more frequent than by chest radiographs.

The distribution of B lines as assessed by the B line score was found to be significantly correlated with the presence of heart failure.

When the semi-quantitative B-line score was analyzed, it showed a good correlation with the Ross Heart Failure class (Gamma = 0.94 and $r=0.87$), but a poor correlation with the radiographic pulmonary venous hypertension (Gamma 0.67 and $r=0.49$).

Conclusion: Lung ultrasound can be safely and easily performed in pediatric cardiology patients. The presence of B lines was strongly associated with the presence and degree of heart failure. Lung ultrasound had better sensitivity in detecting lung collapse and pneumonia.

P-167

Prevalence of Bicuspid Aortic Valve in Turner Syndrome and its Correlation to Karyotype: a Multicentre Prospective Magnetic Resonance Study

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Introduction: Congenital heart disease affects approximately 50% of individuals with Turner syndrome (TS), and significantly contributes to an overall increased mortality in TS. Bicuspid aortic valve (BAV) represents the most common congenital heart defect in TS patients. It proved to be one of the most important risk factor for aortic dilatation and dissection.

Objectives: To determine the correlation between the particular karyotypes and the prevalence of BAV in TS individual.

Methods: 67 TS patients aged 6.6 to 32.5 years (median 14.0 years) followed in three tertiary centres underwent cardiac magnetic resonance imaging study. They were divided into four cytogenetic subgroups: 45,X karyotype; 45,X/46,XX mosaicism; structural abnormalities of the X chromosome; and 45,X/structural abnormality of the X chromosome mosaicism. Prevalence of BAV and odds ratio (OR) compared to the general population in the whole study group; and statistical comparison of the prevalences of BAV among the individual subgroups were determined.

Results: BAV was found in 28.4% of all patients (OR 208.3; p -value 0.0001). Statistically significant difference between the prevalences of BAV in patients with any 45,X cell line in their karyotype compared to the patients with only structural abnormality of the X chromosome was proved (p -value 0.05).

Conclusion: our data confirms the hypothesis that prevalence of bicuspid aortic valve differs between karyotype subgroups of TS. 45,X karyotype is associated with the highest prevalence of BAV. Also, the presence of the 45,X cell line in the mosaic karyotype increases the probability of BAV.

P-168

Elevation of gamma-glutamyl transpeptidase reflects hepatic impairment caused by a variety of mild stresses in Fontan patients

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Background: Hepatic impairment is said to occur after Fontan because of elevated pressure in inferior vena cava (IVCp).

However, we often encounter hepatic impairment without IVCp-elevation in Fontan patients. We sometimes find levels of gamma-glutamyl transpeptidase (GGT) rising after Fontan. We investigated whether we could identify the cause of hepatic impairment by elevation of GGT (GGT-elevation).

Methods: The medical records of 116 patients after Fontan procedure were reviewed. Cardiac catheterizations and blood tests were performed in stabilized period after Fontan between 2004 and 2014. First, we compared GGT levels between before and after Fontan. Second, we defined GGT-elevation as GGT levels in the top fifth of 116 post-Fontan patients ($\text{GGT} \geq 119 \text{ U/L}$). Cardiac performances were determined which affected GGT-elevation.

Results: Levels of GGT were higher after Fontan than those before Fontan (16 vs. 78 U/L, $p < 0.0001$). Both levels of aspartate and alanine aminotransferase were significantly increased after Fontan, the rises of which were slight respectively. After multivariate analysis GGT-elevation was independently associated with odds ratio of 9.0 ($p = 0.001$) for IVCp ($\geq 13 \text{ mmHg}$), 5.3 ($p = 0.006$) for index of pulmonary artery ($< 170 \text{ mm}^2/\text{m}^2$), 4.5 ($p = 0.010$) for ejection fraction of major ventricle ($\leq 50\%$), and 4.1 ($p = 0.016$) for levels of atrial natriuretic peptide as index of elevated intravascular volume ($\geq 54 \text{ pg/ml}$). In monovariate analysis GGT-elevation was significantly related to wedge pressure of pulmonary artery ($\geq 7 \text{ mmHg}$). Levels of brain natriuretic peptide had no association with GGT-elevation. The rate of patients with GGT-elevation increased with the number of risk factors growing from 0 to 5: 5% of patients had GGT-elevation at no risk factor; 14% at 1; 19% at 2; 54% at 3; 80% at 4; 100% at 5.

Conclusion: Our study showed levels of GGT were highly elevated after Fontan procedure, by means of which we could infer the risk factors of liver impairment in Fontan patients. Narrow pulmonary artery, low ejection power, and venostasis, all of which were mild change, were independently related to hepatic impairment other than IVCp-elevation. We should attend GGT-elevation and these risk factors for hepatic involvement, even if Fontan patients did not have IVCp-elevation.

P-169

Correlation Between Right Ventricular Outflow Sizing and Right Ventricular Function and Volumes in Patients With Repaired Tetralogy of Fallot Undergoing Routine MR Follow-up: is There a Better Candidate for Percutaneous Pulmonary Valve Implantation?

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Introduction: cMR is the established tool for routine RV assessment in repaired tetralogy of Fallot (rTOF) patients. Anatomical data of the RVOT are assessed to improve indications for Pulmonary Valve Replacement (PVR), either surgical or percutaneous. Current eligibility criterion for percutaneous treatment (PPVI) is a maximum PT diameter up to 27 mm. The aim of this cross-sectional analysis is to improve the ability of CMR to identify patients suitable for percutaneous pulmonary valve implantation (PPVI).

Methods: Patients with rTOF and significant pulmonary regurgitation (PR) underwent routine CMR. In a sub-group of 31 patients a targeted 3D SSFP navigator sequence set at end-systole was also performed to better assess the pulmonary trunk (PT) morphology, length and dimensions. Transverse Diameter (TD) and Superior-Inferior Diameter (SID) and area obtained from the 3D dataset according to vessel analysis principles at three levels (PV remnant, mid-portion, bifurcation) were measured. We assumed that after

PPVI the geometry of the PT will shift from elliptic to circular, thus we calculated a predicted PT ($pPT = \sqrt{4 \times \text{elliptic area} / \pi}$) circular diameter through geometrical correction of the measured elliptic area at the PV remnant level.

Results: A statistical significant positive correlation was observed between EDV, the area and TD of pulmonary remnant ($p < 0,01$), although no correlation was observed between EDV and DSI, or between SID, TD, the area and RV ESV and RVEF.

The pPT diameter showed a stronger correlation to EDV (both absolute and indexed) as compared to individual observed PT diameters ($R^2 = 0.74$). When comparing the current anatomical criteria for PPVI eligibility to the pPT diameter the number of eligible patient increased from 3 to 11 ($p < 0.05$). In addition, in ROC analysis an EDVi between 134 and 139 mL/m² best identified patients eligible for PPVI according to the pPT diameter ($AUC = 0,44$).

Conclusion: Our study suggests that geometrically predicted PT diameters is more strongly associated with RV EDV as compared to measures currently used in clinical practice. Use of predicted PT diameter improves the identification of PPVI eligible patient. Our study suggests that higher rates of PPVI eligible patients are present when EDVi is between 134 and 139 mL/m².

P-170

Right ventricle behavior over time in repaired Tetralogy of Fallot patients

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Background: The progression of right ventricle (RV) dilation, mainly due to pulmonary regurgitation (PR) after Tetralogy of Fallot repair (rToF) with transannular (TP) or infundibular (IP) patch, varies among patients. The onset of RV dysfunction, due to its progressive dilation, is still difficult to predict. Pulmonary valve replacement (PVR) does not seem to affect RV function, if already impaired. Understanding the rate of progression of RV dilation over time could help identify the right timing for PVR in order to prevent RV dysfunction.

Methods: 118 rToF patients (mean age $19,7 \pm 10,8$ years, 59% men) with TP or IP underwent repeated cardiac magnetic resonance (CMR) exams at two centers in Italy between March 2008 and March 2014. Data regarding date of surgery and type of correction were collected, in addition to CMR parameters (RV/LV dimension and function, pulmonary trunk/arteries stenosis, tricuspid regurgitation and restrictive pattern flow in the pulmonary artery).

Results: In the whole population RV volume barely increased over time, paired with a slight RV ejection fraction reduction. TP patients (86% of whole population) showed a significantly higher rate ($p = 0,002$) of RV dilation (36%) compared to IP ones (19%) and a lower rate of RV reduction (3% vs 25%). RV dilation was not significantly correlated with the type of first surgery (palliative shunt vs repair) or the time elapsed from surgery. Eventually pulmonary trunk/arteries stenosis or restrictive pattern did not significantly influence RV changes over time.

Conclusion: RV dilation did not progress consistently in the majority of our rToF patients, being nearly stable over time. Significant increase in RV dimensions and concurrent impairment in function was observed in a subgroup of patients mainly composed of TP correction, regardless of the presence of a palliative shunt. The mild reduction in RV volume over time occurred in a small subgroup, without any significant correlation with RV restrictive physiology pattern.

P-171

Right ventricle linear measurements z score in children.

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Introduction: Reference linear measures of normal right ventricle (RV) in children is important, especially to detect abnormal RV dimensions in patients with congenital heart disease (CHD). Data relating RV diameters is scant in pediatric population. Z score of RV internal diameters (basal, midcavity and longitudinal) versus body surface area (BSA) is provided.

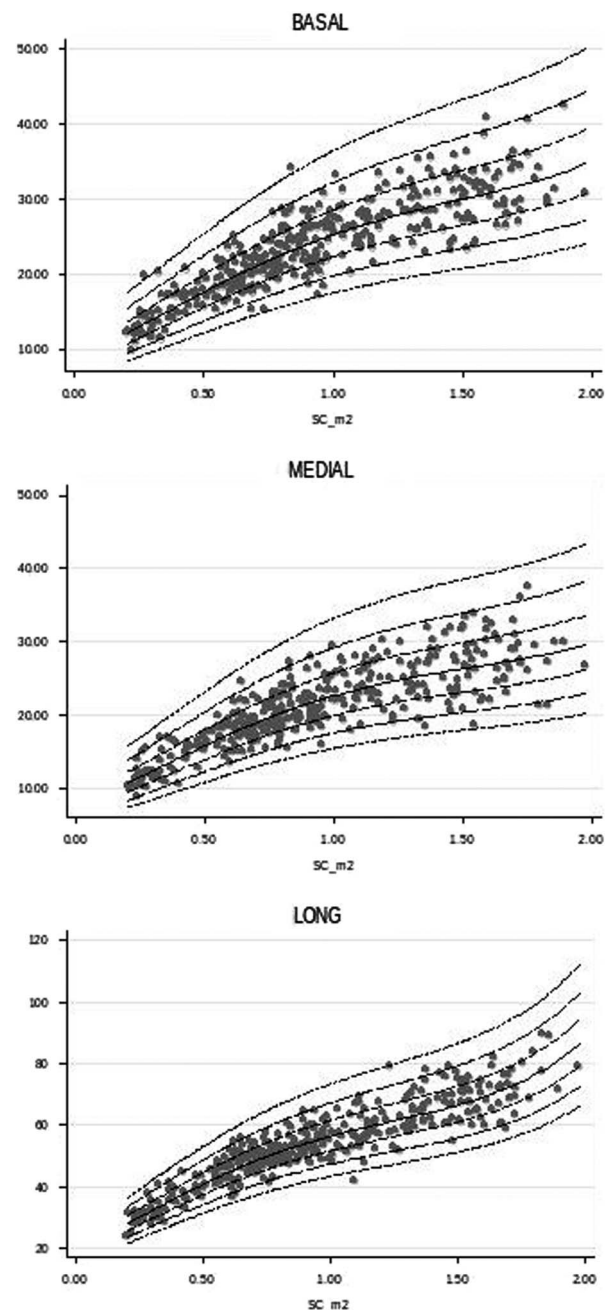


Figure.

Methods: A healthy study cohort of 400 patients (0 days to 18 years old) was enrolled. RV end-diastolic internal diameters (basal, midcavity and longitudinal) was measured in a 4-chamber focused view as 2D RV guidelines recommend. Using body surface area (BSA) as a significant determinant of RV size versus the different diameters allowed the development of normal z-score tables for each of the diameters.

Results: In published recommendations for quantification methods in pediatric echocardiography, RV internal dimension parameters are described to be useful, but normative data is scant. All investigated RV diameters increased from neonates to adolescents in a nonlinear way. There was strong collinearity of BSA with basal diameter (basRV), midcavity diameter (midRV) and longitudinal diameter (longRV), with correspondingly R^2 values of 0.79, 0.77 and 0.87. BSA related -2, 0 and +2 z-score for RV diameters were:

- for 0.5 m² BSA, in basRV: 13.8 mm, 17.6 mm and 22.5 mm respectively, in midRV: 12.3 mm, 15.9 mm, 20.5 mm and in longRV: 34.3 mm, 40.8 mm, 48.6 mm.
- for 1 m² BSA, in basRV: 19.8 mm, 25.3 mm and 32.3 mm respectively, in midRV: 17.6 mm, 22.7 mm, 29.2 mm and in longRV: 47.4 mm, 56.4 mm, 67.2 mm.
- for 1.5 m² BSA, in basRV: 23.5 mm, 30 mm and 38.3 mm respectively, in midRV: 20.4 mm, 26.3 mm, 33.9 mm and in longRV: 55.9 mm, 66.6 mm, 79.3 mm,

as shown in the graphics.

Conclusions: the normal ranges of pediatric RV internal diameters are provided and the z-score is also calculated. This diameters are easy to determine and may be used as noninvasive measurements to study RV size. Normal RV internal diameters z-scores might be important predictors in identifying enlarged RV in patients with congenital heart disease. Determination of RV internal parameters in children with CHDs and its comparison with normal z-score could provide a new insight in follow up and decision making in CHD.

P-172

Impact of exercise and cardiac rehabilitation on echocardiographic functional parameters in postoperated Tetralogy of Fallot children

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Introduction: exercise capacity of children with congenital heart disease (CHD) is often depressed. We developed a personalized cardiac rehabilitation program (CRP) conceived to counteract the deconditioning and comorbidities associated with prolonged sedentarism and physical inactivity after cardiac surgery in children.

The objective of this study was to characterize the effect of a CRP on echocardiographic functional parameters.

Methods: 18 postoperated Tetralogy of Fallot (POTOF) patients were recruited. Our CRP commenced with a comprehensive intake evaluation of the patient including a clinical status review, a baseline echocardiography, exercise test and a post exercise echocardiography. Documentation of nutrition, life-style and cardiac risk factors was also obtained. A supervised exercise training program with both aerobic and respiratory training components is then designed individually. After 3 months of CRP, full assessment is repeated. Relevant cardiovascular outcomes of cardiac rehabilitation are classified as:

- echocardiographic systo/diastolic functional parameters pre and post CRP.

Left ventricle (LV): EF, SF, E/E' and Tei index. Right ventricle (RV): TAPSE, FAC, E/E' and Tei index.

- quality of life outcomes: individual's perception of satisfaction with life, social functioning and general sense of well-being measured by questionnaire.
- ergometric parameters pre/post CRP HR, BP, endurance time and double product.

Results: echocardiographic baseline parameters pre and post CRP were compared. Systolic function improved. SF: mean preCRP 32%, postCRP 35.1% and mean rise +7.76%; EF: mean preCRP 63.84%, postCRP 68.8%, mean increase +5.11%; TAPSE: mean preCRP 13.2 mm, postCRP 15.95 mm, mean rise +10.2 mm; FAC: preCRP 44.5%, postCRP 59.7%, mean increase +13.14%. Mitral diastolic function improved partially while tricuspid E/E' remained the same.

Some of the results were invalid whilst others suffered no significant changes. 83.3% of patients experienced improvement at least in one of the echocardiographic parameters studied. Only 3 children suffered worsening in diastolic function (E/E' increase).

Conclusions: An improvement in cardiac function (quantified by echocardiography) has been demonstrated in POTOF after CRP (that includes education and counseling in addition to exercise and respiratory training). An improvement in ergometric parameters and quality of life was also assessed. Our CRP was performed in the absence of symptoms or cardiac events.

P-173

Assessment of pulmonary regurgitation by cardiac magnetic resonance: is the reverse volume more accurate than the regurgitation fraction?

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Introduction: Pulmonary regurgitation (PR) is a common finding after right ventricular (RV) outflow tract surgery and may result in RV dilation and dysfunction. Cardiac magnetic resonance (CMR) imaging is the imaging modality of choice for the quantification of PR and assessment of RV size and function. The expression of the regurgitant burden as a fraction of forward pulmonary flow is commonplace. However, PR fraction (PRF) may be highly variable in terms of absolute PR volumes (PRV). Therefore, we hypothesized that a volumetric expression of PR would be a better measure of ventricular preload and a more accurate reflection of degree of insufficiency.

Methods: We retrospectively studied 44 patients (mean age 23 ± 11 years, 27 males) with pulmonary valve or right ventricle-to-pulmonary conduit regurgitation due to various congenital heart disease: 21 patients with tetralogy of Fallot, 8 after Ross operation, 6 with pulmonary atresia or critical pulmonary stenosis, 3 with transposition of the great arteries, 3 with double outlet right ventricle, and 1 with truncus arteriosus. CMR were performed before and after surgical valve replacement (14 patients) or percutaneous Melody valve implantation (30 patients). Short axis ECG-triggered cine and phase contrast sequences were acquired to calculate RV volumes and pulmonary flow. The volumes were adjusted for BSA. We calculated both PRF (retrograde to antegrade flow ratio) and PRV (ml/m²) and correlated the results with RV end diastolic volume index (EDVi) before and after procedures, and RV stroke volume index (SVi). A Spearman test was used and p-value ≤0.05 was considered significant.

Results: Overall PRF (%), PRV (ml/m²), RVEDVi (ml/m²) and SVi (ml) were 23 ± 25 , 0.29 ± 0.22 , 99 ± 43 and 45 ± 16 respectively. Measures of RVEDVi were more closely correlated with indexed PRV ($r=0.549$; $p<0.001$) when compared with PRF ($r=0.480$; $p=0.001$). Similarly, RVSVi was more significantly correlated with PRV ($r=0.701$; $p<0.001$) than PRF ($r=0.605$; $p<0.001$). The difference of RVEDVi before and after the procedure significantly correlated both with PRF ($r=0.427$; $p=0.004$) and PRV ($r=0.489$; $p=0.001$).

Conclusions: PR expressed as an absolute volume is better able to predict RV dilation and RV stroke volume than PR expressed as fraction.

P-174

3D printed models for surgical planning in complex congenital heart diseases

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Introduction: A precise understanding of the anatomical structures of the heart and great vessels is essential for surgical planning in order to avoid unexpected findings.

Rapid prototyping techniques are used to print three-dimensional (3D) replicas of patients' cardiovascular anatomy based on 3D clinical images such as MRI and CT.

The purpose of this study is to explore the use of 3D patient-specific cardiovascular models using rapid prototyping techniques to improve surgical planning in patients with complex congenital heart disease.

Methods: This European prospective multicenter study included 8 patients with complex congenital heart diseases (Figure 1). Magnetic resonance imaging (MRI) and computed tomography (CT) were used to acquire 3D cardiovascular anatomy. Images were segmented and 3D mesh was created using AYRA software (IKIRIA, Spain). Fused deposition technique using polylactic acid was used. A Bland-Altman analysis was used to evaluate the

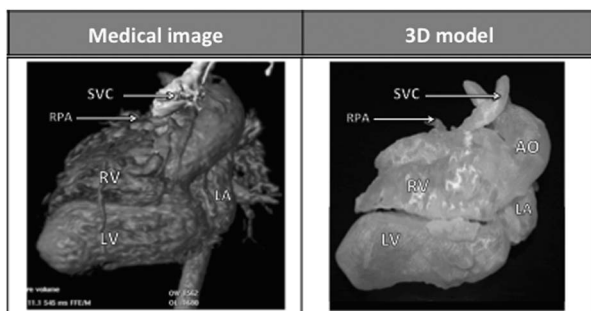


Figure 1.

diameters measurement agreement between the 3D printed model and the patient's MRI and CT. 3D-models were used to plan the surgery. After the procedure, surgeons involved filled a questionnaire form to evaluate the usefulness of the 3D printed model.

Results: The Bland-Altman analysis showed accurate agreement in the diameter between medical images and 3D-models (-0.12 ± 1.40 mm, mean bias \pm standard deviation, Figure 2). 3D-models showed the spatial relationships between the ventricular septal defect and great vessels (Case2, Case-6, Case-7, Case-8), re-appraisal for biventricular repair (Case-1, Case-8), planning of lateral tunnel completion (Case3), re-opening of a restrictive VSD and its relationship with the conductive tissue (Case4) and evaluation of RVOT aneurysm and pulmonary artery origin (Case 5). Surgeons found the 3D models to be very useful for surgical planning with an overall level of satisfaction of 8.5 out of 10, all agreed (score 4 out of 5) that 3D-models they were helpful to decrease possible surgical complications, strongly agree (score 5 out of 5) that would recommend it to other colleagues as well as teaching for trainees.

Conclusions: 3D-printed cardiovascular models accurately replicate the patient's anatomy and are extremely helpful for planning surgery in complex congenital heart disease. They may potentially reduce operative time and morbi-mortality.

P-175

Myocardial deformation imaging for assessing left ventricular function in patients with transposition of great arteries after arterial switch operation

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Background: This study aims to investigate the global and regional myocardial function of the LV by 2DSTE in patients with transposition of great arteries with intact ventricular septum (TGA) after arterial switch operation (ASO).

Methods: Prospective analysis of radial, circumferential and longitudinal deformation from echocardiograms in 2 groups: 58 children (5.7 ± 4 years) after neonatal ASO and 17 healthy controls (7.6 ± 4 years). Apical and basal short axis for 2D images were acquired (frame rate 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 (ϵ_{LL} , ϵ_{CC} , ϵ_{RR}) were extracted using a commercial software built on a 18-segment LV. A parametric paired samples T-test integrated in the statistical software SPSS was used, with $p \leq 0,05$ considered significant.

Results: Global longitudinal strain (GLS) measures of the LV were significantly different between the two groups (GLS $-16,42 \pm 3,08$ vs. $-19,29 \pm 2,17$, $p < 0,001$). There was no significant differences for global circumferential (GCS) and radial strain (GRS). In the measurements of LV function, there was a clear tendency toward a decrease in the ejection fraction ($63,6 \pm 4,1$ vs. $71,1 \pm 3,2$) with a concomitant increase in LV diastolic diameter (LVDD) z-score ($0,7 \pm 1,0$ vs. $-0,3 \pm 0,8$). The regional peak systolic longitudinal strain (LS) was reduced significantly, especially for segments 7 and 13 on anterior wall of LV, as well as the global longitudinal systolic strain and SR. There was no significant difference for regional circumferential (CS) and radial strain (RS).

Conclusions: Reduced GLS after ASO is associated with an unfavourable trend toward reduced pump function of LV. Decreased segmental LS values are a sign of regional hypokinesia with a possible local segmental coronary ischemia during surgery and LS

could be used for predictor of myocardial disturbances after ASO. Lack of significant difference for both circumferential and radial strain may be associated with still normal or slightly impaired global LV function and their changes may predict a newly onset failure of the ventricle.

P-176

Right ventricular longitudinal strain assessment in children with pulmonary arterial hypertension

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Objectives: Prognosis of patients with pulmonary arterial hypertension is closely related with right ventricular function. The aim of the study was to evaluate the role of speckle-tracking echocardiography in assessment of right ventricular function in children with pulmonary arterial hypertension compared to conventional echocardiography.

Methods: We evaluated prospectively 42 children (21 with severe pulmonary arterial hypertension on pulmonary vasodilator therapy – 16 associated with congenital heart disease and 5 idiopathic – and 21 age and sex match controls) using speckle-tracking echocardiography (Q LAB 10.0 software), conventional echocardiography, clinical parameters (6-min walking test) and biomarkers (B-type natriuretic peptide level). The right ventricular free wall and global longitudinal strain were determined by averaging three (basal, medial, apical) respectively seven regional peak systolic longitudinal strain. There were also evaluated the conventional echocardiographic parameters: tricuspid annular plane systolic excursion, right ventricular systolic/diastolic ratio, systolic right ventricular fractional area change, right ventricular myocardial performance index, left ventricular eccentricity index.

Results: Right ventricular free wall and global longitudinal strain were significantly lower (absolute value) in pulmonary arterial hypertension children than in controls ($p=0.0001$, $p=0.0001$ respectively $p=0.0001$). Right ventricular free wall and global longitudinal strain has correlated with right ventricular myocardial performance index, right ventricular fractional area change, left ventricular eccentricity index and also with B-type natriuretic level ($r=0.63$, $r=0.52$, $r=0.66$, $r=0.43$ respectively $r=0.62$, $r=0.48$, $r=0.65$, $r=0.41$ with a $p < 0.05$).

Conclusions: Right ventricular longitudinal strain assessment is useful in noninvasive evaluation of right ventricular function in children with pulmonary arterial hypertension.

Aknowledgement: The present study was carried out in the research project no 27/11.12.2013, financed by UMF Tg. Mures

P-177

Clinical Implication of Mitral Valve Geometry Alterations in Children with Dilated Cardiomyopathy

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Introduction: Functional mitral regurgitation (MR) severity is associated with poor prognosis in children with dilated cardiomyopathy (DCM), and surgical intervention for severe functional MR has been reported. In adult patients, preoperative assessment of mitral valve geometry predicts mitral valve durability after annuloplasty and valvuloplasty. However, few studies have

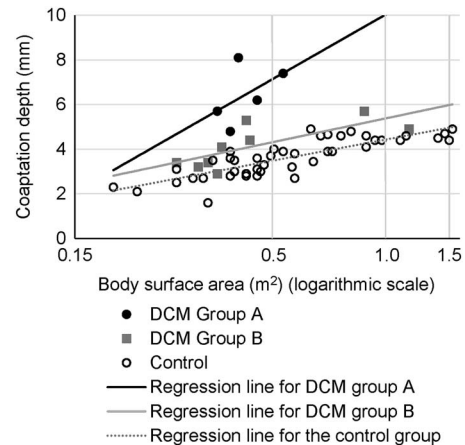


Figure.

described mitral valve geometry alteration in children with DCM. We aimed to elucidate the relationship between mitral valve geometry and clinical characteristics in children with DCM.

Methods: The medical records of 14 children with DCM were reviewed. Mitral valve geometry was evaluated by measuring coaptation depth (CD) using echocardiographic images of apical four-chamber view at the initial presentation. The patients were grouped into the DCM with moderate or severe functional MR (DCM group A, $n=5$) and DCM with less than moderate functional MR (DCM group B, $n=9$) groups. Measurements of 44 healthy children were used as normal controls. Analysis of covariance (ANCOVA) was performed to evaluate mitral valve geometry differences between groups.

Results: The median age of the patients with DCM was 1.2 years (range, 0.4–12.3 years). The figure shows the scatter plots of CD according to log-transformed body surface area (BSA). Both DCM groups showed significantly increased CD compared to the control group ($p < 0.0001$, ANCOVA). In addition, there was a significant difference in CD between DCM group A and DCM group B ($p < 0.001$, ANCOVA). Mitral repair was performed in three of the patients in DCM group A at a median 15 days (range, 13–113 days) after the initial presentation. The mitral repair led to the improved MR in two patients who were discharged on postoperative days 75 and 109. In the other patient, who showed the largest CD, the mitral repair failed to alleviate the MR, so mitral valve replacement was subsequently performed.

Conclusions: There was a significant mitral valve geometry alteration in children with DCM. The BSA-adjusted assessment of CD might be predictive of mitral repair efficacy and useful in determining the need for mitral valve replacement.

P-178

Temporal sequence of right ventricular function in children after tetralogy of Fallot repair: comparison of pulse wave Doppler echocardiography versus tissue Doppler imaging

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Objectives: The aim of the study was a temporal analysis of right ventricular (RV) function and its consequences for hemodynamic disturbances in children after tetralogy of Fallot repair (RTOF).

Methods: 52 RTOF children (mean age 13.7 ± 3.4) and 32 healthy controls (mean age 13.7 ± 2.9) were studied. The pulmonary valve opening time (PVO), ejection time (ET), and pulmonary valve closure time (PVC) were measured from pulmonary systolic outflow pulse wave Doppler patterns (PW). The tricuspid opening time (TVO), relaxation time (RT) and tricuspid closure time (TVC) were evaluated from PW of tricuspid inflow patterns. The contraction time (CT), relaxation time (RT), isovolumic relaxation (IVRT) and isovolumic contraction time (IVCT) by tissue Doppler imaging (TDI) at the RV free wall of the tricuspid annulus site were assessed. Heart rate (HR) was determined in all children on the basis of 3 consecutive echo cycles.

Results: There were not significant differences in HR between RTOF and healthy children. RTOF patients demonstrated increased PVO: 96.4 ± 23.4 ms vs 72.8 ± 17.4 ms ($p < 0.01$), ET: 334.7 ± 41.3 ms vs 236.4 ± 59.6 ms ($p < 0.01$), PVC 408.0 ± 44.6 ms vs 371 ± 44.2 ms ($p < 0.01$), TVO 453.0 ± 74.2 ms vs 419 ± 50.4 ms ($p < 0.01$) and TVC 783.7 ± 132.8 ms vs 745.0 ± 129.1 ms (NS) compared to the controls. RT was reduced in the RTOF comparing the healthy children, respectively 325.5 ± 128.1 ms vs 333.0 ± 102.1 ms (NS). In TDI both CT: 211.5 ± 52.1 ms vs 253.9 ± 38.6 ms ($p < 0.01$) and RT 414.2 ± 111.9 ms vs 420 ± 113.8 ms (NS) were reduced in the RTOF comparing to the controls. IVRT in the RTOF and healthy children was respectively 84.4 ± 41.2 ms vs 39.3 ± 11.3 ms ($p < 0.01$) and IVCT 113.2 ± 58.0 ms vs 60.9 ± 26.4 ms ($p < 0.01$). ET was statistically significantly longer than CT ($p < 0.01$) in the RTOF patients while in healthy controls significant difference between ET and CT was not found.

Conclusions: 1. The temporal sequence of right ventricular function is deeply disturbed in patients after tetralogy of Fallot repair. 2. The discrepancy between ejection time in PW and contraction time in TDI indicates the predominance of passive flow through the right ventricular outflow tract due to impaired right ventricular systolic function.

P-179

Spasm of Patent Ductus Arteriosus During Catheterization

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Background: In patients with patent ductus arteriosus (PDA), current treatment of choice and an alternative to the surgical repair is transcatheter closure. Intermittent spasm and subsequent patency of PDA has been described in the past, but it is rare in older children. We report our experience of ductal spasms that occurred during transcatheter occlusion of a PDA.

Methods: We reviewed all transcatheter PDA occlusions performed in our clinic. Retrospective data included: gestational age, age at catheterization, echocardiographic parameters before catheterization, PDA size (after spasm relief), and device specifications.

Results: Between January 2011 and March 2014, 150 transcatheter PDA closure procedures were performed. Among them, 5 patients had ductal spasm during the procedure. All of these patients receiving oxygen supplement initially during the procedure. Median age at the procedure was and gestational age 17 months. All of patients were born prematurely and median gestational age 30 weeks. Physical examination revealed continuous machinery murmur and precordial thrill over the infraclavicular area in all of the patients. When reauscultated (4 of 5), murmurs were disappeared during spasm. In one patient the device had embolized to the descending aorta and one day later embolized device removed

Table 1. Diagnostic and Demographic Variables of PDA Spasm Patients

Patient	Birth Weight (kg)	Gestational Age (wk)	Age at Procedure (mo)	Weight at Procedure (kg)	Echocardiography	
					PDA MLD (mm)	Left Heart Volume overload
1	2.32	35	17	9	4	Yes
2	1.60	30	12	8.5	4.8	Yes
3	2.10	34	36	17	3.5	Yes
4	1.24	28	16	10	4.5	Yes
5	1.28	28	17	10.3	3	Yes

and another device was placed. All PDAs were closed with an 8-mm or larger Amplatzer Vascular Plug-II devices, finally (Table 1). **Conclusions:** Ductal spasm is a frequent condition, in premature patients receiving supplemental oxygen even during transcatheter PDA closure. Clinicians should consider ductal spasm, if the physical examination and echocardiographic findings of the ductus did not overlap with the catheter angiography findings.

P-180

The evaluation of diagnostic and interventional cardiac catheterization procedures of patients under extracorporeal membrane oxygenation support

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Introduction: The extracorporeal membrane oxygenation (ECMO) support has been using increasingly after cardiopulmonary arrest or postoperatively in cardiac surgery. ECMO is an useful method in patients that cannot be weaned from cardiopulmonary bypass and to adapt to the recent hemodynamic alterations in early postoperative period. Cardiac catheterization might be indicated for clarification of the underlying pathology or for treatment of these patients supported by ECMO.

Methods: Between 2010 and 2014, 1420 patients with congenital heart disease underwent cardiac surgery and 71 of these patients (5%) needed ECMO support after surgery. Diagnostic or interventional cardiac catheterization was performed in 6 (8.45%) of these 71 patients.

Results: The diagnostic catheterization was performed in 2 of the patients while invasive procedures were performed in 4 of the patients. In patients with antegrade flow, ECMO cannulas were clipped before the injections for higher quality. Left pulmonary arterial stenting was performed in 3 of the patients and balloon angioplasty of both pulmonary arteries was performed in one of the four patients during interventional catheterization procedure. BT shunt recanalization was performed simultaneously with left pulmonary artery stenting in one of these patients. There were no complications recorded during transportation to the catheterization laboratory or during catheterization. One of the two patients after the diagnostic catheterization was referred to surgery for pulmonary artery reconstruction and the other one for correction of supra-valvular aortic stenosis and left main coronary artery stenosis. Four patients were successfully weaned from ECMO after the procedures but 2 patients died under ECMO support during the ICU stay.

Conclusions: ECMO can be life saving as a solution of hemodynamic problems after congenital heart surgery. If the patients cannot be weaned from ECMO support due to hemodynamic problems, catheter angiography should be performed urgently.

Suitable transportation conditions, extensive logistic support together with experienced staff, diagnostic and invasive cardiac catheterization can be performed with minimal risk in these patients. We think that for better and high quality views in patients with antegrade flow, who cannot be weaned from ECMO support, ECMO cannulas can be clipped just before the injections.

P-181

Transcatheter atrial septal defect and patent foramen ovale closure experiences in children, evaluation of short, intermediate and long term outcomes

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Objectives: In this study, we aimed to evaluate transcatheter atrial septal defect (ASD) and patent foramen ovale (PFO) closure experiences, short, intermediate and long term results and investigate the effects of different methods on procedural success and complications.

Methods: After the reviewing of archive records for transcatheter ASD or PFO closure procedures between 2000–2013, 506 cases included for study, of 90 cases files that containing data about follow-up and diagnosis before the procedure could not be reached. Patients and procedural characteristics, post-procedural follow up datas were evaluated.

Results: Male:female ratio was 1:1,3, mean age and weight was $8,6 \pm 5,8$ years and $28,9 \pm 16,4$ kg respectively during procedure, mean follow-up period was $6,05 \pm 3,7$ years (1 month – 13,5 years). Mean procedure and fluoroscopy duration was $61,5 \pm 23,5$ and $7,8 \pm 6,7$ minutes respectively. The procedures were performed under transthoracic echo (TTE) guidance in 90 (17,8%) cases, balloon sizing used in 214 (42%) cases. Pulmonary hypertension was observed in 67 (16%) cases. Within closure attempted 416 cases, the procedure was successful in 401 (96,3%). Procedure-fluoroscopy durations were shorter, defect and device sizes were smaller in successful group than unsuccessful group ($p < 0,05$). The presence of deficient rim and use of “balloon sizing” were not influential on procedural success. In balloon sizing group, longer procedure-fluoroscopy duration and lower total septum/device ratio were observed ($p < 0,05$). In selected cases, it was found that TTE guidance shorten the procedure-fluoroscopy duration ($p < 0,05$). Residual shunt was seen in 32,4% and 0,9% of patients immediately after procedure and at the end of the follow-up respectively. Major (rescue surgery, thrombus, erosion) and minor (most frequently rhythm disorders) complications rate were %1,4 and %1,8 respectively. No embolisation and mortality were observed. Benign holter abnormalities were observed in 7,7% of patients.

Conclusions: Transcatheter ASD and PFO closure is a safe and effective method. Reducing the fluoroscopy time and not requiring general anesthesia are the additional advantages of the defect closure with TTE guidance in the presence of sufficient rim and centrally localized defect.

P-182

Transcatheter catheter closure of patent ductus arteriosus in small pediatric patients from an exclusive transvenous approach using combined angiographic and echocardiographic guidance

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Introduction: Transcatheter closure is the treatment of choice for the majority of patients with a patent ductus arteriosus (PDA). However, the standard technique of this procedure may be associated with arterial complications, particularly, in small pediatric patients. The aim of this study was to report experience with catheter closure of PDA in 33 consecutive small children using an exclusive venous approach.

Methods: The age of the patients ranged from 6–36 months and the weight from 6–13 kg. The anatomy and size of PDA were defined by transvenous retrograde aortography using a Pigtail or a Berman catheter. The PDA occluder was implanted through a 7 F (ADO I) and 5 F (ADO II, ADO AS) delivery sheath (DS) in 10 and 23 patients, respectively. The procedure was guided using hand injections of contrast media through the DS and 2D and color Doppler echocardiography from suprasternal and parasternal long and short axis, respectively.

Results: The PDA occluders were permanently implanted in all 33 patients. The mean PDA diameter (at the pulmonary end) was $3,8 \pm 0,9$ mm (range, 2,5 to 5,2 mm). The mean device diameter was 5 ± 1 mm (range 4 to 6 mm) and $4,2 \pm 1,5$ (range, 3–6 mm) for the ADO I and the ADO II occluders. Complete echocardiographic closure of the ductus at 1-month follow-up was observed in all 33 patients (100%). Immediately after the procedure there was a mild left pulmonary stenosis (peak pressure gradient of ranging from 6–8 mm Hg), in 3 patients. Five minor groin hematomas were the only complications of the procedure.

Conclusions: Exclusive transvenous PDA occlusion using combined angiographic and echocardiographic guidance is an effective and safe method that prevents the arterial complications of the standard approach in small children. In contrast to ADO I the ADO II, AS occluders due to their low profile can be delivered through a 5 F delivery sheath which facilitates crossing of PDA and the injections of contrast medium for guidance of the procedure.

P-183

Percutaneous transluminal balloon dilation and surgical valvotomy comparative analysis in non-critical congenital aortic valve stenosis

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Introduction: One third of patients with non-critical congenital aortic valve stenosis need intervention, either transcatheter percutaneous balloon aortic valvoplasty (BAV) or surgical aortic valvotomy (SAV).

Methods: The aim of the study was to evaluate our 26 years (1987–2013) experience in performing BAVs and SAVs. After excluding critically ill newborns, 62 consecutive interventions were included in the study (39 BAVs and 23 SAVs).

Results: BAV patients were older, but there was no difference between groups according to sex, valve function and morphology. Suboptimal acute outcome was registered in 4/39 BAVs. Three patients did not survive early period after surgery. Significant peak instantaneous pressure gradient reduction (47% vs. 50%, $p = 0,211$), and mild aortic regurgitation increment ($0,5 +$ vs. $1 +$, $p = 0,385$) were registered after BAV and SAV. Patients with residual peak instantaneous gradient of ≤ 50 mm Hg (peak to peak gradient of ≤ 31 mm Hg) and none/mild aortic regurgitation after BAV, and patients with none/trivial aortic regurgitation before surgery had longer reintervention free survival. Follow-up period was $7,0 \pm 5,4$ (up to 18,8) years in the BAV group and $9,0 \pm 8,0$ (up to 23,9) years after surgery. There were no late aortic stenosis related deaths in both groups. A peak instantaneous gradient

increase was higher by 8 mm Hg in BAV versus SAV group over time. Similar aortic regurgitation progression was registered in both groups. There was no difference between BAV and SAV groups with respect to long-term echocardiographic, electrocardiographic and radiographic heart function parameters. Left ventricular end-diastolic diameter and aortic regurgitation grade, hypertrophy and pressure gradient, contractility (in the BAV group) and follow-up period associations were showed. Reintervention-free survival was 12.0 (95% CI: 9.3–14.7) and 14.5 (95% CI: 10.0–18.9) years ($p = 0.733$), and survival without aortic valve replacement was 15.2 (95% CI: 12.5–17.9) and 17.4 (95% CI: 13.1–21.7) years ($p = 0.877$) after BAV and SAV respectively.

Conclusions: Both early and late follow-up results in patients with non-critical congenital aortic stenosis are similar after BAV and SAV. However, BAV is a less invasive procedure and long-term results are slightly better after surgery.

P-184

Safety and efficacy of cutting balloon angioplasty for patients with congenital heart diseases

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Background: Cutting balloon angioplasty (CBA) is performed for difficult vascular stenoses that are resistant to standard balloon angioplasty. In this study we investigate the safety and efficacy of CBA performed at our institute in the past 5 years.

Methods: Forty-seven patients underwent a CBA. The most common diagnosis was pulmonary atresia with ventricular septal defect: 59.6%, followed by Williams-Beuren syndrome: 10.6%. The median age was 3.3 years (range 1.5–46.4) and the median weight 12 kg (range 4–88). All procedures were performed under sedation. The CBA was performed in 35 patients in pulmonary artery stenoses, in 5 patients in major aortopulmonary collateral arteries, in 3 patients in the superior caval vein, in 1 in a Pott's shunt, in 1 in an iliac artery and in 1 in a renal artery. The size of the cutting balloons used was 3.5 to 8 mm.

Results: The procedure was successful in 36 patients (76.6%), measured as a) a significant reduction of gradient across the stenosis and/or reduction in right ventricular pressure (20 patients), b) increased oxygen saturation in cyanotic patients (9 patients), or c) recruitment of new small vessels (7 patients). Postprocedural thrombosis occurred in 3 patients (successfully treated with lysis and subsequent balloon angioplasty).

Conclusion: CBA is a safe and effective treatment for resistant stenosis and can be performed without significant complications.

P-185

Preliminary follow up of the use of Valeo stent for coarctation stenting in children.

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Objective: To evaluate the performance of the Valeo stent in the management of coarctation of the aorta in children.

Methods: Between July 2012 and August 2014, coarctation stenting with the Valeo Premounted Re-dilatable Stent was undertaken in 5 children. Data including demographic, angiographic and echocardiographic imaging and clinical outcome were reviewed retrospectively.

Results: Median age at the time of procedure was 5.1 years (range 3.6 to 7.1 years) and median weight was 18.3 kg (range 15.3–27 kg). The youngest patient presented following an intracerebral haemorrhage secondary to severe systemic hypertension; one

patient had William's syndrome with fracture and re-stenosis of previous coarctation stent; one had complex congenital heart disease with re-coarctation after previous surgical repair and two others had isolated coarctation. All stents were 10 mm × 26 mm in size and delivered via a 7 F sheath. There was improvement in median coarctation diameter from 4.9 mm (range 1.8–7.4 mm) to 9.8 mm (range 5.8–12.7 mm), $p < 0.01$; and a reduction in the median peak pressure gradient across the coarctation from 30 mmHg (range 20–40 mmHg) to 8 mmHg (range 0–15 mmHg), $p < 0.01$. Median percent stent recoil in the middle of the stent was 1.5% (range 0.5–19%). All implants were successful with no vascular complications. All patients had a CT angiogram at a median time of 3 months (range 2.7–10.2 months) post procedure which showed no aortic wall complications. Follow-up was a median 16.6 months (range 4.7–30 months). At the last follow up, echocardiographic findings were no different from discharge in 4 patients; one patient had gradual increase in peak gradient of the descending aorta on echocardiography and repeat catheterisation at 13.1 months post procedure showed no stent stenosis but narrowing of the transverse arch. Four children required antihypertensive medications and 1 was off medication at latest follow-up.

Conclusions: The Valeo stent is low profile, has adequate radial strength and can be post dilated up to adult size (20 mm). The use of Valeo stent for treatment of coarctation in childhood is safe and effective in the early term. However, further study is required to determine longer-term stent efficacy.

P-186

Successful Operation of Giant Thrombus Formation One Year after Percutaneous Closure of an Atrial Septal Defect with an Amplatzer Septal Occluder

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Introduction: Although a number of transcatheter closure devices have been developed the most commonly used device is the Amplatzer® Occluder worldwide. The overall incidence of complications associated with these devices has been reported to be between 6–11%. Of these complications, thrombus formation that was usually observed 4 weeks after the procedure was reported as 1.2% in ASD patients. Herein, we report a late presentation of a giant thrombus formation on the left atrial disc of the device presenting 1 year after device implantation.

Case Report: A 17-year-old patient underwent a TEE-guided transcatheter closure of the atrial septal defect under general anesthesia using a 18 mm Amplatzer® Septal Occluder without residual shunt. Aspirin 300 mg/day had been stopped after 6 months.

At the 1-year follow-up, he was evaluated with transthoracic echocardiography which showed a left atrial thrombus attached to the ASD closure device. The diameter of thrombus was 14 × 62 mm. He was taken to surgery for removal of the thrombus and the device. The surgical approach was achieved via a median sternotomy and institution on cardiopulmonary bypass. After right atriotomy, well endothelialized occluder device was seen and excised with large thrombus. There was no device fracture or dislocation. The thrombus and device explanted and sent to pathological examination. The novel created atrial septal defect was closed by pericardial patch. The patient had an uneventful recovery and the studies continues on the etiology of hypercoagulability states.

Conclusion: To our knowledge, this is the first report of late thrombus formation associated with Amplatzer Septal Occluder device. It was emphasized that the patients should be followed-up longer periods after device closure in terms of thrombus formation.

P-187**Transcatheter closure of ventricular septal defect: different types and different devices.**

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Background: The most common congenital heart disease is VSD. Surgical repair is widely accepted, but still carries a small but definite risk of morbidity and mortality.

The aim of this work is to report our initial experience in transcatheter closure of VSD using different types of devices.

Methods: Between January 2013 and December 2014, a total of 40 patients with VSD underwent an attempt of transcatheter closure under trans-esophageal echocardiographic guidance.

Results: The median age was 3.5 years and median weight was 14.25 kg. The median VSD size in echocardiography was 5 mm (ranging from 2.5 to 12 mm), while by angiography it was 5.2 mm. The type of VSD was: perimembranous in 25 cases, high muscular in 7, midmuscular in 4 & apical in 4 cases. Indications for closure were: pulmonary hypertension in 8 patients, LV dilatation in 29 patients, history of previous infective endocarditis in two patients, residual VSD S/P surgical closure in 3 patients and one patient had residual VSD after previous device closure.

Device implantation was accomplished for 33 patients (82.5%) with immediate closure in 30 patients (75%) while three patients showed minimal residual flow. The procedure was aborted for 6 patients due to inability to cross the defect while in another patient temporary heart block developed with implantation of the device. The types of devices used were: ADO I in 15 patients, ADO II in 9 patients, PFM in 6 patients, muscular VSD device in 4 patients and cribriform ASD device in 1 patient. The immediate success rate was: 93.3% with ADO I, 87.5% with ADO II, 80% with PFM and 100% with the muscular device. One patient with a small residual after implantation of a PFM coil, developed prolonged hemolysis, the defect was surgically closed with device removal. Two patients developed new mild mitral regurgite. The approach was from the RV without arteriovenous loop in 14 cases closed by ADOI (94%) so the procedure was shorter with less fluoroscopy.

Conclusion: Transcatheter closure of different types of VSDs seems to be effective and safe using different devices. Long-term results are yet to be evaluated.

P-188**Patent ductus venosus causing cyanosis after Fontan operation - successful transcatheter-based flow reduction to induce growth of rarefied intrahepatic portal veins allowing for ultimate interventional closure**

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Background: Pulmonary arteriovenous fistulas (PAVM) leading to cyanosis after bidirectional superior cavopulmonary anastomosis are considered to be due to hepatic venous blood bypassing the lungs. Here report for the first time that a patent ductus venosus (PDV), a persistent fetal connection between the left portal vein and the inferior cava, should be considered as a cause for PAVM after Fontan operation.

Case report: PAVM were diagnosed in a 9-year-old boy with episodes of dizziness developing cyanosis after modified Fontan

procedure for palliation of pulmonary atresia with intact ventricular septum. During color Doppler examination of hepatic flow a PDV was diagnosed. Via subclavian venous access and a retrograde transcaval route a portal venogram was obtained showing rarefied intrahepatic portal veins (PV). PV pressure increased from 12 to 26 mmHg during temporary PDV balloon occlusion. To avoid PV congestion leading to portal thrombosis, flow reduction of the PDV was achieved by transcatheter placement of a diabolo-shaped covered stent-ensemble consisting of a 5 × 12 mm Formula renal stent centrally placed on a 16 × 41 mm covered Advanta V12 stent yielding a gradient of 5 mm Hg between PV and inferior vena cava. Repeat portal venogram 5 months later showed increased vascularity of the PV system and a minor PV pressure increase from 13 to 16 mmHg during temporary PDV balloon occlusion. After ultimate closure of PDV with an 8 mm AVP II, elevated levated blood ammonia levels normalized, episodes of dizziness disappeared and tcSaO₂ gradually increased from 80-85 to 85-90% during the following 3 months.

Conclusion: PDV should be considered as a possible cause for unexplained cyanosis in Fontan patients with PAVM. Transcatheter-based flow reduction of PDV is feasible and appears to induce growth of rarefied intrahepatic portal veins allowing for ultimate interventional closure.

P-189**Role of persistent arterial duct for treatment of neonatal Scimitar syndrome with suprasystemic pulmonary hypertension**

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Introduction: Right ventricular (RV) failure is one of the causes for a poor outcome of neonatal Scimitar syndrome with suprasystemic pulmonary hypertension (ssPH). Here we report on our strategy to use the persistent arterial duct (PDA) for RV decompression.

Case reports: Two preterm neonates (Pt. 1 31, Pt. 2 35 weeks of gestation) were diagnosed of scimitar syndrome with complete anomalous right pulmonary venous drainage to inferior vena cava, persistent primitive hepatic venous plexus (PPHVP), right pulmonary artery hypoplasia, anomalous systemic arterial supply of parts of the right lung, secundum atrial septal defect (ASD) and PDA. In both patients, anomalous systemic arteries were occluded by transcatheter embolization. In the presence of ssPH despite optimal PH-specific medical therapy, interventional balloon angioplasty of subtotally closed PDA was performed for acute RV decompression. After surgical repair with a pericardial intraatrial baffle and subtotal ASD closure, systolic pulmonary artery pressure declined to half systemic allowing for PDA closure in both patients. In Pt. 1, transcatheter occlusion of a hepatic vein of the PPHVP draining into the Scimitar vein was also performed. At age of 6 months there is no echocardiographic evidence for PH in Pt. 1. In Pt. 2, two restrictive muscular ventricular septal defects leading to increased pulmonary flow were interventionally closed. However, development and recurrence of left pulmonary and Scimitar vein stenosis despite multiple transcatheter interventions led to reappearance of systemic PH in Pt. 2. This patient is in a stable condition at age of 16 months.

Conclusion: In neonatal Scimitar syndrome with suprasystemic PH, balloon angioplasty of subtotally closed PDA may prevent early RV failure and thus improve the poor outcome of this entity.

P-190

Successful transapical Melody valve implant in a mitral bioprosthesis in a 3-year old child with dextrocardia and Fontan palliation

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Background: Fontan failure with consecutive bronchitis fibroplastica or exudative enteropathy might be caused by pre- or postcapillary pulmonary vascular obstruction or by a mixed form. We describe the first-in-child transapical melody valve-in-valve (ViV) implantation in complex congenital heart disease with failing Fontan circulation.

Case report: A 3.5 year old, 15 kg weighing girl was referred for evaluation of heart transplantation. She presented with bronchitis fibroplastica (BF) based on secondary pre- (23 mmHg mean pulmonary artery pressure, PAP) and post capillary (PCWP, 14 mmHg) hypertension within the Fontan circulation. Based on a complex anatomy consisting of situs inversus with dextrocardia, asplenia and imbalanced atrio-ventricular (AV) septal defects and transposition of the great arteries bidirectional Glenn shunt followed by a fenestrated 16 mm extra-cardiac Fontan completion with the need of re-fenestration was performed. Additionally, due to a severe AV regurgitation an artificial tricuspid valve atresia together with implantation of 27 mm mitral valve bioprosthesis became necessary. In a follow-up of six months the artificial valve showed a severe regurgitation, again. Considering the clinical situation, the echocardiography and magnetic resonance data a customized 22 mm Melody valve was implanted through a minimally invasive right intercostal thoracotomy and an 21 F sheath. The intervention was performed without complications, despite the BF she recovered well, and the transthoracic echo at her discharge home revealed a minimal residual regurgitation without any trans-valvular pressure gradient.

Conclusion: The transapical implantation of a Melody valve allowed successful sealing following a ViV, but the anatomical dimension of the patient with body surface area of about 0.65qm was borderline.

P-191

Serum levels after everolimus-eluting stent implantation in infants with pulmonary vein stenosis

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Introduction: Everolimus-eluting stents are used in adults for interventional treatment of coronary artery disease and in small vessel disease to reduce restenosis rate. The antiproliferative substance everolimus is acting locally but is also released into the circulation. Data on their use in pediatrics concerning systemic substance levels and side effects are limited. We report systemic drug exposure after implantation of a single everolimus-eluting stent in three infants (age in months/body weight in kilograms: patient 1: 8/4.6, patient 2: 6/3.9, patient 3: 1/3.5), respectively, which were used for relief of pulmonary vein stenosis.

Methods: Xience® everolimus-eluting stents (Abbott Laboratories) were used. Each stent had different size and device drug amounts (stent diameter x stent length in mm and device drug amount in µg: patient 1: 4 × 18 mm, 113 µg; patient 2: 2.5 × 8 mm, 40 µg; patient 3: 3 × 8 mm, 40 µg). Everolimus levels were measured by

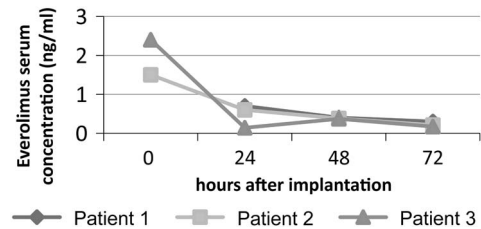


Figure.

liquid chromatography-tandem mass spectrometry (LC-MS/MS) 1 hour (except patient 1), 24, 48 and 72 hours after stent implantation.

Results: Patient 1 was exposed to a maximal calculated everolimus dose of 24.6 µg/kg, patient 2 to 10.3 µg/kg and patient 3 to 11.4 µg/kg, respectively. The highest serum everolimus level was measured 1 hour after stent implantation in the youngest and smallest child (patient 3; 2.4 ng/ml) followed by patient 2 with 1.5 ng/ml. Subsequently, serum everolimus levels decreased continuously (Figure). After 48 hours, everolimus levels were below the lower laboratory limit (<0.5 ng/ml) at our institution in each of the patients, but everolimus was still reliably detectable by repeated LC-MS/MS. At anytime, everolimus levels were below the immunosuppressive therapeutic range of 3–6 ng/ml. Complete blood count, creatinine, C-reactive protein, and liver enzymes did not change as compared to patient laboratory parameters prior to catheter intervention. Drug toxicity was not observed.

Conclusions: After everolimus-eluting stent implantation in infants we documented always subtherapeutic serum levels. Highest everolimus levels were measured in the first hour after implantation, thereafter rapidly decreasing. These data suggest that at least one everolimus-eluting stent can safely be used in small infants for therapy of pulmonary vein stenosis without systemic adverse effects.

P-192

Immediate results of PDA stenting in duct dependant pulmonary circulation. Egyptian experience

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Introduction: BT shunt used to be the first line for palliative treatment in duct dependant pulmonary circulation lesions. However, this procedure has a lot of complications and long waiting list. The aim of our study was to assess the efficacy, safety and immediate outcome of ductal stenting in neonates & young infants with duct-dependent pulmonary circulation.

Methods: All patients with duct dependant pulmonary circulation who were unable to undergo a shunt operation and were referred to Cairo University Pediatric Cardiology Division, during the period from April 2008 to February 2011 were included in the study. Patients with bleeding tendency, sepsis, Hyaline Membrane Disease and pneumonia were excluded.

Results: Ductal stenting was attempted for 56 patients with duct dependant pulmonary circulation. 46 (82.1%) patients had univentricular physiology and 10 (17.9%) patients had biventricular physiology. The mean age was 33.7 days (5–210 days), and the mean weight was 3.6 ± 0.76 Kg.

The stent diameter ranged from 3–4 mm, stent length ranged from 12 – 28 mm (19 ± 3.2). 55/56 cases were done through a femoral

access and one case through an axillary axis, fluoroscopy time was 24.7 ± 18 min, total procedure time was 88.5 ± 36.5 min.

The success rate was 64% (36/56), 23/26 were in horizontal straight ducts, 3/5 in vertical straight ducts, 1/2 in horizontal tortuous ducts and 9/25 in vertical tortuous ducts, Oxygen saturation improved post stenting from $65 \pm 5.79\%$ to $85.2 \pm 19.9\%$. Mean post stent hospital stay was 10.6 ± 4.5 days. There were 3 mortalities in the 36 stented cases (8.3%), femoral artery occlusion occurred in 6/36 cases (16.6%), stent displacement in 2/36 cases (5.5%), acute stent thrombosis in 2/36 cases (5.5%), septicemia occurred in one case (2.7%) with no evidence of pulmonary overshunting in any of our cases.

Conclusion: PDA stenting is a life saving procedure, that should be attempted even in tortuous ducts. Proper preparation of the patient, anaesthetic management and post-stenting ICU management are mandatory for success. Surgical intervention should be done early after successful stenting to guarantee good surgical outcome before desaturation.

P-193

Transcatheter interventions in neonates with critical pulmonary stenosis in the era of duct stenting

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Introduction: Critical pulmonary stenosis (CPS) is defined as severe PS in a newborn with cyanosis or signs of low cardiac output at presentation. Pulmonary blood flow is mostly duct dependent. We present our experience with CPS between 2005 and 2014.

Method: 57 patients aged between 2-28 days (median 7 days) underwent pulmonary balloon valvuloplasty (PBV). Duct stenting was performed immediate after in patients that hypoxemia persists (<75%) and had smaller RV and RVOT. In good RV morphology or non-constricted PDA, we waited for one week on prostaglandin (PGE) infusion. If hypoxemia persists longer, stent was implanted.

Results: In 57 patients, 47 of them were duct dependent, 10 of them have significant hypoxemia but duct had been occluded. The procedure was successful in 56 of 57 (98%). In one patient we couldn't cross the valve and underwent to surgery. Predilation with coronary balloons in 10 and snare assisted technique was needed in two for crossing the valve with the final balloon. 20 newborn needed duct stenting; 14 in the same and 6 in subsequent session. Duct spontaneously occluded in two when waiting on PGE, recanalized and stented in one and surgical shunt was performed in another. The mean Z scores and valve diameters in duct stent group were significantly lower for both tricuspid and pulmonary valve than the others'. In one patient in whom pericardial effusion was developed, effusion was drained and the procedure was completed. One patient developed persistent complete heart block; PBV was performed retrograde through the duct while RV pacing. The patient died in spite of successful PBV on the fourth days in intensive care unit. During the follow up (median 57 months), transcatheter reintervention was performed in 9; PBV due to recurrent PS in 5, stent redilation in 3, transcatheter shunt occlusion in one. Surgical interventions were needed in 4; RVOT reconstruction in two, Glenn anastomosis in two. Severe pulmonary regurgitation was seen in two but no need valve replacement yet.

Conclusion: Although additional interventions are not uncommon in early and intermediate time after the procedure, PBV should be the first choice in newborn with CPS.

P-194

Transcatheter closure of secundum ASD with Cocoon septal occluder in children; early and intermediate term results

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Introduction: The aim of this study is to investigate the efficacy and safety of Cocoon septal occluder (CoSO) in ASD closure, which has similar characteristics with Amplatzer septal occluders (ASO) but low cost than it.

Methods: From January 2004 to December 2014, 724 patients underwent transcatheter ASD closure with various devices. Initially we used ASO in 144 patients. The other devices used were 434 Cardiofix septal occluders, 29 Ceraflex, 13 Cardia, 9 Figulla Occlutech, 7 Gore Helex, 2 Hyperion, 2 PFM septal occluders later. Nowadays, we have been using predominantly CoSO in ASD closure. We evaluated the efficacy and safety of CoSO in ASD closure in patients whose ages <18 years and compared with the patients' that ASO was used in regarding to procedural success, complications, procedure and fluoroscopy times, complete occlusion rate in one month. Only transthoracic echocardiography (TTE) was performed before the procedure in all, but TTE or transesophageal echocardiographic guidance was preferred according to complexity of the defect or echogenicity of the patient during the procedure.

Results: We have used CoSO in 76 and ASO in 87 children. Age and weight of the patient in CoSO and ASO groups were comparable as 8.5 ± 3.9 vs 8.2 ± 3.3 years, 29.8 ± 15.5 vs 27.1 ± 11.2 kg, respectively. Mean size of defects and devices were not statistically different between the groups; 15.1 ± 5 vs 15.2 ± 3.8 , p: 0.85 and 19 ± 5.5 vs 20 ± 4.8 , p: 0.95; respectively. The number of TTE guidance was 35 in CoSO and 45 in ASO. The complex defect rate and deployment technique was similar in both groups. Procedural success rate was %100 in both groups. Procedure and fluoroscopy time were significantly lower in CoSO group (48.6 ± 21.6 vs 66 ± 28.1 ; p: 0.008 and 8.8 ± 5.7 vs 16.6 ± 11.6 ; p < 0.001, respectively). Complications were minimal and 2 in CoSO and 2 in ASO. Complete occlusion rate was similar as %97 in CoSO and %99 in ASO in one month. No complication has occurred during the intermediate term follow-up in both.

Conclusions: Our early and intermediate term results showed that CoSO is efficient and safe alternative in transcatheter ASD closure. It can be preferred because of low-cost.

P-195

Interventional treatment of postsurgical Gerbode-type defect

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Introduction: The communication between the left ventricle and right atrium, known as Gerbode type defect, is rare cardiac anomaly. There is increasing number of adult patients with postsurgical defects after mitral or aortic valves replacement. The traditional method of treatment is surgical closure of defect.

Material and Method: A 11-year-old male was referred for increasing deterioration of physical activity. He was born with congenital cardiac anomaly: d-transposition of the great arteries, inter-ventricular defect and severe subpulmonary stenosis. At 21 months of age, he underwent Rastelli operation. After 6 years next corrective operation was performed due to increasing stenosis in the

right and left ventricle outlet tracts. Two years later he was admitted to our hospital again due to mild deterioration of physical capacity and the incidences of supraventricular arrhythmias. The left ventricle to the right atrium shunt with a defect measuring 5,5 mm was diagnosed. Cardiac catheterization was performed to evaluate the hemodynamic state with intention of percutaneous therapy. A pulmonary to systemic blood flow ratio was 1,9:1,0. We decided to implant Amplatzer Duct Occluder II 6-4. Unfortunately after placing the device in the defect both angiography and echocardiography revealed significant shunt through the implant. Therefore the implant was removed and Amplatzer Duct Occluder 10/8 (9-PDA-006, St Jude) was introduced from the right femoral vein successfully during the same procedure.

Results: The retention skirt was placed just below aortic valve and the right sided part of implant did not interfere with tricuspid valve. Both echocardiography and ventriculography showed good position of the device with minimal residual shunt. No arrhythmia or conduction disturbances were noted during whole procedure. At follow-up the patient remains free of symptoms. At the beginning the small residual leak was observed but at the last follow-up it disappeared completely.

Conclusion: The development of cardiac surgery is associated with the formation of various complications. Each subsequent surgical correction increases the operational risk. Percutaneous treatment of iatrogenic Gerbode type defects is an attractive and feasible method of treatment.

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Percutaneous closure of the complex interatrial defects using the modified balloon technique

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Introduction: Percutaneous closure of interatrial defects is more widely used method of treatment. It also applies to complex type defects. There are used various technical modifications in these situations.

Materials and Methods: In the period of 43 months in 27 patients with complex interatrial defects aged from 35 months to 67 years (mean $23,6 \pm 25,3$ years) there were performed unconventional methods of percutaneous closure of these defects. Stretched diameters of defects ranged from 19 to 35 mm (mean $26,9 \pm 5,1$). The smaller defects occurred mainly in the smaller children. In 19 patients (70%) there was a lack of aortic rim.

In all these patients, despite the use of multiple attempts (classical, the use of different curvatures of delivery system, left pulmonary vein and left atrial roof technique), we could not get correct position of discs of the implants in relation to the septum, because the left atrial disk constantly prolapsed into the right atrium. Therefore, the second contralateral venous access was obtained, through which the calibration balloon was introduced. When the balloon was partially inflated, both discs of the implant were released. Then the desinflation was made, the balloon and guidewire were removed subsequently.

Results: The defects were closed successfully in 25/27 (92,6%) patients. The trivial or small residual leak was observed in 7/25 (28%) patients, which disappeared in all cases during the follow-up (mean $16,5 \pm 11,4$ months). In the remaining 2 patients, despite many attempts to percutaneous closure, it was impossible to get proper placement of the implant. Therefore, the procedure was abandoned. In both cases, the surgical correction was performed.

Conclusion: Modified balloon technique is extremely useful method that significantly increases the effectiveness of the percutaneous closure of complex interatrial defects. This method is applicable both in adult patients and small children.

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ADO II in Percutaneous VSD Closure of Pediatric

Patients:

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Background: It is hard to find an ideal device to use for every VSD successfully. If inappropriate device was chosen; complication rate increases, procedure time gets longer that prolongs exposure to ionizing radiation. Therefore interventionalists are in the search for new ideal devices. Main aim of our study to show that ADO-II device can be used for small ventricular septal defects successfully, safely with low complication rates.

Methods and Results: Between the dates April 2011- October 2014, 17 VSD closures with ADO-II device. Actually there were 16 patients but one of the patient had 2 perimembranous defects which were closed separately. Patients having muscular and perimembranous VSD with hemodynamically significant left to right shunt detected by clinical examination and echocardiography were included in the study.

Results: Age of patients ranged between 3-18 years. Weight of the patients was between 14-76 kg. VSD diameter ranges between 2-6.7 mm (3.75 ± 1.25). One of them was muscular, eighteen of them were perimembranous type. Fourteen of the perimembranous defects were aneurysmatic, tunnel shaped. We have used mostly venous route (12 patients) for closure. One of the patients had two separate VSDs. The distance between two defects were 7 mm. Therefore we have used two separate devices to occlude them. One of the defect was 3.2 mm, occluded with ADO II sized 5×4 from arterial side. The other one was 3.4 mm width and closed with 5×6 mm ADO II from venous side. All cases were successfully closed, no major complications were reported. There was no incidence of left bundle branch block, P-R prolongation, or complete heart block.

Conclusion: Perimembranous aneurysmatic ventricular septal defects are difficult, risky for percutaneous closure because of its proximity to aortic, atrioventricular valves, conduction tissue. We have showed that ADO-II devices (infact they are off-label use) can be used safely, effectively in such defects. Up to our knowledge this is the only study includes largest number of pediatric patients whose VSD were closed by ADO-II.

P-198

Trans-hepatic implant of a trimmed Melody® valved stent in tricuspid position in a 1 year old infant: first in man.

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Introduction: The trans-hepatic approach provides safe alternative route for cardiac interventions in children with complex congenital heart disease. We describe a transhepatic implant of a custom trimmed valved stent (Melody) in a 1 year old infant. This is the first procedure of his kind in man.

Method and Results: Infant with prenatal constriction of arterial duct leading to massive tricuspid regurgitation. Neonatal two surgical plasty attempts failed and therefore a De Vega plasty of the tricuspid valve failed as did an aortic bioprosthetic valve was implanted on day 30 (19 mm Epic™ aortic valve, St Jude Inc.). At the age of 1 year again a massive tricuspid regurgitation (3–4/4) was present. A surgical replacement was deemed to be too aggressive and too soon. Therefore a new bioprosthetic valve was implanted percutaneously. A transhepatic access was used deal with the size mismatch of the 20 Fr delivery and the vessels of an infant weighing 9,7 kg. The Melody valve was surgically trimmed by cutting some zigzags to 21 mm length. Echo guided puncture of hepatic vein with a Chiba Echotip® Biopsy Needle (Cook®), a 0.014” wire was placed into the right atrium. An 8 Fr introducer sheath was placed into the IVC. A 4 Fr right coronary catheter with a 0.035” J-tip Terumo® wire was advanced into the right pulmonary artery. Exchange for a 0.035” Amplatzer Super Stiff wire. Exchange for a 20 Fr sheath (Edwards Lifescience®) with a downsized introducer. The trimmed Melody® valved stent was delivered on a 16 mm Tyshak® balloon with the support of 8 Fr Mullins sheath. The 20 Fr sheath was pulled back into the hepatic vein until the hepatic parenchyma was reached. Two vascular plugs: 8 and 10 mm Amplatzer Vascular Occluder were deployed in tandem position in the hepatic veins. The skin was closed with 1 Donati stitch.

Discussion: This is the first trans-hepatic implant of a tailored downsized Melody valve in an infant. The therapeutic spectrum for complex procedures has been enlarged.

P-199

Radiofrequency perforation of the pulmonary valve: an efficient low cost solution

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Introduction: radiofrequency perforation of the pulmonary valve requires special equipment; price and availability are important. We assessed the feasibility and safety of “standard cathlab tools” in off-label use.

Methods: A co-axial telescopic microcatheter system was prepared (off-label use for all items): a 0.014” PT2 coronary guidewire (€160; Boston Scientific, USA) was chosen because the radio-paque tip is straight and has proven electrical conductance; for insulation the wire was put into a Progreat micro catheter (€250; Terumo, BE; profile 2.7 F=0.025”; lumen 0.021”) with 2 mm protrusion at the distal end; this was delivered to the aortic pulmonary valve through a standard prebended 4 F Ri Judkins coronary catheter (lumen 0.035”); gentle forward “push” was maintained on the RF wire complex while burning; radiofrequency RF energy was delivered with a standard surgical electrocauter system (Erbe ICC 80, Tubingen, DE; 5–10 W for 3 sec cutting mode).

Results: In vitro testing in a submerged sheep heart demonstrated perforation of semilunar valves in cutting mode at 5 W. Perforation of the pulmonary valve was performed in 5 patients (median age 3 days, weight 2.6 ± 0.3 kg); antegrade 3, retrograde 2; 10–15 W for 2 – 5 sec. After perforation of the valve, the microcatheter was easily pushed across the valve allowing to obtain a good position for the 0.014” wire (through duct into descending aorta or RA) for the ensuing balloon dilation and/or stent placement. In 1 patient a perforation to the pericardium was made; this was sealed by retracting the PT2 wire while applying RF coagulation mode.

Conclusions: The microcatheter telescopic system PT2-Progreat is a low-cost valuable tool in order to deliver radiofrequency energy in selected targets.

P-200

Complications after transcatheter ASD closure with the Amplatzer Septal Occluder

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The aim of this study is to report complications of transcatheter ASD closure with the Amplatzer Septal Occluder (ASO) (St Jude Medical). From November 1997 to December 2014, 792 patients underwent ASD closure with the ASO. Closure was mostly performed under general anaesthesia and transoesophageal echocardiography control. Choice of the device diameter was established after balloon sizing and measurement of the stretched diameter.

Mean age of the patients was 31.4 ± 22 years (0.5 month – 84 years). The stretched diameter was 22.4 ± 6.6 mm (4 – 43 mm) and device dimension 21.8 ± 6.7 mm (4 – 40 mm). Duration of the procedure was 41 ± 15 minutes and fluoroscopic time 7.39 ± 6.57 minutes (median 5.9 minutes). Dose of radiation was 18.1 ± 22 Gy.cm2 (median 11 Gy.cm2).

Implantation succeeded in 92.4% of pts and failure was mainly related to deficient rim. No device related death was noticed. Embolization occurred in 4 pts (0.5%): 1 in the aorta, 1 in the left ventricle, and 2 in the pulmonary artery. All but one underwent surgical extraction and ASD closure. The patient with aortic embolization had percutaneous device extraction and underwent subsequently successful implantation with a larger device. No patient required blood transfusion for any groin hematoma. One patient without aortic rim had hemopericardium one month after implantation; this was corrected by drainage with no recurrence and ASD full occlusion was noticed on colour Doppler control. No late complication was observed. The rate of full occlusion on Doppler control is more than 90%, and the remainings have trivial shunt.

Transcatheter ASD occlusion with the Amplatzer Septal Occluder is a safe and effective procedure. The rate of immediate complication is very low and need for immediate surgery following the implantation is rare (<1%). No device related late complications were reported up to 17 years after implantation. The risk of aortic perforation in absence of anterior rim (observed in about 20% of pts) is trivial and not a real limitation in clinical practice.

P-201

Outcomes following percutaneous intervention to obstructed right ventricle to pulmonary artery shunts after the modified Norwood procedure

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Introduction: Stenosis of the right ventricle to pulmonary artery (RV-PA) shunt following modified Norwood operation leads to early oxygen desaturation and may contribute to interstage morbidity and mortality. We sought to evaluate the efficacy of percutaneous interventions for severely desaturated patients with RV-PA shunt stenosis. **Material and Methods:** Between 01/2006 and 06/2014, 22 patients (17 with true HLHS) underwent 24 catheterizations for dilation of obstructed RV-PA conduit at a median interval of 163 days (range 3–434) after the Norwood operation. Median age and weight was

157 days (range 19–450) and 5.4 kg (2.8–10.1) respectively. Four patients required intervention early after the Norwood operation, and 2 patients required dilation of an interstage replacement RV-PA conduit. Eighteen patients had discreet obstruction: 11 at the proximal end, one in the mid portion and 6 at the distal end. The remaining 4 patients had multilevel stenosis.

Results: Balloon dilatation was performed in 17 patients, followed by stent implantation in 3 patients. Two patients had isolated stent implantation. Minimum shunt diameter of the stenosis and arterial oxygen saturations increased from 2.4 ± 0.5 mm to 4.3 ± 0.6 mm and $58.7 \pm 15.9\%$ to $79.4 \pm 9.9\%$, respectively. The mean diameter of the right and left pulmonary artery and pulmonary index was 5.75 ± 1.7 mm, 5.6 ± 1.8 mm and 185.7 ± 94.4 , respectively. Nine patients required additional interventions including pulmonary artery balloon dilation (5), aortic arch balloon dilation (2), pulmonary artery (1) or atrial septal (1) stent implantation. There were 4 in-hospital deaths, with a mean hospital stay for the remainder of 21.8 ± 14.6 days. Two patients died after discharge before the second stage of palliation, and 2 patients had RV-PA shunt exchange after interventional dilation due to inadequate pulmonary artery growth. Of the remaining 16 patients, 12 have undergone bidirectional Glenn.

Conclusions: Percutaneous treatment of a stenotic RV-PA shunt is effective and may facilitate a reduction in shunt-related surgical interventions. Isolated balloon angioplasty is often sufficient, however selected patients may require stent implantation to maintain conduit patency.

P-202

Percutaneous interventions for pulmonary artery obstructions in patients with HLHS after bidirectional Glenn operation

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Introduction: Pulmonary artery (PA) obstructions are one of the commonest sequelae after surgical treatment of hypoplastic left heart syndrome (HLHS). After Norwood operation, the small access to PAs through the systemic-to-pulmonary artery shunt limits treatment options. After the second stage of palliation, percutaneous interventions play a key role in management of stenotic PAs. We sought to evaluate our fifteen years of experience with interventions for PA stenosis in patients with HLHS after bidirectional Glenn (BDG) operation.

Methods: We retrospectively reviewed our database to select patients with HLHS after BDG operation, which required interventions for PA obstructions. Immediate results of balloon dilation (BD) and/or stent implantation, complications including stent fractures and follow-up reinterventions were evaluated.

Results: Between 09/2000 and 10/2014 one hundred two patients underwent interventions for PA obstructions at a median interval of 23.4 months (0.03–68.5 months) after BDG operation. Patients' median age and weight were 32.2 months (5.4–85.4 months) and 12 kg (4.1–23 kg) respectively. Fifty-five patients presented with isolated left PA stenosis, 4 patients had right PA stenosis and 43 presented with bilateral obstruction. Thirty-six patients (35%) had systemic-to-pulmonary artery shunt patent at the time of the intervention. Isolated BD was performed in 21 patients, a further 18 patients had subsequent stent implantation and 63 patients had

stent implanted as the first treatment. Altogether 81 patients received 85 stents and in 42 patients (52%) the stent was immediately post-dilated. Mean diameter of the stenosis increased from 3.9 ± 1.4 to 7.9 ± 1.9 mm (P.0001). Mean PA pressure proximal to the stenosis and mean gradient decreased from 16.7 ± 3.8 to 15.6 ± 3.5 mmHg (P.061) and 1.6 ± 1.5 to 0.2 ± 0.4 mmHg (P.0001) respectively. Fifty-five patients (54%) underwent 84 follow-up catheterizations at a median interval of 13.9 months (0.03–66.7 months). Twenty-six additional stents were implanted, 56 previously placed stents were redilated and in 2 patients isolated BD was performed. Seventeen stents fractures (20%) were noticed, 8 stents were stabilized with BD, the remaining 9 required further stent implantation.

Conclusions: Percutaneous interventions resulted in improvement in PA diameters and pressures. Isolated BD produce satisfactory results only in selected patients, the majority requiring stent implantation. Apart from stent redilation, stent fractures were a frequent indication for reintervention.

P-203

Pulmonary perfusion assessment in HLHS patients after Glenn operation and left pulmonary artery stenosis treated with stent implantation - preliminary study

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Introduction: Left pulmonary artery (LPA) stenosis is a common complication in patients with hypoplastic left heart syndrome (HLHS) after Norwood operation. LPA stenting is a well-established treatment in those patients.

Aim: Assessment of pulmonary perfusion in patients with HLHS after bidirectional Glenn (BDG) operation and pulmonary artery stenting using planar lung perfusion scintigraphy.

Material and Methods: Planar pulmonary perfusion scintigraphy was performed in ten patients (6 boys and 4 girls) at the median age of 56 months (21–104 months), after BDG operation. Mean Nakata index was 239 (118–341). Eight patients underwent stent implantation of which 7 had LPA stenting and the remaining one had RPA treated. Mean time from the stent implantation to scintigraphy was 19 months (3 days - 7 years).

Results: Pulmonary perfusion asymmetry was noted in 9/10 (90%) patients. Hypoperfusion of the left lung was noted in 7 cases. Contribution of the left lung to the global lung perfusion ranged from 11 to 59% (mean 28%). Hypoperfusion of the left lung was diagnosed in two patients (age 21 months and 30 months) with no previous pulmonary artery interventions. After LPA stenting both patients presented with improved left lung perfusion; from 18 to 37% and from 28 to 34%, respectively.

In one patient (8 years old) after previous LPA stent implantation, hypoperfusion of the left lung (25%) was noticed, which was related to restenosis due to stent fracture. Despite successful stent implantation with significant improvement in the diameter of the obstruction from 6.25 mm to 12.5 mm, there was no improvement in the follow-up scintigraphy performed 3 days after the intervention.

Conclusions:

1. Patients with HLHS present with pulmonary perfusion abnormalities due to non-physiological pulmonary flow and pulmonary artery obstructions.

2. Hypoperfusion of the left lung is the most common finding even after successful LPA stenting.
3. Pulmonary artery stent implantation in young patients shortly after BDG operation brings the biggest improvement in pulmonary perfusion.
4. Planar lung perfusion scintigraphy may be a useful method for outcomes assessment of percutaneous interventions for pulmonary stenosis in HLHS.

P-204

Transcatheter Melody® valve implantation in pulmonary position. Expanding anatomical indications

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Transcatheter pulmonary Melody® valve implantation: immediate results and mid term follow in diverse RVOT anatomies

Background: Transcatheter pulmonary Melody® valve implantation was originally designed to treat surgical RV-PA conduits. We describe our experience in Melody implantation in different Right Ventricular Outflow tract (RVOT) anatomies,

Methods: retrospective review of clinical records of the 17 patients who underwent percutaneous Melody valve implantation in our center between February 2012 and October 2014. Indications, RVOT anatomies, technique, immediate and mid term results were evaluated.

Results: The *indication* was combined stenosis and regurgitation (n = 13), severe pulmonary regurgitation (n = 3), severe stenosis (n = 1). The *anatomy of the RVOT* was previous surgical conduit (n = 6), native outflow tract (n = 6), pulmonary biological prosthesis (n = 3) and single right pulmonary artery (n = 2). Venous access used for the Melody implantation was femoral (n = 13) and jugular (n = 4). Pre-stenting was performed in 16 patients.

Immediate results: successful implantation was achieved in all 17 cases. The peak hemodynamic gradient fell from 28.3 ± 11.8 to 10.88 ± 6.2 mmHg and the RV/Ao pressure ratio from 0.57 ± 0.11 to 0.41 ± 0.10 , and only one patient had and mild residual periprosthetic regurgitation. One patient developed a small aneurysm in the conduit distal to the prosthesis, that remained stable during follow up. **Follow up.** In a mean follow-up of 1.15 ± 0.74 years (maximum 2.4 years), the valves remained competent and all patients maintained the improvement in their functional class. One patient developed moderate gradient needing balloon redilatation of the Melody 4 years after the implantation. One patient had bacterial endocarditis 6 months after the implantation, resolved with antibiotic treatment.

Conclusions: Percutaneous Melody® valve implantation is an alternative to surgery not only for dysfunctional surgical conduits, but also for other anatomic types of RVOT, with good immediate results and in the medium term follow up.

P-205

Catheterization in the Early Postoperative Period Following Pediatric Cardiac Surgery: Security and Efficacy

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Introduction: Cardiac catheterization in postoperative critical care unit period after cardiac surgery is generally perceived as high risk

and often debated; to date there is little published data regarding this setting.

The aim of this study is to examine the indications, safety and efficacy of catheterization performed early after congenital heart surgery.

Methods: All catheterizations performed within six weeks after surgery between January 2011 and December 2014 were retrospectively reviewed. Morphological, surgical and catheterization data, including mortality and reintervention were analyzed.

Results: 75 patients, median age 5 months (0–169), median weight 6 kg (1.5–65) underwent 83 catheterizations on median post-operative day 8. Procedures were either interventional (n = 63) or non-interventional (n = 20). Primary diagnoses were heterogeneous, but the majority had complex intracardiac anomalies, and 43.4% had functional univentricular physiology.

Main indications for cardiac catheterization included: low cardiac output (51%), residual lesions by echo (25%) and persistent hypoxemia (13%). Twenty-seven children required extracorporeal cardiopulmonary support. Intervention procedures included: stent implantation (n = 41), angioplasty (n = 13) and vascular/shunt occlusion (n = 11). Most of these interventions (67%) involved a recently created suture line. Ten catheterizations were associated with complications (one acute renal failure, two stent migrations, four arrhythmias and two superior caval vein perforations). There were no complications related to patient transport, and there was no procedural mortality. 31% of patients died during ICU postoperative period with an hospital discharge survival of 60%. Non-interventional catheterization (p = 0,012), and extracorporeal cardiopulmonary support (p = 0,025) were risk factors for death.

Conclusions: In our experience, transcatheter interventions can be successfully performed in the early postoperative period. Catheterism also allows to find and treat undiagnosed residual lesions, which may have a positive impact on patients outcome. These procedures should be always performed supported by a multidisciplinary team.

P-206

Results of a systematic transcatheter closure of secundum atrial septal defects with the Amplatzer Septal Occluder: a ten year single paediatric center study

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Introduction and Methods: Percutaneous closure has become the treatment of choice of Ostium Secundum Atrial Septal Defects (ASD) but follows variable selection criteria from one group to another. In August 2000, our group adopted the policy to systematically attempt percutaneous closure of all significant isolated ASDs presenting to our institution, irrespective of the anatomy of the defect (size and presence of sufficient rims) and size of the patient. This study evaluates retrospectively the safety and efficacy of this policy during the first ten years of its application, using the Amplatzer Septal Occluder (ASO) in a consecutive paediatric population from a single institution.

Results: A total of 322 patients were evaluated for ASD treatment at a mean age of 6.2 ± 4.2 years (range 0.3 - 18) and a mean weight of 23.5 ± 16.8 kg (range 5–135). Only one family elected surgical ASD closure upon advice from its treating cardiologist. Percutaneous ASD closure was systematically attempted and was successful in the remaining 321 patients. Mean procedure time was 121 ± 35 minutes (range 40 - 240) and mean fluoroscopy time 21.1 ± 9.8 minutes (range 6 - 66). Mean device size was 19 ± 6 mm (range 6 - 38). A small residual shunt was noted in 39 patients (12%) at 24 hours, and in 11 (3.4%) at final follow up of 33 ± 25 months. Minor transitory

complications occurred in 46 patients (14.3%), but no deaths or permanent sequelae from complications were related to the procedure, and no device had to be explanted.

Conclusion: In our experience, all anatomical variations of Ostium Secundum ASD can be effectively and safely treated with the ASO in paediatric patients. This report supports our policy of systematically attempting percutaneous closure in all patients, leaving failed procedures the only clear indication for surgical treatment.

P-207

Indication for percutaneous transluminal pulmonary angioplasty is predicted by oxygenation index and pressure of vena cava superior on super-acute stage after Glenn

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Background: Stenosis of pulmonary artery (PA), which has to be mended by percutaneous transluminal pulmonary angioplasty (PTPA), frequently occurs in early infants on acute stage after Glenn, particularly after Glenn with Norwood following bilateral PA banding. We investigated whether PTPA was predicted by pulmonary circulation factors on super-acute stage in patients after Glenn.

Methods: The medical records of 54 early infants after Glenn were reviewed. Arterial carbon dioxide pressure (PaCO₂) and arterial oxygen pressure (PaO₂) on respirator was measured next morning after Glenn. Alveolar-arterial oxygen difference (AaDO₂) was calculated with fraction of inspiratory oxygen (FiO₂). If patient was free from respirator next morning, these three values were adopted ones just before ventilator weaning. Three indexes were employed as follows: AaDO₂/PaO₂ (RI), PaO₂/FiO₂ (PFR), and pressure of vena cava superior (SVCp). The area under a receiving operating characteristics (ROC) curve was calculated to determine the best discriminating each factor for predicting PTPA. Next, positive predictive value (PPV) and negative predictive value (NPV) were calculated for predicting PTPA.

Results: Twenty-three infants underwent PTPA between 1 to 50 days (median 8 days) after Glenn; 16 of them had got through Norwood-Glenn procedure. The area under ROC curve of RI, PFR, and SVCp for predicting PTPA were 0.85 (95% confidence interval: 0.74-0.94), 0.84 (95% CI: 0.74-0.94), and 0.78 (95% CI: 0.66-0.90) respectively. For predicting PTPA PPV of RI (>15) was 73%; PFR (≤ 72) 71%; SVCp (≥ 18 mmHg) 72%. If patients did not meet every one of three criteria as above, 87% of them got away from PTPA (NPV 87%). If we employ criteria as below, the value of NPV for predicting PTPA becomes very high respectively: NPV of RI (<10) was 94%; PFR (≥ 70) 100%; SVCp (≤ 13 mmHg) 100%.

Conclusion: Our study showed RI, PFR, and SVCp on super-acute stage after Glenn were good indexes for predicting PTPA. Stenosis of PA would be formed just after the operation is completed. Measurement of blood gases and SVCp can be repeated on super-acute stage after Glenn. These pulmonary circulation factors might be useful for screening of PA stenosis on super-acute stage after Glenn.

P-208

Selective propensity of bovine jugular vein material to bacterial adhesion and impact of percutaneous pulmonary valve implantation procedural steps in the genesis of infective endocarditis: an in-vitro study

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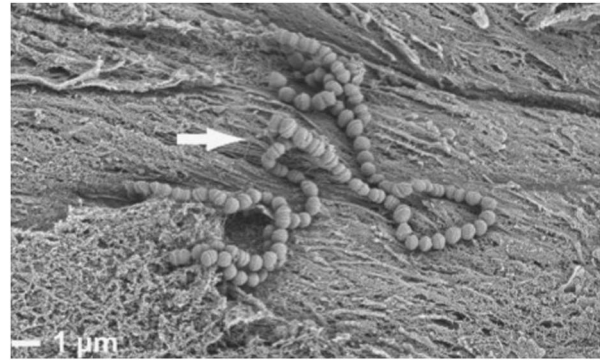


Figure 1.

Introduction: Percutaneous pulmonary valve implantation (PPVI) using bovine jugular vein made Melody valve is safe and effective. However, infective endocarditis have been reported for unclear reasons. We sought to assess the impact of PPVI procedural steps on valvular histology, selective bacterial adhesion and leaflet mechanical behaviour.

Methods: Three valved stents (Melody valve, homemade stents with bovine and porcine pericardium) were tested in-vitro in 4 conditions: I) control group, II) crimping, III) crimping + inflation of low-pressure balloon and IV) condition III + post dilatation (high-pressure balloon). For each condition, valvular leaflets (and venous wall sample for Melody stents) were taken for histological analysis, bacterial adhesion using *Staphylococcus aureus* and *Streptococcus sanguinis* strains and mechanical uniaxial tests of valve leaflets.

Results: Among Melody valves, incidence of transverse fractures was significantly higher in traumatized samples compared with control group ($p < 0.05$) whereas, incidence and depth of transverse fractures were not statistically different between the 4 conditions for bovine and porcine pericardial leaflets. Bacterial adhesion was higher on bovine jugular venous wall for *S. aureus* and on Melody valvular leaflets for *S. sanguinis* in control groups and significantly increased in traumatized Melody valvular leaflets with both bacteria (I vs IV: $p = 0.05$). Figure 1 shows scanning electron microscopy evidence bacterial adhesion of *S. sanguinis* on Melody valve leaflet (white arrow). Bacterial adhesion was lower on bovine pericardial leaflets.

Conclusions: Valved stent implantation procedural steps induce histological lesions on Melody valve leaflets. Adhesion of *S. aureus* and *S. sanguinis* pathogenic strains to Melody valve components was noted on safe tissue and procedural steps of implantation increased it.

P-209

Percutaneous duct occlusion in very low weight children: a safe and feasible alternative to surgical approach. Follow-up in 12 cases

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Background: Pre-terms neonates and newborns with severe co-morbidities are nowadays a great challenge in neonatal ICU. From a cardiovascular point of view, patent ductus arteriosus (PDA) represents the most frequent cause of heart failure. Treatment options includes: medical therapy and/or surgical intervention. Surgery is safe but, involving a thoracotomy, carries morbidity and mortality. The risk rises considerably in those

neonates with additional comorbidities. Catheter closure option can potentially overcome several of these limitations. Thanks to new materials with smaller profile, the great limit of vascular access caliber is overcome.

We present our experience and follow-up in weight babies (≤ 3 Kg). **Materials and method:** From March 2012 to November 2014 we treated 12 newborns, weight ≤ 3 Kg (mean 2,56 Kg) with percutaneous approach. All babies presented with relevant PDA and important co-morbidities: 8/12 preterms; 4/12 with severe bronchodysplasia; 1/12 was previously operated of aneurysm of Galeno vein; 1/12 with congenital Rubella; 1/12 with diaphragmatic hernia; 3/12 had other congenital cardiac defects associated (ASD and VSD); 1/12 Trisomy 18 and another one affected by a plurimorfomation syndrome. In 8/12 patients medical therapy with indometacine was attempted: in four there was evidence of NSAID therapy complication, one of them had retinal hemorrhage. All patients but one received furosemide, three were in ACE-I therapy and two children received sildenafil for bronchodysplasia and pulmonary hypertension.

Results: We register no mortalities nor morbidities due to catheterization.

We used Amplatzer Duct Occluder II Additional Size in 9/12. In three cases we used ADO II.

Mean fluoroscopy time was 15 minutes and mean contrast volume was 15 ml.

No residual shunt at the ultrasound was found.

Along follow-up there were 0 death. The heart failure therapy was stopped in all patients after procedure.

Conclusions: Thanks to materials improvement percutaneous occlusion of PDA in very low weight children is now feasible and safe alternative to the surgical gold standard.

P-210

Goose neck snare assisted percutaneous ASD closure: a safety procedure for very large and complex ASDs

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Background: ASD transcatheter closure is a widespread procedure. However, in some cases ASDs may be large and with soft rims. Furthermore, the tension between the delivery system and the septum can be associated to a significant change in position of the device at release. A potential risk exists for device malposition/embolization.

Methods and Patients: When TEE evaluation and balloon sizing showed large defects with floppy rims the chosen Amplatzer device was implanted in a standard way. Before release a 5 mm goose-neck snare with its 4 Fr catheter were placed across the delivery cable and advanced in parallel up to the screwing mechanism. The goose-neck snare was placed across it and fixed in order to catch the screwing mechanism.

Then the delivery cable was unscrewed and the device reached its final position without any tension. The goose neck snare is very soft and allows a tension-free position of the device. If needed the device can be retrieved and/or replaced or repositioned.

If the position was considered satisfactory the device was released from the goose-neck snare by opening the loop of the snare.

Between January 2000 and October 2014, 14 patients had a snare assisted ASD transcatheter closure (10 females, median age 22 years (range 10-62 years)).

Results: Median device size was 20 mm (range 16-36 mm).

In three patients, the device after release from the delivery cable changed significantly its position and a residual shunt due to

prolapse of the left atrial disc was seen. The right atrial disc was recaptured with the help of the goose neck snare system and the left atrial disc repositioned properly.

In two subjects with multiple ASDs, a second fenestration looked quite significant with the device still attached to the delivery cable. However, after release (with the device still attached to the snare system) the second fenestration looked significantly smaller at TEE and at Qp/Qs evaluation.

Procedure was successful. No complications occurred during the procedure and during follow-up

Conclusions: Snare-assisted Amplatzer ASD device placement is a new method for ASD percutaneous closure. It adds safety to the procedure and it proved to be safe.

P-211

Atrial Septal Defect Morphology and Stenting in Hypoplastic Left Heart Syndrome after Hybrid Palliation

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Introduction: Five morphologies of atrial septal defect (ASD) have been described in hypoplastic left heart syndrome (HLHS). The most common is the ostium secundum ASD. Restrictive physiology occurs when ASD is small (either secundum-type or primum-type ASD) or sometimes in case of malaligned ASD, due to the small left atrial size.

Methods: Between October 2011 and November 2014 forty-three patients (25 males; mean weight 3 Kg, from 1,9 to 4,2 Kg) with HLHS underwent a hybrid procedure in our institution. Mean age of the patients at the time of operation was 3,6 days (1 to 7). Restrictive physiology of ASD occurred in 10 patients (23%). Diagnosis of restrictive ASD was based on continuous Doppler through the ASD recording a gradient higher than 7 mmHg and clinical signs of overloaded pulmonary flow.

Results: Only one patient showed malaligned ASD, while the other patients had ostium secundum ASD. Three of them presented with restrictive physiology shortly after birth and underwent transcatheter balloon atrioseptectomy along with the hybrid procedure. Three patients needed balloon dilatation of

Table 1.

ASD MORPHOLOGY	ANATOMY	INTERVENTION	TIMING (days from birth)	TRANS-ATRIAL PRESSURE GRADIENT mmHg	BALLOON/STENT TYPE
Critically small	MS/AA	Rashkind + Surgery	7/8	12 → 10	
Critically small	MS/AA	Rashkind + Stent	4/59	17 → 5/12-2	Rashkind/Genesis 10/19
Critically small	Shone Complex	Rashkind	6	---	Rashkind
OS ASD	MA/AA	Balloon	107	8 → 5	Advance 8x20
OS ASD	MA/AA	Stent	52	23 → 1	Genesis 25/10
OS ASD	MA/AA	Stent	57	17 → 1	Genesis 19/10
OS ASD	MA/AA	Stent	52	32 → 5	Genesis 19-10
OS ASD	MS/AS	Balloon + Stent	20/96	3 → 1/23 → 2	Tyshak II 8/20 / Genesis 10/19
OS ASD	MS/AS	Stent	80	20 → 5	Genesis 10/19
OS ASD	MS/AS	Surgery	86	---	None
LDSP	MS/AS	Balloon + Stent + Surgery	48/53	22 → 20/22 → 1	Fox 8/20/ Genesis 10/19

the ASD. Of these, two required ASD stenting. A total of 7 patients received transcatheter ASD stenting. Only two patients underwent surgical atrioseptostomy. In the other case surgery was performed right after migration of the stent in the right atrium due to the malalignment of the septum primum that made the stent unsupported by the antero-superior septal tissue. Characteristics of patients with restrictive ASD are reported in table 1. In our experience, a crucial aspect of transcatheter ASD stenting concerns the technique of deployment of the stent: we prefer to first withdraw and inflate the distal half of the stent on the left side of ASD, then withdraw the long sheath and complete the stent deployment on the right side of the ASD, obtaining more stability of the stent.

Conclusions: ASD stenting is commonly required in HLHS palliation and is nearly always feasible for the ostium secundum ASD. Conventional transcatheter approach may not be effective in the presence of different ASD morphologies.

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Esophageal perforation due to an Amplatzer Vascular plug IV

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The Amplatzer vascular plug IV (AVP IV) is a self-expandable, replaceable occluder made of Nitinol wire mesh, which allows a safe and effective interventional occlusion of medium size vessels.

We report about an infant diagnosed with PA-VSD and multifocal collateral lung perfusion through 4 MAPCAs. A central aorto-pulmonary shunt was performed at the age of 4 months. Because of postoperative pulmonary hyperperfusion one of the MAPCAs was closed interventionally using a 5 mm AVP IV. This MAPCA originated from the descending aorta (DAO) near the 5th thoracic vertebra and coursed behind the esophagus to the right lower lung lobe. The MAPCA was closed near its origin from the DAO. Four weeks later the patient presented an episode of severe gastrointestinal bleeding, caused by perforation of the AVP IV into the esophagus. The occluder was extracted surgically, the MAPCA was clipped and the esophageal injury was oversewn. A broad antibiotic therapy was successful to prevent severe mediastinitis and the esophageal perforation healed without complication. Later on the patient underwent unifocalisation and biventricular repair.

Until now there are no reports describing esophageal perforation due to an AVP IV. In our case the perforation was favored by the fact, that the AVP IV was implanted near the aorta in a MAPCA segment that was located directly in front of the spine and behind the esophagus. Another possible factor in our patient was the chronic requirement of a gastrointestinal feeding tube. Although the occluder is soft and flexible, the spindle-shaped ends can become potentially traumatic if they are located in close relationship to other structures. If the implantation of the AVP IV is considered in vessels originating from the descending aorta, one should be aware of potentially dangerous anatomical relations of the target vessel with the spine and adjacent organs. Close spatial relations might favour late perforations due to erosion by the relatively stiff ends of the vascular plug.

P-213

Does Fontan circulation engender progressive liver dysfunction?

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Introduction: Total cavopulmonary connection (TCPC) forces systemic venous blood into the lungs, equalizing caval and pulmonary pressure. Chronic hepatic stasis generates a progressive liver dysfunction, eventually leading to cirrhosis. We investigated prospectively the hemodynamic changes and the liver status after TCPC.

Methods: From March 2013 to December 2014, 64 TCPC patients (pts) underwent cardiac catheterization and liver examination (blood tests, ultrasound and gastroscopy) at our center.

Results: Median age was 10 (5-32) yrs, median distance from TCPC was 10 (1-19) yrs. Catheterization showed the following data: pulmonary arterial pressure (PAP) $11,6 \pm 2$ mmHg (>15 mmHg in 10 pts), ventricular end-diastolic pressure (VEDP) $6,67 \pm 2,58$ mmHg, pulmonary vascular resistances (PVR) 2 ± 1 WUm2 (>2 in 18 pts); cardiac index (QSI) $3,15 \pm 1,27$ ml/min/m²; systemic O₂ saturation $94 \pm 4\%$ ($<95\%$ in 18 pts), QP/QS $0,9 \pm 0,2$.

37 interventions were performed in 27 patients. We found the following significant correlations: interval from TCPC vs QSI ($-0,30$, $p < 0,001$), interval from TCPC vs VEDP ($0,3$, $p < 0,01$), PVR vs QSI ($-0,81$, $p < 0,001$). Nor PAP, PVR or QP/QS were significantly related with age or interval from TCPC. Trans-hepatic gradient was $2,33 + 1,10$ mmHg; 20 pts had major venous collateral vessels from the liver.

Gastroscopy showed oesophageal varices in 6 pts (0,09%) having PVR >2 WUm². Conversely, all pts with PAP >15 mmHg had either venous collaterals or oesophageal varices.

Liver function was normal in all pts. Hepatomegaly was found in 23 pts; the liver was nodular and/or inhomogeneous in 10 and 35 pts. Stiffness was $16,63 + 5,96$ KPa and significantly related to time from TCPC ($r:0,33$, $p < 0,01$).

A subgroup of patients showed a negative trend very early after TCPC.

Conclusions: This is the largest prospective series showing that TCPC engenders a progressive decrease of QSI and increase in VEDP, with a tendency to PAP and PVR to raise. This is balanced by the progressive development of venous collateral vessels, from both caval and hepatic systems. The hepatic stiffness increases with time, but cirrhosis and esophageal varices are found in few patients. Only a multidisciplinary approach will be able to identify patients at risk.

P-214

Early experience with Valeo Balloon Expandable Vascular Stents in treatment of aortic recoarctation in small children

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Background: The Valeo stent as a low profile, open cell balloon expandable stent that can be postdilated to 20 mm diameter seems to be an alternative for treatment of growing children with

different vascular narrowings, but its low radial force can limit its use to compliant lesions.

Objectives: To describe the single center experience and outcome from Valeo Vascular Stents (Bard) implantation in small children with postoperative coarctation of the aorta (rCoA).

Methods: Retrospective data collection was analyzed. Primary endpoints were peak systolic catheter gradient reduction, stented segment diameter increase. Early follow-up results were recorded.

Material: Between 2013 and 2014, 12 pts with rCoA (rCoA/dAo diameter $0,43 \pm 0,17$, including 6 pts after Norwood- Sano operation for HLHS underwent Valeo stent implantation following the unsatisfactory result of balloon angioplasty due to elastic recoil. Median patients age was 5,5 mths (2-120). Arterial (6 pts) or venous (6 pts) approach using short 6-7 F sheath were performed. The stent diameter was equal to diameter of aorta on the level of diaphragm and length dependent on the morphology of stenosis. In one patient immediate stent redilation to 12 mm was performed to opposite the stent to the wall of the aorta.

Results: All implantations were successful with no complications. There was significant improvement ($p < 0.001$) in pre versus post stenting aortic diameters ($3,5 \pm 1$ mm (1-4,6, med.3,35) vs $7,3 \pm 1,9$ mm (6-12, med.6,6) and systolic gradient ($31,83 \pm 13$ mmHg (12-50, med.25) vs $3,3 \pm 4,9$ mmHg (0-10, med.0). Successful reduction in the post stenting gradient was achieved in 100% of primary procedures. During follow-up period $9,6 \pm 6,9$ mths (3-18, med.6) all stents were patent with no signs of restenosis (noninvasive pressure measurements and echo-doppler), no stents fractures (chest x-ray).

Conclusions: 1. Valeo Balloon Expandable Vascular Stents is useful in treatment of compliant recoarctation of the aorta in small children with good acute and early results. 2. Longer follow up is needed.

P-215

Single center early experience with Valeo Balloon Expandable Vascular Stents in treatment of pulmonary arteries stenosis in small children

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Objective: To describe single center experience with a low profile, open cell, balloon expandable Valeo Vascular Stents (Bard) implantation in small children with pulmonary arteries stenosis.

Methods: Retrospective data collection was analyzed. Primary endpoint was stented segment diameter increase (due to diverse group of univentricular and biventricular defects plus additional RVOTO in several patients, pressure gradient reduction was not chosen as an adequate endpoint). Early follow-up data based on echo-doppler were recorded.

Material: Between 2013 and 2014, 21 pts with pulmonary arteries stenosis (8 pts after Glenn/Fontan operation, 7 pts after Tetralogy of Fallot correction with pulmonary regurgitation, 2 pts after univentricular to biventricular conversion, 4 pts with complex congenital heart defects after various surgical procedures) after unsuccessful balloon angioplasty due to elastic recoil or with tubular stenosis had 23 Valeo stents implanted through 6-7 F short sheath. The femoral vein approach or jugular approach in post Glenn patients were performed. The stent diameter was equal to the diameter of normal segment of the pulmonary artery and length 18 or 26 mm dependent on the length of stenosis. Median patients age was 5 yrs (0,6-16).

Results: All stents were implanted with no major complication. There was significant improvement ($p < 0.001$) in pre versus post stent pulmonary artery diameter ($3,95 \pm 1,32$ mm (2-6, med.3,7) vs $7,5 \pm 1,5$ mm (4-10, med.7,4). Successful dilatation was achieved in 100% of primary procedures. Acute minor complication with stent dislocation stabilized with the 12 mm diameter Tyshak valvuloplasty catheter which occurred in one patient. During follow-up period $9,5 \pm 5,8$ mths (2-28, med.6) all stents are patent with no signs of restenosis – judgment on the base of echo-doppler examination, with no signs of stent fractures or collapse in chest x-ray.

Conclusions: 1. Valeo Balloon Expandable Vascular Stents are useful in treatment of compliant pulmonary arteries stenosis in young children, with good acute and early results. 2. Longer follow up is needed to evaluate their role in the final treatment of pulmonary arteries stenosis.

P-216

Transcatheter closure of ruptured sinus of Valsalva aneurysm with new types of nitinol wire mesh PDA occluders – short and midterm results

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Introduction: Ruptured sinus of Valsalva aneurysm (RSOVA) is a rare shunt lesion frequently treated percutaneously. Lately for this purposes have been used also Chinese PDA nitinol wire mesh devices very similar to Amplatzer Duct Occluder type I (ADO). Experience with this occluders is scant.

Aim: To present results of transcatheter closure of RSOVA with Chinese PDA occluders taking in consideration short and midterm results.

Methods: From September 2010 to August 2014, 8 patients (pts) from 17 to 72 years old (mean age 40 y) have closed their RSOVA with nitinol wire mesh PDA occluders (produced by 3 different Chinese companies). All but two pts had congenital sinus of Valsalva aneurysm. Two pts had acquired RSOVA after previous cardiac surgery (one after aortic valve replacement, another after surgery of tight subaortic stenosis – LVOTO). In all pts arteriovenous loop was created and PDA devices were implanted transvenously. There were used devices 2-6 mm bigger than orifice of RSOVA. There were 7 connection between right coronary or noncoronary sinus with right atrium and 1 between noncoronary sinus and right ventricle.

Results: All PDA devices (sizes from 12/10 to 18/16) were successfully implanted in RSOVA. In one pt with iatrogenic RSOVA (after LVOT operation) device have been retrieved because of massive aortic regurgitation after implantation provoked by the device. In 72 y old woman, after aortic valve replacement, Chinese duct occluder was applied in proximal entrance to the RSOVA. Because of the presence of important residual leak on the edge of the implant the procedure had to be supplemented by closing of the distal RV orifice of RSOVA with 10 mm Muscular VSD Occluder. In one pt after embolization of ADO to pulmonary artery and its transcatheter retrieval, bigger device (Chinese one) were applied. In another pt after ADO implantation 2 y later (during pregnancy) recanalization of SOVA occurred treated successfully by Chinese PDA occluder after delivery. In follow-up (ranged from 0,5 till 4 years) no complications were observed in any pt.

Conclusions: Transcatheter closure of ruptured sinus of Valsalva aneurysm with new PDA nitinol wire mesh occluders are safe and effective procedures.

P-217**The evaluation of nosocomial infection prevalence and risk factors in pediatric patients with extracorporeal membrane oxygenation support**

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Objectives: Extracorporeal membrane oxygenation (ECMO) to provide temporary respiratory and cardiovascular support in intensive care units has become a standard technique over the past few decades. We undertook a review of the data of patients who received ECMO support to determine the risk factors and causal organisms related to acquired infections in pediatric ECMO patients.

Methods: Sixty-six patients who received ECMO support in pediatric cardiac intensive care unit at Mehmet Akif Ersoy Thoracic and Cardiovascular Surgery Center in years between January 2011– June 2014 were included in this retrospective study. Demographic, echocardiographic, hemodynamic features and surgical procedures were reviewed.

Results: A total of 66 patients received a total of 292.5 days of VA-ECMO support were revealed in pediatric ICU. Sixty of them were postoperative patients, 6 patients were internalized by different indications. 45 patients weaned from ECMO support with an ECMO survival of 68.2%. There were 13 (37.2%) blood stream infections (BSI), 10 (29.4%) respiratory tract infections (RTI), 9 (25.7%) urinary tract infections (UTI) and 2 surgical site infection (5.7%). The rate of infection was 116.2/1000 ecmo days. Gram negative bacteria accounted for 44.1% (15/34), gram positive bacteria 26.5% (9/34), Candida 29.4% (10/34) of the total culture positive pathogens. When infected and non infected patient groups were compared; prolonged ICU stay, duration of ventilation and the duration of ECMO were associated with the development of nosocomial infection in patients received ECMO support ($p=0.045$, $p=0.030$, $p=0.040$ respectively). Multivariate logistic regression analysis revealed only the duration of ECMO as an independent risk factor for nosocomial infections in patients received ECMO support (0 R:1.318 (CI:1.066–1.63; $p=0.011$).

Conclusions: ECMO is a life saving modality in perioperative cardiac patients. The establishment of a prophylactic antibiotic regimen covering the most prevalent microorganisms in ICU and a standardized protocol for ECMO practice, with the more strict application of ECMO indications would lead an improvement in infection incidence along with hospital surveillance of the patient group.

P-218**Surgical Management Following Initial Palliation with Ductal Stent and/or Radiofrequency Pulmonary Valve Perforation/Pulmonary Balloon Valvuloplasty in Patients with Intact Ventricular Septum-Pulmonary Atresia/IVS-Critical Pulmonary Stenosis**

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Introduction: Our approach is to stent the patent ductus arteriosus and to perforate or balloon the pulmonary valve in patients with Intact Ventricular Septum (IVS) – Pulmonary Atresia (PA) or IVS – Critical Pulmonary Stenosis (CPS) as an initial palliative procedure. We investigated the surgical strategies as a second procedure following this palliation.

Methods: Medical records were reviewed in patients with IVS-PA or IVS-CPS who had undergone surgery as a second procedure at our institution between 2010 and 2014. All patients had mono or bipartite right ventricles and ductus dependent pulmonary circulation. The influence of ductal stenting, RF perforation or BD of pulmonary valve and balloon atrial septostomy (BAS) on the second stage surgical management and mortality was examined.

Results: Of 35 patients, 28 (80%) had IVS-PA and, 7 (20%) had IVS-CPS. Twenty-four (69%) patients had bipartite and 11 (31%) patients had monopartite right ventricles. All patients had ductal stent but one. RF perforation and BD were performed in 17 and 4 patients, respectively. BAS was done in 11 (%31) patients. Twenty-six (74%) patients survived following the initial palliation. Seven patients (27%) have been followed up without surgery. Two patients lost to follow-up and 17 (65%) patients (48% of all patients) underwent second stage surgical procedures. Single, one-and-a-half and 2 ventricle repairs have been performed in 7, 6 and 4 patients, respectively. Patients who had undergone one-and-a-half ventricle repair had 2 right ventricle outflow tract (RVOT) reconstructions and 2 pulmonary valvulotomies, Two-ventricle repair patients had 3 RVOT reconstructions and 1 pulmonary valvuloplasty. There was 1 early and 1 late death because of cardiac and non-cardiac reasons, respectively. All the patients with monopartite ventricles who had not undergone BAS at the initial procedure died.

Conclusions: Ductal stenting is effective for pulmonary artery development in patients with IVS-PA or IVS-CPS and provides a straightforward surgery without dealing with pericardial adhesions at the second stage. A sufficient RF perforation or BD of pulmonary valve is necessary for the future one-and-a-half or 2 ventricle repairs. It is important to perform BAS as a part of the initial procedure if the right ventricle is monopartite.

P-219**Evaluation of Tracheostomy patients in Pediatric Cardiac Intensive Care Unit: Five years experience from single center**

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Objective: Recently, rapid extubation after pediatric cardiac surgery is a preferred approach but not possible in some cases and prolonged endotracheal intubation may lead to local trauma, nosocomial infections and psychological problems which will increase morbidity and mortality rates. Tracheostomy is an alternative method to prevent these problems. Still, there is no consensus for timing of tracheostomy, after pediatric cardiac surgery. In this study we aimed to evaluate, pediatric patients undergoing cardiac surgery, who required tracheostomy.

Patients and Methods: All pediatric cardiac patients (under 18 years of age) who had cardiac surgery and required tracheostomy

between January 2010 and November 2014 were reviewed retrospectively. The time of tracheostomy, duration of mechanical ventilation and intensive care, clinical status, demographic, echocardiographic features and additional pathologies were recorded. **Results:** After cardiac surgery, 19 (1.3%) of 1450 patients with a median age of 19.3 ± 43.3 (range 1.8–192) months during surgery and weight of 8.5 ± 11 (3.8 to 51) kg required tracheostomy. Ten patients (53%) were female. Median duration between surgery and tracheostomy was 38.2 ± 13.3 (20–77) days. Mean extubation trial before tracheostomy was 4 ± 1 times. Genetic syndromes were present in 5 (26%) of the patients (3 Down syndromes, one Di George syndrome, one Danon disease). Eight (42%) patients had neurological sequela. No procedure related complication was occurred in any patient except a patient with surgical site bleeding who needed surgical revision. After the tracheostomy, 12/19 patients (63%) were weaned from ventilators and 3/19 patients are still in the hospital. Totally 9/19 patients were discharged from the hospital and 5/9 were successfully decannulated (1, 4, 6, 8 and 11 months later). 8 patients died (7 during hospital stay, 1 at home) in total.

Conclusion: The mortality of tracheostomy patients is still significant but this may be related to primary cardiac disease and underlying other clinical problems. On the other hand tracheostomy which is fairly a safe procedure may facilitate discharge from the intensive care unit.

P-220

Postoperative Chylothorax Following Paediatric Cardiac Surgery

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Introduction: Chylothorax is a rare condition that generally occurs after thoracic and cardiac procedures. The present study aims to determine the incidence, risk factors, and the impact of chylothorax on the early postoperative outcome after paediatric cardiovascular surgery.

Method: The hospital records of all paediatric patients with postoperative chylothorax following congenital heart surgery at our institution between June 2011 and June 2014 were reviewed retrospectively.

Results: There were 1123 cases operated during the study period. Of these, 35 cases (3.1%) were complicated by chylothorax in the postoperative period. Nineteen patients (54%) were males, while 16 (46%) were females. The most common surgery complicated by chylothorax was the single ventricle repair (11 cases, 31%). It was followed by aortic arch repairs (9 cases, 25%); Tetralogy of Fallot (4 cases 11%); atrioventricular septal defect repairs (4 cases, 11%); arterial switch (3 cases 8%), ventricular septal defect repairs (2 cases 6%) and others (2 cases, 6%). Nutritional management included low lipid diet, enteral feedings enriched with medium-chain triglycerides and parenteral nutrition. Fourteen patients were treated with octreotide, 4 with thoracic duct ligation, and 3 with pleurodesis. Sepsis was observed in nine cases and three patients died.

Conclusion: Chylothorax after paediatric cardiac surgery is not a rare complication. It occurs more commonly after single ventricle repair and has a significant impact on the postoperative clinical course and morbidity.

P-221

Late Primary Arterial Switch Operation in Patients with D-transposition and Intact Ventricular Septum

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Introduction: The primary arterial switch operation (p-ASO) has become the standard treatment choice for correction of transposition of the great arteries with intact ventricular septum (TGA-IVS) in the first few weeks of life. Some studies suggested that the p-ASO is a feasible strategy for patients with TGA-IVS up to age 2 months. The aim of this study was to assess the surgical outcome of the p-ASO in children with TGA-IVS presenting beyond 3 weeks of age.

Method: We analysed the clinical records of 12 children older than 3 weeks with TGA-IVS who underwent p-ASO at our institute from 2010 to 2014. Median age was 43 days (range 22–125 days). Detailed transthoracic echocardiography of the left ventricular (LV) geometry (dimensions, shape, wall thickness, inter ventricular septal motion) was performed. 5 patients restrictive atrial septal defect (ASD) and 4 patients had large patent ductus arteriosus (PDA) at preoperative period. 9 patients had well-preserved LV geometry. Preoperative cardiac catheterization was performed in 3 patients with small or squashed (banana-shaped) LV. All of these patients were underwent p-ASO. Two patients had balloon atrial septostomy earlier in life before their transfer to our center. All patients were cyanotic.

Results: There was no early or late mortality. Two patients needed of peritoneal dialysis because of transient acute renal failure. The mean length of mechanical ventilation, intensive care unit (ICU) and hospital stay was 120 hours, 10 and 18 days, respectively. One patient needed Extra Corporeal Life Support (ECLS). There was no correlation between the preoperative end-systolic, end-diastolic diameters or wall thickness of the LV and the postoperative course. The only important preoperative finding was the non-spherical shape of the LV.

Conclusion: The primary ASO for patients with TGA/IVS still can be tolerated beyond the third week of life. The duration of postoperative ventilation, ICU and hospital stay is prolonged in the late ASO patients. The presence of a large PDA and/or a restrictive ASD is important for the preservation of left ventricular preload. The requirement of ECLS was not different from other patients but the probability of the necessity should be kept in mind.

P-222

Comparison of the systemic hemodynamics of sevoflurane and ketamine for anesthesia induction in children with congenital heart disease

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Objective: Sevoflurane and ketamine are commonly used for pediatric anesthesia during cardiopulmonary bypass (CPB). Comparison of the two agents on hemodynamics has been largely based on clinical indirect indicators. Pressure recording analytical method (PRAM) is a new and direct systemic hemodynamic monitoring technique based on mathematical analysis of the peripheral arterial waveform. We aimed to compare the effects of sevoflurane and ketamine on systemic hemodynamics using PRAM in children undergoing CPB.

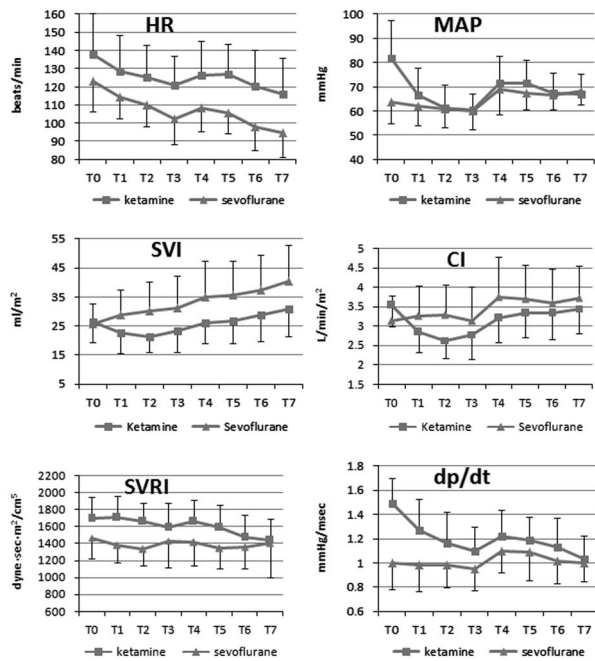


Figure.

Methods: Thirty-two children (2.2 ± 1.1 years) with ventricular septal defect undergoing CPB were randomized into two groups to receive either inhaled sevoflurane (Group S) or intramuscular ketamine (Group K) for basal anesthesia. After basal anesthesia, all children were induced by combined intravenous anesthetics and then intubation. Hemodynamic data were monitored by PRAM including heart rate (HR), systolic, diastolic and mean arterial pressure, stroke volume index (SVI), cardiac index (CI), systemic vascular resistances index (SVRI), the maximal slope of systolic upstroke (dp/dt_{max}) after basal anesthesia, 1,2,5 min after combined intravenous anesthetics, and 1,2,5 and 10 min after intubation. Rate-pressure product (RPP) and cardiac power output (CPO) were calculated using standard equations.

Results: HR, arterial pressures showed a significant decrease during induction ($p < 0.05$), then a small and significant increase at intubation ($P < 0.0001$), followed by a gradual decrease ($p < 0.0001$) in both groups. As compared to Group S, in Group K, the decreases in arterial pressures during induction were significantly faster ($p < 0.01$). Group K had significantly higher HR, arterial pressures, SVRI, dp/dt_{max} , RPP and CPO and lower SVI ($p < 0.01$) during the entire study period. CI was not significantly different ($p > 0.05$). For each increase in CPO, RPP was significantly greater in Group K ($p < 0.0001$).

Conclusions: Compared with sevoflurane, ketamine exerts highly unstable and significant adverse effects on systemic hemodynamics and myocardial energetics during the basal anesthesia in children undergoing CPB. Further studies are warranted to examine the advantageous and disadvantageous effects of various anesthetic agents on systemic hemodynamics in neonates and infants with complex congenital heart diseases during CPB.

P-223

Fontan completion during winter season is not associated with significantly higher mortality or severe morbidity in the early postoperative period

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Objectives: Previous studies suggested that completion of total cavopulmonary connection (TCPC) during winter season was associated with higher early mortality and morbidity. The aim of our study was to compare postoperative outcomes after TCPC completion between patients who were operated during winter and summer season.

Methods: We retrospectively studied 173 patients who underwent extracardiac TCPC completion at our institution between 1995 and 2013. 61 (35%) patients were operated during winter (november-to-march) and 112 (65%) patients were operated during summer season (april-to-october). Baseline characteristics (e.g. age, sex, weight, preoperative oxygen saturation, ventricular morphology or presence of heterotaxy syndrome) were not significantly different between both groups. We compared the following postoperative outcomes: early mortality (i.e. within 30 days postoperatively), intubation time (hours), length of pleural effusions (days), incidence of low cardiac output (i.e. need for intravenous catecholamine therapy longer than 72 hours), intensive care unit (ICU) and hospital stay (days). Fisher's exact test and Mann Whitney test were used for statistical analysis. Data are presented as percentage or median values, respectively.

Results: Early mortality (8% vs. 6%, $p = 0.76$), incidence of low cardiac output (21% vs. 11%, $p = 0.09$), intubation time (13.5 vs. 13 hours, $p = 0.19$), length of pleural effusions (8 vs. 7 days, $p = 0.23$) and ICU stay (4 vs. 3 days, $p = 0.22$) were not significantly different between patients who were operated during winter and summer season. However, hospital stay was significantly longer in patients operated during winter season (16 vs. 14 days, $p = 0.048$).

Conclusion: In our patient series, TCPC completion during winter season was not associated with higher mortality or severe morbidity in the early postoperative period. These results suggest that TCPC completion during winter season might be performed at no significant additional risk for patients.

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Is Surgery Still Needed for Patent Ductus Arteriosus in Preterm Babies?

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Introduction: Patent ductus arteriosus is associated with significant morbidity and mortality in preterm infants but the indications of pharmacological and surgical intervention have been controversial due to the lack of overall survival benefits. Hence there is a wide variation in management of patent ductus arteriosus in preterm infants.

Table.

	Pharmacological intervention	Surgery	Not treated
n	52 (+36)	57 (36 + 21)	91
Males n (%)	21 (40.3%)	36 (63.1%)	50 (54.9%)
Median gestation in weeks (Q3-Q1)	26 (25-27)	28 (26-30)	27 (26-30)
Median Birth weight in grams (Q3-Q1)	850 g (730-1010)	990 g (707-1345)	1010 g (810-1347)
Median NICU stay in days (Q3-Q1)	69 days (37-106)	44 days (20-97)	31 days (9-56)
Median left atrium/aortic ratio (Q3-Q1)	1.76 (1.47-2)	1.61 (1.48-2)	1.6 (1.4-1.8)
Ductus closure rate	37 (43%)	57 (100%)	80 (88%)
Necrotising enterocolitis (NEC)	4/52 (7.6%)	13/57 (22.8%)	17/91 (18.6%)
Intraventricular haemorrhage (IVH)	7/25 (28%)	7/22 (31.8%)	13/53 (24.5%)
Chronic lung disease (CLD)	17/25 (68%)	16/22 (72.7%)	25/53 (47.1%)
Death	8/52 (15.3%)	5/57 (8.7%)	13/91 (14.2%)

Methods: We reviewed all preterm neonates diagnosed with a haemodynamically significant patent ductus arteriosus between 1992 and 2013 at the University Hospital of Wales. We compared the clinical status, and outcomes of neonates being managed conservatively versus with pharmacological intervention or surgery.

Results: A total of 200 cases were included. 88 babies had pharmacological intervention. Along with 21 sickest preterm infants an additional 36 cases with failed pharmacological intervention underwent surgery. Despite being the sickest, with higher NEC, IVH and CLD rates, the babies undergoing surgical ligation (57 cases) had a lower mortality rate when compared with pharmacologically treated or non-treated babies. The results are summarised in the table.

Conclusion: Our study showed that the babies underwent surgical PDA ligation had a shorter hospital stay and better survival rates indicating that surgery still has a place in the management of PDA in sick preterm babies.

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Volatile sedation in children following congenital cardiac surgery is safe and efficient – a case series

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Introduction: Spontaneous breathing is essential in children following congenital cardiac surgery – especially with (Hemi-)Fontan circulation or restrictive right ventricular physiology. However, sedation with conventional drugs as benzodiazepines and opioids, regularly impedes appropriate spontaneous breathing. Volatile sedatives (e.g. Isoflurane), applied by the AnaConDa® device, allow reduction of other sedatives and facilitate spontaneous breathing. Isoflurane is a halogenated hydrocarbon, major side effects are systemic vasodilation and potentially elevated fluoride levels. Effects on neurological outcome remain unclear, thus it is a final rescue option if other sedative drugs fail. Aim of our study was to analyze spontaneous breathing and the safety of this method.

Patients: Retrospective analysis of the electronic records of 12 children (5 with Fontan, 2 with Hemifontan, 5 with restrictive RV) with prolonged or difficult sedation following congenital cardiac surgery. Analysis of (1) time to achieve spontaneous breathing and extubation, (2) hemodynamic and metabolic parameters, (3) doses of conventional sedatives and (4) fluoride levels (before starting isoflurane, after 48 h and afterwards weekly). As volatile anesthetics are not licensed for sedation in children parental consent was obtained.

Results: Age 2–30 months. Median duration prior to Isoflurane sedation: 6,8 days, median duration of Isoflurane sedation: 7,9 days. Patients with >50% spontaneous breathing: 29% after 6 hours and 50% after 18 hours. Extubation: 6 patients within 1 hour, 3 patients within 3 hours and 10 patients within 8 hours. Stable hemodynamics (MAP: +1% ± 21%, heart rate: -10% ± 30% after 120 min), ScvO₂, lactate levels, urine output and inotropic indices. Withdrawal of all conventional sedatives and dose reduction of opioids. Fluoride levels <30 µmol/l (median 12 µmol/l). No relevant side effects.

Conclusions: Volatile sedation provides initiation of effective spontaneous breathing and timely extubation in patients with congenital heart disease and prolonged or difficult sedation. Hemodynamics remained stable, fluoride levels were low during and after therapy. However, sedation with Isoflurane remains a rescue option as its effects on long-term neurologic outcome are unknown. Further investigations concerning this topic are necessary.

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Early And Late Results Of Surgically Managed Congenital Vascular Rings

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Objectives: Persistent respiratory symptoms or feeding problems in children may be associated with a congenital vascular ring. Increasing awareness for this problem exists with pediatricians, but the long-term outcomes are not well described. This study aims to analyze the clinical presentation and surgical treatment of a series of vascular rings, and to evaluate risk factors for mortality and late outcome.

Patients and Methods: Since 1993, 58 vascular ring patients (55% male) were treated surgically at our center. Median age at operation was 1 year (range 3 days–27 years). Presenting symptoms were mainly respiratory (86%), including need for preoperative ventilation in 16%, 32% had feeding problems, 2 patients were asymptomatic. The most common diagnosis was double aortic arch (55%), followed by right arch with aberrant left subclavian artery and ductal ligament (36%), left arch with right arteria lusoria (7%), and pulmonary artery sling (2%). Associated anomalies (cardiac, gastrointestinal, chromosomal) were present in 36%. Left thoracotomy for interruption of the vascular ring was the preferred access in 90% of cases.

Results: Median time to extubation and hospital stay were 0,4 days (range 0–8) and 5 days (range 3–371) respectively. Mean follow-up was 7,8 ± 5,7 years, and was 100% complete. Mortality was 8,6% (5 patients), occurred during early follow-up (within 1,5 years postoperatively), and was associated with anatomical diagnosis ($p < 0,05$), preoperative intubation ($p < 0,0001$) and concomitant anomalies ($p < 0,05$). Freedom from symptoms at 1 month and 6 months was 64% and 81% respectively. Freedom from inhalation therapy at last follow-up was 83%. Dysphagia symptoms always disappeared. A significant relationship was found between the freedom from symptoms at 1 month, at 6 months, and freedom from inhalation therapy at last follow-up, with preoperative ventilation ($p < 0,0001$) and anatomical diagnosis ($p < 0,05$).

Conclusion: Surgical relief of tracheo-oesophageal compression by a vascular ring is usually effective, with a swift disappearance of symptoms. In 17% of children however, respiratory symptoms persist, necessitating chronic inhalation therapy. Associated anomalies may compromise the short-term outcome. Patients with a double aortic arch, especially when ventilated before operation, are at higher risk to remain symptomatic during long-term follow-up, particularly at the occurrence of infectious exacerbations.

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Fate of the pulmonary autograft and left ventricle after Ross-operation in the pediatric population

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Introduction: Retrospective study to evaluate postoperative results in children after the Ross operation with special respect to the pulmonary autograft and left ventricular (LV) dimensions.

Methods: 31 children <20 years-old (22 male) after Ross operation (1994–2008) were included in the study. The first postoperative and last follow-up echocardiograms were used to measure: endsystolic dimension of left ventricle (LVESD), enddiastolic dimension of left ventricle (LVEDD), thickness of interventricular septum in diastole (IVS) and of LV posterior wall (HW) (M-mode) and aortic valve ring, aortic root, sinutubular junction in diastole in the parasternal-long axis.

In order to evaluate our results, Z-scores were compared to normal values of patients and children in regression models of Daubeney et al. (1999) and Petterson et al. (2008). t-tests were used to determine significant differences between Ross patients' and normal values.

Results: Mean age at Ross operation was 11.1 years (0.5 – 20 years), mean follow-up time 5.6 years, range 10 – 168 months. Indication for operation was aortic stenosis in 4, aortic insufficiency only in 4, and both in 23 patients. Operative technique was modified root inclusion. One patient died early postoperatively, one was lost to follow-up. At last follow-up, 28 patients had normal (n = 13) or mild valve dysfunction of the autograft (n = 15), one patient was re-operated at the autograft while primary indication for reoperation was homograft replacement.

The z-scores of the aortic ring and STJ did not show a significant change at last follow-up. Z-scores of the aortic root decreased, meaning approaching normal values – according to both reference models.

No change of z-scores occurred for LVESD, IVS and PW, while a significant decrease was noted for the LVEDD.

Conclusions: The autograft demonstrates potential for somatic growth parallel to values of normal patient population, the feared dilation of the aortic root does not occur, even normalization of aortic root dimension was observed with this technique.

Parameters of the LV also did not show an increase of z-scores and LVEDD approached normal values in mid-to long-term follow-up.

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Stent extraction and coarctation repair after palliative stenting in neonates with critical coarctation

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Objectives: Surgery is the treatment of choice for critical aortic coarctation. Surgical repair should be performed as early as feasible, but can be problematic in premature, low birthweight newborns and is associated with a higher (peri-) operative risk in these patients. Therefore palliative coarctation stenting in these newborns or infants in cardiogenic shock can be a feasible approach in order to stabilize these patients until corrective surgery.

Methods: A retrospective data analysis of newborn patients who presented with a critical aortic coarctation in our institution from April 2011 to August 2014 was performed.

Results: In total, ten infants with critical aortic coarctation underwent this two-staged procedure. Median age at stent implantation was 22 days (median weight 3.2 kg), median age at surgery was 4.1 months. Hypoplastic aortic arch was described in all but one patient, who had an interrupted aortic arch type B. Five patients had a bicuspid aortic valve. Six patients were diagnosed with concomitant intracardiac anomalies (ventricular septal defects, atrial septal defects, mitral stenosis or mitral dysplasia).

Follow-up data was available for nine patients for up to 36 months (median 19 months) after surgery and showed a stable outcome without reoperations. Two patients required reinterventions at the distal aortic arch due to recurring stenosis. One preterm infant with several comorbidities and an atrioventricular septal defect died one month after surgery due to sepsis.

Conclusion: Palliative stenting followed by surgical stent removal and coarctation repair are safe and efficient procedures in infants with critical coarctation who are not suitable for primary aortic coarctation repair.

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Right ventricular remodeling after pulmonary valve replacement in children with repaired Tetralogy of Fallot and transannular patch

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Objectives: Right ventricular dilation due to severe pulmonary valve regurgitation is the most frequent reason for re-operations in patients with repaired Tetralogy of Fallot and transannular patch. To preserve the right ventricular function, patients should undergo pulmonary valve replacement as soon as right ventricular end-diastolic volume exceeds 150 ml/m². In this study we assessed right ventricular remodeling after pulmonary valve replacement in children with repaired Tetralogy of Fallot using cardiac magnetic resonance imaging (MRI).

Methods: 30 children with repaired Tetralogy of Fallot and transannular patch underwent cardiac MRI before and after pulmonary valve replacement. Mean age was 128,13 ± 40,29 months, the mean interval of cardiac MRI before and after pulmonary valve replacement was 193,27 ± 135,54 days and 436,83 ± 230,33 days respectively. When MRI data showed a right ventricular end diastolic volume of more than 150 ml/m², pulmonary valve replacement was performed on average 122,76 ± 33,17 months after primary repair.

Results: Significant reduction in end-diastolic and end-systolic right ventricular volume, as well as pulmonary regurgitation fraction could be shown by cardiac MRI. Right ventricular end-diastolic volume decreased from 167,48 ± 40,93 ml/m² to 100,42 ± 26,83 ml/m² (p < 0,05) right ventricular end-systolic volume from 92,47 ± 25,03 ml/m² to 56,83 ± 20,85 ml/m² (p < 0,05) and pulmonary regurgitation fraction from 43,60 ± 14,28% to 3,35 ± 7,77% (p < 0,05). Right ventricular ejection fraction (42,61 ± 7,89% to 44,16 ± 8,37%) and left ventricular volumes did not change significantly.

Conclusion: In children with corrected TOF, transannular patch and RV dilatation of more than 150 ml/m² pulmonary valve replacement led to a rapid, significant RV remodeling as assessed by MRI.

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Legitimate Wedding/A Happy Marriage - Initial experience of a surgical center for children with heart disease in Angola

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A large underserved population of children with congenital heart disease (CHD) is present in many developing countries as in Angola. Children have very limited or no access to cardiovascular surgical care in Africa and will face irreversible heart and lung damage or death. Methods to provide and improve cardiovascular surgical care in developing countries are needed. In recent years,

several strategies have been implemented to address this need. These strategies include transferring children to first-world countries for surgical care, organizing surgical trips to developing countries, and, more recently, creation of local cardiovascular surgical programs. The latter strategy promotes a shift towards taking care of children in their local environment and allows more children to be treated at a fraction of the cost. Unit “twinning” has been proposed rather than the relatively inefficient “medical safaris”. We implemented a dual-nation marriage and a collaborative initiative between local major medical facilities, state health authorities to extend modern pediatric cardiac care to the country.

Goals of the pediatric cardiac care program were to improve outcomes of children with CHD in Angola by training local staff to become independent of foreign aid. Three core functions form the basis of our operations: service delivery, training, and research. Between April 2011 and November 2014, we performed 1517 procedures on 1460 patients (531 infants, 67 neonates); overall mortality of 3,9%.

Problems peculiar to the African context, are discussed. Programmes to qualified pediatric cardiac team are explained. We have participated in research leading to publication of papers in peer-reviewed journals. In spite of our achievements, we recognize the enormous challenges faced by the continent in terms of pediatric cardiac care.

In summary, an attempt has been made to quantify CHD in Angola, to guide planning and training while simultaneously giving real access to first world medical care on a permanent basis.

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Coronary anomalies in children with transposition of the great arteries (TGA) and their impact on long term follow up after arterial switch operation

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Introduction: Coronary anomalies are frequently associated with the TGA. Current method of choice for treatment - arterial switch operation (ASO) includes switch of the great vessels and coronary arteries transplantation thus the coronary anomalies increase the difficulty and force operator to modify the reimplantation sites what may cause significantly higher risk of severe postoperative complications.

The aim of this study was to establish occurrence of the coronary anomalies in patients with TGA after arterial switch operation and their impact on long term follow up including coronary complications.

Methods: We reviewed all 690 arterial switch procedures performed between years 1991–2014 in Department of Cardiosurgery of Polish Mother's Memorial Hospital in Lodz including patients with simple TGA (424pts-62%), TGA associated with VSD (182pts-26%), TGA with aortic arch anomalies (64pts-9%) and 2 stage operation with pulmonary artery banding prior to the ASO (20pts-3%). All of the operations were performed with modifications introduced by JJM. The patients' medical records were reviewed retrospectively to gather clinical and echocardiographic data before surgery, during the perioperative period and postoperatively.

Results: The overall mortality was 7,2% and mean clinical follow was 9,4(SD:5,9) years. Coronary anomalies was observed in 236 patients (34%). The most frequent anomalies are: Cx extending

from RCA (1:LCA;2RCA-Cx) - 49% (115 cases); 1:LCA,RCA;2:Cx - 15% (35 cases) and all arteries arising from right sinus (1:0;2:RCA,LCA) - 11% (26 cases). Among 206 routine coronarography examinations, performed usually between 5 and 10 years after ASO, in 10 cases (5%) mild disturbances were observed with no need of interventions or reoperations. Patients with coronary anomalies had significantly higher risk of reinterventions (RR-3,16;CI95%:1,28-7,79; p=0,012), however they were not related to the coronary arteries. There was no significant correlation between presence of the coronary anomalies and early and late deaths (p=0,59), perioperative complications (p=0,89) and reoperations (p=0,17). During follow up there was 1 reoperation related to the coronary arteries (LCA occlusion) in patient without coronary anomaly.

Conclusions: Coronary anomalies in children with TGA are common finding and occur in about 1/3 of patients. The most frequent anomaly is Cx extending from the RCA. Coronary anomalies increase the risk of reinterventions during follow up.

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Reinterventions and reoperations in patients with transposition of the great arteries after arterial switch operation (ASO)

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Introduction: ASO is currently the method of choice for treatment of the transposition of the great arteries (TGA). Good early and late outcomes are well proved, however still remaining complications which needs to be followed. Neo-aortic regurgitation, supra-aortic pulmonary stenosis and coronary insufficiency are the most important ones.

The aim of this study was to establish occurrence and the most frequent reasons for reoperations and reinterventions in patients with TGA after ASO.

Methods: We reviewed retrospectively all 690 arterial switch procedures performed between years 1991–2014 in Department of Cardiosurgery of Polish Mother's Memorial Hospital in Lodz including patients with simple TGA (424pts;62%), TGA associated with VSD (182pts;26%), TGA with aortic arch anomalies (64pts;9%) and 2 stage operation with pulmonary artery banding prior to the ASO (20pts;3%). All of the operations were performed with modifications introduced by JJM. Patients after ASO are followed in Department of Cardiology and the complete TGA patients database contain clinical, operation and laboratory data as well as results of echocardiographic and other imaging examinations. This data are updated prospectively during each ambulatory or clinical visit.

Results: The overall mortality after ASO was 7,2% and mean clinical follow was 9,4(SD:5,9) years. In the early, perioperative period (30 days) 31 pts (4,5%) needs emergency operation. In this group 45% procedures were related to the postoperative bleeding, 29% - delayed closure of the sternum, 10% - severe condition and hypotension, 10% - ECMO and 6% - tamponade. During late follow up 23pts (3%) were reoperated (29 procedures) - most frequently because of re-coarctation of the aorta (reCoA;20%) and supra-aortic pulmonary stenosis (SVPS;17%). Interventional treatment was performed in 22pts (3%;31 procedures). Major part of this procedures constitute balloon plasty of SVPS (39%) and reCoA (39%). Among analyzed risk factors in multiple regression analysis only associated cardiac anomalies were an independent risk factor for reoperation (RR-2,94;CI95%:1,81-4,81;p<0,001).

Cardiac anomalies associated with TGA (RR-2,3;CI95%:1,5-3,53;p < 0,001) and coronary anomalies (RR-3,16;CI95%:1,28-7,79;p = 0,012) were independent risk factors for reinterventions. **Conclusions:** Frequency of reoperation and reintervention in patients with TGA after ASO remains low. Majority of the procedures are performed because of SVPS and reCoA. Cardiac anomalies associated with TGA have significant impact on the incidence of reoperation and reinterventions.

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Prognostic factors for early outcomes in patients with total anomalous pulmonary venous return

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The aim of this study was to assess postoperative outcomes and prognostic factors of children with total anomalous pulmonary venous return (TAPVR).

Methods: all cases diagnosed with TAPVR from 1973 to 2014 were included. Clinical, echocardiographic, surgical data were collected and analyzed. Patients were divided into 4 groups according to decades of follow-up and comparisons were made between groups to assess changes over time.

Results: 180 patients were included = 78 supracardiac 43%, 48 intracardiac 27%, 35 infracardiac 19% and 19 mixed 11%. Pulmonary venous return was obstructed in 45%, more frequently in infracardiac type (69%). Mean age at diagnosis was overall 88 days, lower in infracardiac group (11 days). Age at surgery was 107 days, decreased from 111 days to 18 days over time. Heart failure was the leading symptom at diagnosis (111/180 cases) and 34% of the cases needed mechanical respiratory support before surgery. Associated lesions were present in 33 cases = 18.3% (the most frequent was VSD: 15/33). Mean weight at surgery was 4.1 kg (min 1.8 kg). Delayed chest closure was observed in 58% of infracardiac cases (overall 18.3%). Mean postoperative duration of MVV, CICU stay and hospital stay were respectively 7.4, 10.1 and 18.4 days. Postoperative in-hospital mortality decreased over decades from 42% before 1980 to 7.4% currently. Decade, duration of bypass, body weight, suprasystolic pulmonary pressure and preoperative acidosis were risk factors for in-hospital death. Arrhythmias and pulmonary hypertension acute crisis occurred in 11% and 10.7% during the early post-surgical course. Mean pulmonary pressure decreased from 68 mmHg preoperatively to 40 mmHg within the 1st week after surgery. Inhaled NO was used since 1992 and had a significant beneficial impact on survival (20-year survival 85% versus 55%, p < 0.0001).

Conclusion: Overall prognosis of patients with TAPVR improved over decades and NO significantly contributed to improve survival. Patients with infracardiac TAPVR have more severe clinical presentation and worse early outcomes.

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Pre-operative Sildenafil administration in children undergoing cardiac surgery; a randomized placebo-controlled preconditioning study

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Introduction: Sildenafil has strong cardiac preconditioning properties in animal studies and is known to have a safe side effect profile in children. Therefore, we evaluated the use of sildenafil preconditioning to reduce postoperative myocardial injury in children undergoing surgical ventricular septum defect (VSD) closure. **Methods:** Randomized, double blind study. Children (1-17 years) undergoing VSD closure were randomized into three groups: Placebo (Control group), preconditioning with 0.06 mg/kg (Sild-L group) or 0.6 mg/kg sildenafil (Sild-H group). Measurements of cardiac injury (CK-MB and troponin I), inflammatory response (IL-6 and TNF- α), bypass and ventilation weaning times, inotropy score, and echocardiographic function were made pre-and postoperatively.

Data expressed as median (range), p < 0.05 considered significant. **Results:** Thirty-nine patients were studied (13/group). Aortic cross clamp time was similar with 27 (18-85) and 27 (12-39) minutes in the Control and Sild-L groups, respectively, but significantly longer with 39 (20-96) minutes in the Sild-H group. Area under the curve of CK-MB release was 1105 (620-1855) ng/mL in the Control group, showed a tendency to be higher in the Sild-L group with 1672 (564-2767) ng/mL, and was significantly higher in the Sild-H group with 1695 (1252-3377) ng/mL. There were no differences in inflammatory response markers, cardiopulmonary bypass and ventilation weaning times, inotropy scores, and echocardiographic function between the groups.

Conclusion: Sildenafil does not reduce myocardial injury in children undergoing cardiac surgery, nor does it alter cardiac function, inotropic needs, or postoperative course.

A paradoxical subclinical increase in cardiac enzyme release after sildenafil preconditioning cannot be excluded.

Clinical Trial Registration number: CTRI/2014/03/004468.

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Stented porcine pulmonary bioprosthesis for repair of the dysfunctional right ventricle outflow tract. 15 years of experience

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Objectives: Surgical results of porcine pulmonary stented bioprosthesis for repair the sequela/residual lesions after primary surgery on the right ventricle outflow tract: Mortality and risk factors; Associated surgical procedures; Morbidity; Prosthetic dysfunction and related factors; Incidence of endocarditis; Analysis of volumes and function of the right ventricle.

Methods: Included all patients who received a porcine pulmonary bioprosthesis between 1999-2014. Primary heart disease: Fallot, pulmonary atresia/stenosis with intact ventricular septum, pulmonary atresia with ventricular septal defect, transposition with ventricular septal defect and pulmonary stenosis, truncus, Ross surgery sequela. Prosthetic dysfunction criteria: new surgical/percutaneous replacement, peak gradient > 50/40 mmHg (echocardiographic/hemodynamic), or pulmonary regurgitation > II.

Results: 91 bioprosthesis in 91 patients. Male: 49 (54%). Age: 30 \pm 13 (1-61) years, 73 adults (>18 years), 18 children (\leq 18 years). More frequent original heart disease: Fallot 68 (75%). Bioprosthesis implanted: 21 (23%) Biocor-St Jude[®]; 70 (77%) Mosaic-Medtronic[®]. Prosthesis size: numbers 19-29, mainly implanted n^o27 in adults and n^o25 in children.

Associated surgical procedures: (1-5) in 67 (74%), more common tricuspid valvuloplasty.

Hospital mortality 2 (2,2%). No mortality in isolated pulmonary replacement. P values for patient's age, sex, cardiopulmonary

by-pass and aortic cross-clamp times, number of previous surgeries, number of associated procedures, right and left ventricle function, were not significant for mortality.

Morbidity in 30 (33%), more frequent tachyarrhythmia.

Follow-up: $3,8 \pm 3,7$ (0,1-15) years. No lost patients. Late mortality 1 (1,1%), owed to complications prosthetic endocarditis related.

Echocardiography peak transpulmonary gradient: 20 ± 9 (4-53) mmHg.

None of the bioprosthesis required reoperation, but 3 patients were reoperated for other indications. Interventional catheterizations: 4 (2 valve-in-valve, 2 not bioprosthesis related).

Prosthetic dysfunction: 4 (2 valve-in-valve, 1 gradient > 50, 1 pulmonary regurgitation III). None of the dysfunction criteria were statistically significant.

Incidence of late prosthetic endocarditis: 3 (3,3%)

Improved right ventricle volumes after surgery resulted significant ($p < 0,001$). Neither right ($p = 0,14$) nor left ($p = 0,76$) ventricular ejection fraction improvement were significant.

Conclusions: Surgical repair of dysfunctional right ventricle outflow tract by porcine bioprosthesis is performed with low mortality. Unlike percutaneous techniques, surgery allows associated procedures and bioprosthetic endocarditis is uncommon. Survival curve without reoperation related to bioprosthesis reaches 15 years.

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Mitral valve replacement in children after failed valvuloplasty. The last technical option

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Objectives: Mitral valve replacement (MVR) is the last option after a failed valvuloplasty for children with a severe mitral lesion. We present our surgical results, technical solutions, mortality risk factors and follow-up.

Methods: 23 mechanical MVR in 21 patients <18 years old, operated during 2002-14. Statistical: SPSS 15.0.

Results: Age at MVR 67 ± 58 (7-200) months, women 17 (74%), weight: 19 ± 16 (5-65) Kg. Preoperative diagnosis: 1 mitral regurgitation (MR) in L-transposition of great arteries; 2 rheumatic disease; 6 residual MR in atrioventricular septal defects; 14 congenital mitral lesion (isolated or in a Shone complex). Surgical indication: mitral stenosis (MS) 7 (30,5%), MR 10 (43,5%), MS-MR 6 (26%). Previous surgery: 15 (65%). Age at previous surgery: 19 ± 23 (0,1-79) months. Number of previous surgeries: $1,3 \pm 1$ (0-4). All patients had one/more valvuloplasty, before or during MVR time. NYHA preoperative status: III-IV 91%.

Preoperative echocardiography: peak transmitral gradient $19,7 \pm 12$ (3-40) mmHg; medium $11,6 \pm 9$ (1-30). MR: no 3 (13%); mild 2 (9%), moderate 4 (17%), severe 14 (61%). Mitral annulus: 23 ± 7 (13-39) mm. Leaflet anomalies: 9 (39%). Single papillary muscle: 1 (4%).

Concomitant moderate or severe pulmonary hypertension in 17 (74%). Left ventricle ejection fraction (LVEF): normal 20 (87%), dysfunction 3 (13%).

Surgical approach: transeptal 7 (30%), left atriotomy 14 (61%), superior-septal 2 (9%). Prosthesis size: 20 ± 4 (16-31).

Conservation of papillary muscles: 7 (30%). Supra-annular implantation: 5 (22%).

Cardiopulmonary by-pass time: 159 ± 45 (90-237) minutes, aortic clamp time: 113 ± 32 (65-181).

In hospital results: mortality 2 (8%); intubation: 92 ± 176 (1-600) hours; intensive care unit stay 11 ± 12 (2-46) days and hospital stay

23 ± 18 (7-69). Morbidity in 14 (61%), most frequently an additional pacemaker: 3 (15%).

Follow-up: 31 ± 39 (0,5-176) months. Late mortality: 4 (20%). Reoperation in 2 (10%), because patient overgrowth/mismatch and prosthesis thrombosis. NYHA status: I-II 87%. Echocardiography: peak transmitral gradient 21 ± 12 (8-51) mmHg, medium 9 ± 6 (3-25). All patients have no MR and good LVEF. Moderate or severe pulmonary hypertension in 45%.

Whereas total mortality (6 patients) mostly happened in the youngest, p value for patient's weight, age and prosthesis size were not significant for mortality.

Conclusions: MVR is the last option, even in small annulus, with inhospital mortality rate (8%) similar at expected mortality in RACHS-1 risk category, but higher (20%) in follow-up, due to anticoagulation troubles. NYHA status and pulmonary hypertension improved. MR disappears in all, but re-stenosis is still a matter of concern.

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Outcomes in Fontan surgery: the left ventricular hypoplasia as an independent risk factor

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Objective: The group of patients with hypoplastic left heart syndrome (HLHS) palliated with Fontan surgery now comprise a larger proportion thanks to the improvement in survival. The aim of the study is to identify the impact of HLHS in postoperative Fontan results and analyze risk factors for morbidity and mortality.

Methods: Retrospective study of patients palliated with Fontan between 2003-2013 at a single center. Preoperative variables, surgical variables and postoperative complications were collected. Risk factors for acute failure, chronic failure and complications during follow-up were analyzed. HLHS results were compared with those obtained from all patients with Fontan.

Results: 137 patients were recruited. Of these, 41 patients with HLHS (41/137 30%) were identified. The survival was 87.8% (36/41). Mean follow-up: 5 years (1.9 to 11.6). Type of surgery: Fontan in two stages, extracardiac conduit 88% (37/41), lateral tunnel 12% (5/41), fenestrated 90% (37/41), Norwood Sano 73% (30/41). We compared the results with other congenital heart defects: mortality of 12.2% (5/41) vs 4.1% (4/96), acute failure 14.6% (6/41) vs 4.1% (4/96) and chronic failure 12.1% (5/41) vs 2% (2/96). Patients with HLHS were younger before surgery (5.8 ± 1.5 vs 7 ± 3), they had worse ventricular function (10.5% vs 1%) and greater distortion of the pulmonary branches (28.6% vs 4.7%). After surgery HLHS patients presented: longer stay in PICU (15.5 vs 6 days), greater need for early reoperation (33.3% vs 15.2%), higher rate of intervention (40% vs 12.8%), longer pleural effusion (12 vs 9 days), higher incidence of chylothorax (31.6% vs 11.5%), thrombus (20.5% vs 8.8%) and prolonged antiarrhythmic treatment (22.5% vs 11.2%).

Conclusions: Patients with HLHS have a significant impact on morbidity and mortality. This is the Spain's longest series. In this series HLHS have three times more mortality and acute failure and five times higher incidence of chronic failure. The study of risk factors may be useful to optimize treatment and improve postoperative outcomes.

P-238

Subaortic stenosis surgery in children: our experience with simple versus complex forms

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Introduction: Subaortic stenosis in children is the second most frequent cause of left ventricle outflow obstruction. Usually is progressive and clinical symptoms are mild. There is no consensus about the best moment to operate these patients considering the subaortic gradient and aortic insufficiency progression. We present our experience during the period 2007-2014.

Material & Methods: Retrospective study of 43 surgeries in 40 children aged ≤ 15 years. Main indication of surgery was subaortic stenosis (isolated membrane or fibromuscular tunnel). Depending if they had previous cardiac/great vessels surgery or not, two groups were defined respectively named complex (n = 20) and simple (n = 23).

Results: Complex group patients have worse subaortic stenosis with higher gradients (89 ± 22 mmHg in complex, 72 ± 22 in simple) and 75% tunnel predominance (70% membrane in simple group). Children of the complex group have more associated lesions (bicuspid aortic valve 20%, aortic coarctation 60%), and need surgery at a younger age (57 ± 41 months) compared with simple group (110 ± 54 months). Most of them were asymptomatic in both groups. Operations were performed under extracorporeal circulation. All cases received membrane/tunnel resection \pm Morrow miectomy, and also in complex group were required more aggressive techniques (20% modified Konno or Ross-Konno) with longer by-pass and aortic cross-clamp times. Hospital mortality: 4,3% in simple, 0% in complex group. Morbidity related to pacemaker implantation because of postoperative atrioventricular block was 20% in complex and 0% in simple group. Mean follow-up: 35 ± 24 months. Late mortality: 4,5% in simple, 5% in complex group. Simple group had lower reoperation rate during follow-up (9%) than complex group (21%). Aortic insufficiency increases over time in both groups.

Conclusions: Subaortic stenosis resection during childhood has good results in simple and complex forms. Surgery does not prevent progression of aortic insufficiency, neither recurrence of subaortic stenosis. Our mortality is low in both groups, but morbidity and reoperation during follow-up are higher in complex group.

P-239

Extubation has acute positive hemodynamic effects in the unstable patients after total cavopulmonary connection

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Introduction: After 2009 we aimed early extubation in all patients following Total cavopulmonary connection (TCPC), including the unstable ones. We investigated the acute hemodynamic effects of timely postoperative extubation in the unstable patients following TCPC.

Methods: Between 2009-2013 145 patients have received TCPC in our institution; 50 exceeded the 75th percentiles for volume requirement or inotrope score (135 ml/kg and 19 respectively) in the first 24 postoperative hours and were defined as unstable. The unstable patients, who were successfully extubated in the first 12 postoperative hours, were included in the study (n = 43). The volume requirements, inotrope score, arterial pressure, cavopulmonary pressure, arterial oxygen saturation and partial CO₂ pressure were recorded in the first 24 postoperative hours at fixed time points related to extubation. Their changes were analyzed with Pearson (r-coefficient) correlation and with paired T-test comparison of the values. Presented is the comparison between the values 1 hour before and 1 hour after extubation.

Results: Median ventilation time of the included 43 patients was 5 (range 2-12) hours. Mean arterial pressure increased during the first 24th postoperative hours ($r = 0.50$, $p < 0.001$) with the most significant change occurring between 1 hour before and 1 hour after extubation - 52 ± 7 vs. 58 ± 9 mm Hg, $p < 0.001$. Cavopulmonary pressure decreased with time ($r = -0.34$, $p < 0.001$), but no acute drop was observed with extubation - 18 ± 5 vs. 17 ± 4 mm Hg, $p = 0.22$. Volume requirements decreased with time ($r = -0.651$, $p < 0.001$) with strongest differences between 1 hour before and 1 hour after extubation - 17.3 ± 15 vs. 3.4 ± 5.5 ml/kg, $p < 0.001$. Similar changes were observed with the inotrope score ($r = -0.47$, $p < 0.001$) with strongest reduction around extubation - 12.0 ± 6.6 vs. 9.7 ± 5.4 , $p = 0.008$. Arterial oxygen saturation decreased slightly ($r = -0.12$, $p = 0.03$) and dropped significantly with extubation (93 ± 6 vs. $89 \pm 8\%$, $p = 0.005$). Partial CO₂ pressure remained stable ($r = 0.06$, $p = 0.27$) with a temporary increase in the first hour after extubation - 45 ± 7 vs. 50 ± 8.4 mm Hg, $p = 0.002$.

Conclusion: Extubation of the hemodynamically unstable patients in the first hours following TCPC has beneficial hemodynamic effects and enables an immediate reduction of the volume and inotrope treatment. Thus, extubation can be a powerful management tool in the stabilization of these patients.

P-240

Pulmonary valve replacement after previous TOF repair in childhood

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Objectives: Patients born with Tetralogy of Fallot (TOF) being the most common form of cyanotic congenital heart disease frequently survive into adulthood. As pulmonary valve insufficiency is a well-known long-term complication after surgical TOF repair in childhood therapeutic strategies for these patients must be established. For a long while homograft or Contegra-graft implantation was the only therapeutical option but nowadays the implantations of biological prostheses in pulmonary position lead to convincing results, too. Implantation of these valves provides the opportunity to perform valve-in-valve procedures later on.

Material: From 2000 to 2014 in the Department of Cardiothoracic Surgery of the University of Muenster, 81 patients with previous TOF repair in childhood needed pulmonary valve replacement due to severe pulmonary insufficiency. Either homografts (n = 46) or Contegra-grafts (n = 17) were routinely used till in 2011 pulmonary valve replacement (PVR) with biological valve prostheses (n = 18) was established. Perioperative data of these patients with PVR was analyzed retrospectively.

Results: 18 patients of either sex at a mean age of 29 [range 5-72] years were surgically treated for pulmonary insufficiency. The primary operation was performed at an average of 23 [range 5-48] years ago in childhood. Now either St. Jude Medical Trifecta™ (n = 8) or Carpentier Edwards (CE) Perimount™ prostheses (n = 10) were implanted. Mean size of the implanted biological prostheses was 23 [19-27] mm.

Postoperative echocardiography was used to exclude paravalvular leakages (n = 0). A mild insufficiency of the pulmonary valve could be identified in 5 patients (28%) - the central jets based on the tissue valve design of the biological prostheses. Stenoses of the valves with a mean v_{max} of 2.2 [1.6-2.5] m/s did not lead to clinical symptoms or surgical interventions, respectively.

In-hospital-mortality was 1.2% (1 pt.). This patient, the oldest one of all (72 years) died 40 days after surgery due to a complicated ICU-stay with long-term ventilation and renal failure. All other patients were discharged home.

Conclusion: Pulmonary valve replacement using biological prostheses in patients with TOF repair in childhood is a safe and clinically reliable therapeutic option to treat severe pulmonary insufficiency properly.

P-241

Long term outcome of cyanotic newborns with complete atrioventricular septal defect combined with tetralogy of Fallot requiring a staged repair is not worse than primary repair

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Objectives: Primary repair is the preferred strategy for surgical treatment of complete atrioventricular septal defect combined with tetralogy of Fallot (CAVSD-TOF). However, in cyanotic newborns, a staged procedure may be required. Long term outcome of these patients was compared to primary repair.

Methods: Data of 56 patients with CAVSD-TOF who underwent surgery between 1974 and 2013 were reviewed. Endpoints of the study were mortality and reoperation. The patients were divided into group A: staged repair, and group B: primary repair. Indication for a staged repair was mostly a cyanosis in neonates.

Results: Group A, and B comprised 31, and 25 patients, respectively. In group A, 8 patients died before reaching complete repair, and 3 are currently awaiting repair. The patients in group A were younger at time of initial surgery ($p = 0.02$), and exhibited more often cyanosis (21 vs. 5 patients, $p = 0.003$) compared to group B. Survival at 10 years following initial surgery was $76.4 \pm 8.2\%$ in group A, and $87.1 \pm 7.0\%$ in group B ($p = 0.3$). The freedom from reoperation for regurgitation of the atrioventricular valve (AVVR) at 10 years following repair was $82 \pm 9.4\%$ in group A, and $77 \pm 9.0\%$ in group B ($p = 0.5$). Moderate or more AVVR prior to repair was the only risk factor for reoperation ($p = 0.01$).

Conclusions: Cyanotic neonates who require an urgent treatment exhibit a similar long-term outcome after a staged repair compared to patients after primary repair.

P-242

Extracorporeal membrane oxygenation (ECMO) to aid cardiopulmonary resuscitation in children for cardiac arrest occurring on the paediatric cardiology ward

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Introduction: The use of ECMO rescue therapy for children suffering cardiac arrest (ECPR) has been mostly limited to arrests occurring within the ICU, operating room or catheterisation laboratory. At present, published evidence does not support ECPR for arrest occurring on the ward. We hypothesised that with good quality CPR and rapid ECPR deployment, similar survival would be achievable in patients with cardiac disease who arrest on the ward. Since 2005, we have had a policy of activating ECPR for paediatric cardiology ward arrests. Our objective is to review the outcome in this population.

Methods: Freeman Hospital is a paediatric cardiac surgical centre that undertakes 250-300 open surgical cases per year and one of two centres in UK undertaking heart transplantation.

Ward nursing and medical staff underwent simulation training in ECPR and were taught to activate the team for cardiac arrest of duration >5 min. Perfusion and surgical staff were in-house during weekdays and on-call during weekday out-of-hours and weekends. Patients were transferred to ICU or operating room for cannulation.

Patients aged < 16 years who suffered cardiac arrest on paediatric cardiology ward and received ECPR between 2005 and 2015 were identified. ECPR was defined as ECMO established during CPR. In addition, a patient suffering intermittent multiple arrests during cannulation was also defined as ECPR. Survival and Paediatric Cerebral Performance Category at hospital discharge were determined from the medical record.

Results: Five children, aged 2d to 13 y, received ECPR after cardiac arrest on the cardiology ward during 10-year period. Of these, 2 had end-stage heart failure complicating congenital heart disease, 2 were medical patients and one was post-surgery. The median duration of CPR prior to ECMO was 57 min (range 45-93 min). Of the five patients, 2 (40%) survived to discharge. The two survivors were neurologically intact at hospital discharge (PCPC 1). Both survivors underwent transplantation during the admission following ECPR. Two patients developed brain death complicating cerebral oedema and one patient died due to VAD-related haemorrhage during bridge to transplantation.

Conclusion: Although a rare event, children who suffer cardiac arrest on cardiology ward can survive neurologically intact with ECPR despite prolonged periods of resuscitation.

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Current Trends in the PostAnesthetic Management of Pediatric Patients with Pulmonary Arterial Hypertension: Results of a Survey

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Introduction: Rates of perioperative cardiac arrest and death in children with pulmonary arterial hypertension (PAH) are reported to be 20 fold or greater than rates for all children undergoing anesthesia for all procedures, including catheterizations. Recent advances in the understanding of PAH, have resulted in expanded therapeutic options and dramatic improvement in disease prognosis and survival. This may impact the postanesthesia care management and disposition of these patients.

Methods: A survey regarding postanesthetic practices for patients with PAH was sent to congenital cardiac programs across the United States via emails, text and personal communication.

Results: Thirty institutions were surveyed. 5/30 (17%) were private practice and 25/30 (83%) were academic. 1/30 (3.3%) did not reply. 23/29 (79%) institutions made decisions on a case by case basis. 6/29 (21%) had formal guidelines. The guidelines of 4/6 institutions solely address those with moderate to severe pulmonary hypertension. The responses show that postanesthetic decisions are based on a combination of the following: (1) Severity, (2) Etiology –PAH secondary to congenital heart disease, chronic lung disease of prematurity and congenital diaphragmatic hernia being considered less fragile than primary PAH, (3) Procedure complexity –specific mention of airway procedure or intubation as intensive care unit (ICU) admission. Most guidelines recommend bypassing the postanesthetic care unit (PACU) when admitting to ICU for postanesthetic recovery. If PACU stay happens prior to ICU admission, a detailed sign off is emphasized.

No institution specifically reported to have a “blanket policy” for admission of every patient with PAH, although 2 institutions report that almost all of their patients with PAH stay in house postanesthetic, with most of them admitted to the ICU.

Conclusions: As the medical management of pediatric PAH continues to improve, the perianesthetic care of these patients must evolve with it. Even in this high risk patient group, an indiscriminative overly conservative approach for every patient with this diagnosis is unlikely to be in their best interest. The national trend in current practices gathered from the survey seems to reflect this evolution of care. As this paradigm shifts, evidence-based guidelines are needed to guide these children’s perianesthetic care.

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It was not discrete examination findings but disadvantageous conditions before repair which was related to postoperative pulmonary venous obstruction in patients with simple total anomalous pulmonary venous connection

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Introduction: Pulmonary venous obstruction (PVO) occurs after operation of total anomalous pulmonary venous connection (TAPVC) despite surgeon’s ingenuity. We empirically suppose postoperative PVO (postop-PVO) has relevance to preoperative PVO (examination findings which indicate obstructive lesion). We investigated what preoperative conditions are related to postop-PVO.

Methods: The medical records of 44 TAPVC patients after repair were reviewed (supracardiac 20, infracardiac 14, cardiac 8, and mixed 2). All surgeries were performed by a same operator between 1993 and 2008. We defined PVO as either vertic vein velocity exceeding 2.0 m/s or vertical vein flow being continuous. Patients were divided into two groups: postop-PVO (n = 11); no postop-PVO (n = 33). Preoperative conditions influencing postop-PVO were determined.

Results: In univariate analysis postop-PVO was related to following factors: prolongation of preoperative respiratory support (>1 day, 36% vs. 3%; p = 0.010), early ages at repair (≤ 12 days, 72% vs. 27%; p = 0.012), light body weight at repair (≤ 3.0 kg, 55% vs. 18%; p = 0.05), small cardio-thoracic ratio (≤ 0.49, 45% vs. 6%; p = 0.067), and inotropic support (27% vs. 6%; p = 0.090). After multiple logistic regression analysis postop-PVO was independently associated with an odds ratio of 21.9 (p = 0.016) for respirator support on a prolonged time and 6.3 (p = 0.032) for light body weight at operation. Following preoperative factors had no relation to postop-PVO, such as type of TAPVC, dimension of left ventricle, echocardiographic PVO, degree of pulmonary congestion, intensity of pulmonary hypertension. Similarly, operative method, and learning level of operator were not significantly related to postop-PVO. If TAPVC patients had no risk factors, only 4% of patients had PVO after repair. However, if TAPVC patients had two or more factors, 75% of patients had postop-PVO.

Conclusions: Contrary to our exception, factors which indicated preoperative PVO (small left ventricle, and pulmonary congestion), and even direct evidence of PVO by echocardiograms were not significantly related to postop-PVO. Postop-PVO was associated with bad conditions as pre-operation. Patients with more than two risk factors suffered postop-PVO with high rate. We should mind more than one disadvantage rather than every single finding of examination to predict post-op PVO in patients before TAPVC repair.

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Unbalanced pulmonary stenosis and pulmonary circulation disturbance persists in patients after Fontan who underwent Norwood-Glenn following bilateral pulmonary banding

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Introduction: In patients with hypoplastic left heart syndrome (HLHS) and the relative disease, we perform bilateral pulmonary artery banding (BPAB) as 1st palliation. As 2nd palliation we previously selected Norwood-Glenn (N-G). On acute stage severe stenosis of left PA frequently occurred. We assessed whether pulmonary circulation disturbance persisted after Fontan in patients who underwent N-G following BPAB.

Methods: We defined BPAB group (n = 15) as patients who underwent BPAB as 1st palliation, N-G as 2nd palliation, and attained to Fontan procedure. We also defined non-BAP group (n = 72) as patients who got to Fontan through strategy other than BPAB during the same period (between 2007 and 2013). Cardiac catheterization was performed in stable periods after Fontan. The following indexes were calculated with the arterial carbon dioxide pressure and arterial oxygen pressure (PaO₂) which were measured during cardiac catheterization: value of alveolar to arterial oxygen tension (AaDO₂), RI (AaDO₂/PaO₂), and P/F (PaO₂/0.21). Cardiopulmonary functions were compared between two groups.

Results: As for pulmonary circulations diameter of left PA was smaller in BPAB after Fontan (59% vs. 86%, p < 0.001). Index of PA was also smaller in BPAB (162 vs. 248 mm/m², p = 0.0063). However, there were no significant differences in diameter of right PA. In BPAB AaDO₂ was significantly higher (35 vs. 29 mmHg). In BPAB the number of patients with inferior RI (>40) was larger (80% vs. 32%, p < 0.001); that with inferior R/F (<322) was larger (40% vs. 13%, p = 0.028). Consequently, saturation oxygen of aorta was lower in BPAB (92% vs. 94%, p = 0.0077). Smaller major ventricles significantly existed more in BPAB. Other cardiac functions were not significantly different. In BPAB levels of gamma-glutamyl transpeptidase were increased (98 vs. 68U/L, p = 0.024), and levels of prothrombin time decayed (67 vs. 74%, p = 0.028).

Conclusions: Our study showed stenosis of left PA persisted after Fontan in patients who underwent N-G following BPAB. Unilateral stenosis of PA would cause ventilation-perfusion mismatch, which resulted in oxygenation disturbance. Furthermore, considerable smallness of PA might lead to venostasis, which is related to hepatic disturbance. From a long-term perspective on pulmonary circulation, we should select strategy by which pulmonary arteries are grown.

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Results of atrioventricular valve repair in patients with functional single ventricle

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Introduction: Significant atrioventricular valve (AVV) regurgitation is known as a risk factor for mortality and poor long-term outcome following modified Fontan operation in patients with functional single ventricle.

Methods: Between 2000 and 2014, 24 consecutive Fontan candidates underwent AVV repair. A retrospective study was carried out

to evaluate the indications for AVV repair, the techniques of repair and the clinical outcome.

Results: All patients had a functional single ventricle: left-dominant (9 pts-37%), right-dominant (6 pts-25%), unbalanced atrioventricular septal defect (4 pts-17%) and double inlet ventricle (5 pts-21%). AVV regurgitation was evaluated by echocardiography and graded as significant (> grade 2) in all patients. A staged strategy for Fontan was applied; 18 patients (75%) reached Fontan completion. Primary AVV repair was performed at an age ranging from 11 days to 16 years (median: 5.4 yrs) and the timing was variable: before bidirectional Glenn (BDG) in 2 pts (8%), at the time of BDG in 9 pts (38%), between BDG and Fontan in 5 pts (21%) and at the time of Fontan completion in 8 pts (33%). Various techniques for AVV repair were used: annuloplasty (11 cases), commissuroplasty (9 cases), closure of cleft, fenestration or indentation (9 cases), edge-to-edge repair (3 cases), chordal shortening (1 case) and valve closure (4 cases).

There were two deaths (8.3%): one early cardiac death and one late non-cardiac. 6 patients (25%) underwent 8 reoperations for recurrent regurgitation. The first reoperation included re-repair in 3 patients and AVV replacement in 3 patients. The second reoperation included repair in 1 patient and replacement in 1 patient. At last follow-up (mean follow-up: 9.9 years), 3 patients had significant (> grade 2) residual AVV regurgitation, 4 patients had valve replacement, ventricular dysfunction was present in 5 patients and one patient had had heart transplantation.

Conclusions: In Fontan candidates, early repair of significant AVV regurgitation is essential to minimize volume overload and preserve ventricular function. AVV repair provides acceptable results and the need for reoperation is relatively low. Valve replacement, if needed, remains a valuable option.

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Double Outlet Right Ventricle with Non-Committed Ventricular Septal Defect

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Objective: The management of Double Outlet Right Ventricle (DORV) associated with anatomically non-committed Ventricular Septal Defect (NCVSD) constitutes a surgical challenge. The limits for, and the specific outcomes after anatomical versus univentricular repair still remain to be established.

Methods: Between 1993 and 2011, 35 consecutive patients presenting with DORV/NC-VSD and 2 adequately sized ventricles were included into the study at two centers forming the National Referral Center. The selection criteria included the absence of outflow tract VSD: 21 inlet (4 complete atrio-ventricular septal defect (AVSD)), 9 muscular and 5 perimembraneous. RVOTO was present in 18/35 (51%). Twenty patients had undergone 25 initial palliative procedures.

Results: Anatomical repair by means of intraventricular baffle construction was performed in 23 (Group I) at a median age of 10.5 months. VSD was surgically enlarged in 11 (48%). An associated RVOT reconstruction was required in 11 and Arterial Switch Operation (ASO) was done in 5. The remaining 12 patients underwent univentricular palliative repair (Group II). There were 4 hospital deaths (11.4%): 3 in Group I and one in Group II ($p = .06$). 8/20 survivors of group I patients underwent 13 reoperations after a median delay of 24 months, with subaortic stenosis being the main cause for reoperation (6/8). There was one

late death in group 2. At last visit, all survivors were in NYHA class I-II. Ten years actuarial survival rate and freedom from reoperation were respectively $74.7 \pm 5\%$ and $58 \pm 5\%$ in Group I, and $80 \pm 7\%$ and $71 \pm 7\%$ in Group II. Univariate analysis showed that AVSD and/or isolated mitral cleft were associated with death ($p = .04$) and need for reoperation ($p = .038$).

Conclusions: Despite the need for complex procedure and the high incidence of reoperation for subaortic obstruction, our results suggested the long-term advantages of anatomical repair in DORV with NCVSD. The presence of associated AVSD and/or isolated mitral cleft was the only risk factors for mortality and reoperation.

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Early and Midterm results of aortic root translocation using pivot rotation technique for complex forms of TGA

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Objectives: To clarify the early and midterm results of aortic root translocation (ART) using "pivot rotation technique" in children with complex forms of TGA.

Methods: A retrospective review of 6 patients who underwent ART in our institute from January 2006 to December 2013. ART has been applied for patients with d-TGA (3), DORV (2), ccTGA (1) having LVOTO and small/remote VSD. Seven palliative procedures had been performed in 5 patients previously. The principal details of the operation are as follows; The aortic root was partially excised from the RV and only one coronary artery button was detached. Then the root was translocated over the LV by rotating it around the un-detached coronary pivot. After the re-implantation of detached coronary artery and VSD closure, RVOT is reconstructed.

Results: Median age and body weight at ART was 1.8 (0.6~5.0) years and 10.1 (7.5~14.6) kg, respectively. Their aortic and pulmonary valvular size was 4.47 (2.47~6.08) and -3.76(-5.45~-0.9) in z-score. RVOT reconstruction was achieved with a tricuspid handmade ePTFE graft (3) or native pulmonary valve (3) (= double root translocation:DRT). One patient required pacemaker implantation for complication of complete AVB. With a median follow-up of 2.0 (1.3~7.6) years, there were no early or late deaths. One patient required reoperation for pulmonary valvular stenosis 4.3 months after DRT. None of the patients have developed aortic regurgitation, LVOTO, nor coronary problems during the follow-up.

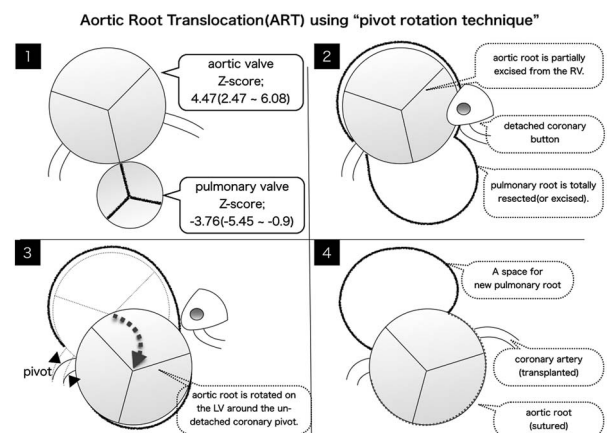


Figure.

Conclusions: ART using pivot rotation technique will be a good surgical option for complex forms of TGA. Long-term benefits need to be evaluated with a longer follow-up.

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Recovery of the left ventricle and mitral valve function long after repair of anomalous origin of left coronary artery from pulmonary artery

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Background: The aim of this study is to evaluate long term results after surgical repair of anomalous origin of left coronary artery from pulmonary artery (ALCAPA) in children. We focused on the function of both left ventricle and mitral valve.

Methods: Medical files of patients operated for ALCAPA for twenty seven years period (1987–2014) were retrospectively analyzed. Echocardiography measurements of left ventricle (LV) and mitral valve (MV) function preoperatively, at discharge, at 3 and 12 months after operation and at last visit were collected. The results in two groups – survivals and nonsurvivals were analyzed and compared. Data are presented as medians with range or means \pm standard deviation. A parametric paired samples T-test integrated in the statistical software SPSS v.19 was used. A value of $p \leq 0,05$ was considered significant.

Results: For a period of 27 years 26 children underwent surgery for ALCAPA. Median age at surgery was 5,5 months (range 0–111 months). All patients underwent establishment of a two-coronary circulation by direct reimplantation ($n = 16$) or by intrapulmonary tunnel technique ($n = 10$), with concomitant mitral valve repair in two cases. 6 patients died. The age of nonsurvivors was lower than survivors: 3,5 (2–6) versus 9 (0–111) months, $p = 0,05$. All nonsurvivors had moderate or severe mitral regurgitation (MR) preoperatively and higher LV diastolic diameter (LVDD) z score than survivors: 11,3 (9–14,6) versus 6 (1,9–13,1), $p = 0,003$. Median follow up was 79 months (4–178). At last follow up, all survivors were asymptomatic with LVDD z-score near normal: 0,3 (0–2,4) versus 6,8 (1,9–14,6) preoperatively $p = 0,001$; and normal LV EF: 66% (61–78) versus 40,5% (16–70) $p = 0,001$. 22 patients had moderate or severe mitral regurgitation at initial presentation and it eventually regressed to non significant in all survivors $p = 0,001$. No subsequent interventions on the coronary arteries or mitral valve were needed.

Conclusion: In our institution operative treatment of ALCAPA with establishment of two coronary circulation without MV repair leads to normalization of LV dimensions and systolic function and improvement of MR in surviving patients. Mortality is related to low age and associated higher degree of LV dysfunction.

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Lung Ultrasound: A New Toll for the Diagnosis of Retro-Sternal Clots in Children Undergoing Cardiac Surgery

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Background: Lung ultrasound (LUS) is gaining consensus as a useful tool for detecting many pulmonary abnormalities; however, its use in pediatric cardiac surgery remains extremely limited.

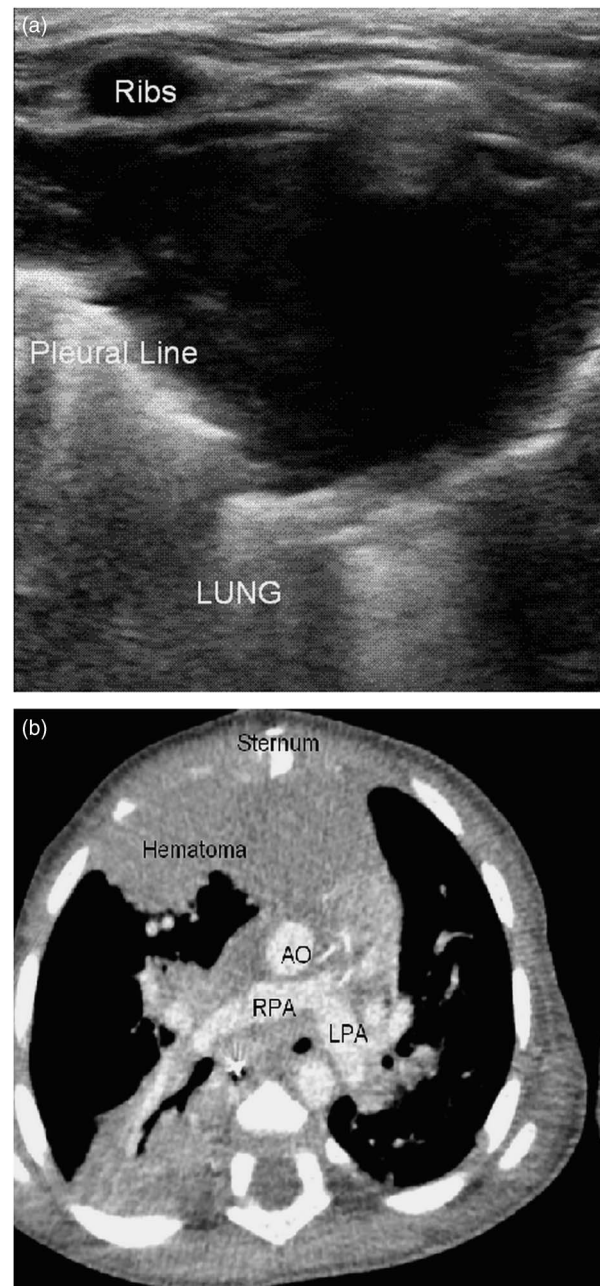


Figure. Case-1. A large (40×25 mm) retro-sternal clot, mainly located in the right hemithorax with no clear relationship with heart is visualized at LUS and was confirmed at CT scan (B). The clot is delineated superior by the sternum/ribs and inferior by the pleural line.

Methods: From January 2014 to October 2014, 270 LUS examinations has been performed in 85 children and young adults (mean age 24.8 ± 73 months) after cardiac surgery for congenital heart disease at a single Center. LUS was performed with a linear 9 MHz probe (Philip IE-33) following the 8-zone scheme. At the beginning LUS was performed to evaluate common pulmonary complications after cardiac surgery (i.e. pleural effusion, diaphragmatic paralysis, pneumothorax). With time, particularly after the first case of incidental diagnosis of retro-sternal clot, we extended to the evaluation of retro-sternal clots/hematomas.

Results: Retro-sternal clots were diagnosed in 21 (25%) at different times after surgery.

In 2 cases clots were of large size squeezing the lung and causing difficult to extubation and required surgical revision. In 6 cases clots were moderate to large causing a mild compression to the heart and/or lungs. Since patients were only mildly symptomatic we decided on echographic follow-up alone and a reduction of clots was noted. In the remaining cases, the majority of our serie (70%), clots were small with no haemodynamic consequences. All clots with no heart compression, although voluminous, were not diagnosed at trans-thoracic echocardiography, and at x-Ray they were misinterpreted as atelectasis/effusion. In cases with heart compression the presence of clots was usually suspected at trans-thoracic echocardiography, but only LUS allowed to clearly define their nature, the spatial relationship and linear quantification.

Conclusions: LUS may lead to a new diagnosis of unknown retro-sternal clots as well as a better definition of those incidentally detected at echocardiography, potentially leading to less need for more complex, ionizing and expensive examinations. Prospective studies are needed to clarify the potential/limitations of LUS in pediatric cardiac surgery.

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Total Anomalous Pulmonary Venous connection combined with Congenital Diaphragmatic Hernia and Left Lung Hypoplasia

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Introduction: Total anomalous pulmonary venous connection (TAPVC) with congenital diaphragmatic hernia (CDH) and left lung hypoplasia is a rare disease entity. Severe respiratory compromise will happen after birth. Here, we reported a newborn case with these complex problems successfully treated surgically. Currently, over 13 years after surgery, the patient is a teenager now and is in functional class I of New York heart association.

Method: A male newborn, body weight 2.2 kg, suffered from respiratory distress with desaturation after birth. Endotracheal tube was inserted and the ventilation was kept acceptably by the effort of pediatric doctors. Chest x ray showed haziness of left lung field where bowel gas was noted. CDH was diagnosed. After general condition was stabilized, repair of left hemi-diaphragm was performed at 4 days of age. During surgery, a left posterolateral defect of diaphragm about 5 cm times 3 cm was noted. The abdominal organ was pull down. The defect was repaired with prolene suture continuously. However, after surgery, the patient was still desaturated. We arranged echocardiography which showed cardiac type of TAPVC, atrial septal defect, patent ductus arteriosus (PDA) and hypoplastic left pulmonary artery (LPA). We arrange cardiac repair for him at 7 days of age. TAPVC repair with PDA ligation were performed smoothly. Because the LPA was very diminutive, we could not do anything for that. Sternum was approximated immediately after surgery. This patient recovered uneventfully. When he was 1 year old, he received another surgery for pyloric stenosis and adhesion ileus.

Result: Currently, he is over 13 years old with good activity and is in functional class I of New York heart association and is pinkish. CT scan was followed up 13 years after repair of CDH and TAPVC and showed growth of LPA from diminutive size to 4 mm in diameter.

Conclusion: TAPVC with CDH and left lung hypoplasia is a rare disease entity with a very critical condition. Surgery is the only treatment of choice. If the patient can stand the surgeries well, the long-term survival can be anticipated.

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Impact of postoperative hemodynamics on cerebral blood flow, microcirculation and oxygen metabolism in neonates and infants after repair or neonatal palliation of complex congenital heart disease

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Objectives: Cerebral protection is a major issue in the perioperative treatment of infants with complex congenital heart defects (CHD) or functional univentricular heart (UVH). Combined laser-Doppler spectroscopy and photo-spectrometry allows transcranial monitoring of regional cerebral oxygen saturation (cSO₂) and relative cerebral blood flow (rcFlow). The aim of this study was to analyse differences in cerebral blood flow (CBF), cerebral microcirculation (rcFlow) and cerebral oxygen metabolism (CMRO₂) after biventricular repair or palliative surgery.

Methods: In 43 neonates and infants after repair of biventricular CHD (n = 30) or neonatal palliation of UVH (n = 13) cSO₂, rcHb and rcFlow were measured 24-36 hours after surgery. CBF was assessed by duplex sonography. The amount of diastolic run-off (DRO) was calculated by a quotient of systolic and diastolic blood pressure. Cerebral fractional tissue oxygen extraction (cFTOE) was determined. CMRO₂ was calculated from CBF and approximated CMRO₂ (aCMRO₂) was calculated from rcFlow.
Results: cSO₂ (54.64% (35.67-64.02) vs 59.72% (44.47-81.70); p = 0.000325), CBF (20.55 ml/100 g/min (9.70-38.60) vs 29.40 ml/100 g/min (9.10-54.00); p = 0.002) and SaO₂ (81.70% (71.70-91.70) vs 98.00% (91.40-99.50); p = 0.000000) were significantly lower after neonatal palliation of UVH compared to biventricular repair of complex CHD, whereas Hb (14.95 g/dl (10.50-17.00) vs 12.10 g/dl (9.00-17.30); p = 0.000001) was significantly higher in patients after neonatal palliation. rcFlow (69.72 AU (42.49-165.25) vs 76.96 AU (41.23-168.12); p = 0.064339), cFTOE (0.34 (0.24-0.82) vs 0.38 (0.17-0.55); p = 0.627186) showed no significant difference. aCMRO₂ was significantly lower in patients after neonatal palliation compared to biventricular repair (3.96 AU (2.07-6.31) vs 4.93 AU (2.15-15.62); p = 0.017792). CMRO₂ showed a trend towards lower values after neonatal palliation (1.59 ml/100 g/min (0.64-3.05) vs 1.79 ml/100 g/min (0.24-3.17); p = 0.214227). There was a significant negative correlation between cSO₂ and CBF and DRO (p = 0.000702; p = 0.044526).

Conclusions: Cerebral oxygen saturation, CBF and aCMRO₂ are significantly lower in patients with UVH after neonatal palliation as compared to children following biventricular repair. These findings provide an explanation for the increased cerebral vulnerability to minor changes in the hemodynamics in infants with UVH.

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Abstract withdrawn

P-254

Meta-Analysis of Carvedilol versus Conventional Treatment in Children with Systemic Ventricle Systolic Dysfunction

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Introduction: Numerous randomized clinical trials in adults with chronic heart failure demonstrated favorable effect of beta-blockers. Treatment in children with systemic ventricle systolic dysfunction includes beta-blockers (carvedilol), although benefit was not recognized sufficiently (mainly due to limited number of pediatric patients).

Methods: We performed meta-analysis, aimed to compare carvedilol and conventional treatment (i.e. digoxin, diuretics, ACE inhibitors) with respect to clinical and echocardiographic outcome in children with chronic heart failure due to impaired systemic ventricle systolic function. We have systematically searched the Medline/PubMed and Cochrane Library for the prospective/observational clinical trials on carvedilol and standard treatment efficacy in pediatric (up to 18 years) heart failure patients. Mean differences for continuous variables, odds ratios for dichotomous outcomes, heterogeneity between studies and publication bias were calculated using Cochrane Review Manager (Rev Man 5.2).

Results: After screening of 391 publications, total of 8 prospective/observational studies (with 516 patients) met established criteria. At the start of studies, there were no differences in the left ventricular end-diastolic diameter ($p = 0.17$), ejection fraction ($p = 0.99$) and fractional shortening ($p = 0.30$) between groups. However, at the end of studies, significantly better ejection fraction (difference 5.2%; 95% CI: 2.4–7.9%; $p = 0.0003$) and fractional shortening (difference 3.4%; 95% CI: 1.7–5.1%; $p < 0.0001$) was demonstrated in the carvedilol vs. control group. Decrease in the end-diastolic diameter was better during the carvedilol treatment, but without statistical significance. Odds ratio for chronic heart failure related mortality/heart transplantation secondary to carvedilol was 0.52 (95% CI: 0.28–0.97; $p = 0.04$), with non-significant heterogeneity between studies and no impact of publication bias. Our analysis showed that carvedilol could prevent 1 death/heart transplantation by treating 14 pediatric patients with impaired systemic ventricle systolic function. In addition, odds ratios for clinical improvement and worsened clinical outcome secondary to carvedilol were 1.54 (95% CI: 0.98–2.43; $p = 0.06$) and 0.60 (95% CI: 0.36–1.00; $p = 0.05$) respectively.

Conclusions: To the best of our knowledge, our meta-analysis demonstrated, for the first time, clinical outcome and mortality/heart transplantation benefits in carvedilol vs. conventional treatment group of pediatric patients with chronic heart failure.

P-255

Clinical characteristics and outcomes of cardiac disease in Barth Syndrome: the 10 year UK experience

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Objective: Barth syndrome (BTHS) is an X-linked disorder characterized by cardiomyopathy, neutropenia, skeletal myopathy, and growth delay. This study aims to describe our clinical experience and outcome of cardiac disease in BTHS.

Methods: A retrospective study was performed to evaluate the cardiac status in all patients with BTHS in the UK over a 10 year period. Data collected included demographics, clinical course, echocardiographic and electrographic parameters. Serial echocardiographic data were analysed to assess trend in myocardial function and morphology. Strain analysis was performed during the most recent echocardiogram.

Results: 26 patients were included in the study. The median age of the 22 surviving patients at the time of evaluation was 12.6 years (range 2.0 to 23.8 years). Seven patients underwent cardiac transplantation at a median age of 3.6 years (range 0.33 to 11.9

years) and 6 recipients are alive at last follow up with good cardiac function. Four patients died at a median age of 2.13 years (range 0.67–4.22 years), and all deaths were related to cardiomyopathy. The 5 year survival rate was 85%, and no deaths were observed after 5 years of age. 92% (24/26) of patients presented with cardiomyopathy; whilst one had profound lactic acidosis and another had a positive family history of BTHS. Left ventricular diastolic dimension and systolic function measured by fractional shortening tended to normalise and stabilise after the first 3 years of life in the majority of patients. However, patients with BTHS ($n = 16$) had statistically significant reduction in global longitudinal and circumferential strain compared to controls ($n = 18$), $p < 0.001$, despite normal fractional shortening and ejection fraction. There was also significantly reduced apical rotation or reversed apical rotation, $p = 0.001$; and reduced LV torsion, $p = 0.046$, in the patient group.

Conclusion: There is a broad spectrum of cardiac phenotype in BTHS, however longer term outcome in our cohort suggest good prognosis after the first 5 years of life, with the majority of patients remaining asymptomatic and recovering normal cardiac function by conventional echocardiographic measures independent of heart failure medication. However, strain imaging may provide valuable insight into the abnormal myocardial mechanics in Barth cardiomyopathy and potentially aid management.

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A neonatal case of myocarditis associated with Campylobacter jejuni enteritis

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Introduction: In Western countries, bacterial causes of myocarditis are uncommon. Case reports in adult population are rare. To the best of our knowledge no neonatal cases or treatment guidelines are available. We present our experience in a neonate who developed a myocarditis, associated with a *Campylobacter jejuni* enteritis and bacteraemia.

Case: Six days after the onset of acute diarrhoea, a three-week-old, previously healthy female neonate presented with tachypnoea and feeding difficulties. At the emergency department she developed a cardiorespiratory arrest. Advanced cardiac life support was initiated, with on-going need for inotropic support at the intensive care. A wide spectrum antibiotic treatment was administered. A positive blood culture confirmed bacteraemia with *Campylobacter jejuni*. Transthoracic echocardiography revealed a dilated left ventricle with poor contractility. Holter monitoring revealed numerous runs of atrial tachycardia. Clinical diagnosis of bacterial myocarditis was based upon signs of heart failure combined with left ventricular dilatation, leucocytosis, arrhythmia and a positive *Campylobacter jejuni* blood culture. Upon early clinical and echocardiographic recovery, within a few days since her admission, a relapse occurred. Inotropic support was mandatory over a period of 2 months, without any signs of recovery. High dose and long-term intravenous antibiotics were administered. Forty days upon admission, immunoglobulin therapy at 1 g/kg/day was initiated during three days. Gradually inotropic support could be weaned successfully. Further treatment included an angiotensin converting enzyme inhibitor and a loop diuretic. One year after the event she recovered a normal heart function.

Conclusions: Underlying pathophysiology of myocarditis caused by *Campylobacter jejuni* is unclear. The time interval between the enteritis and the initial cardiac involvement was short, suggesting an initial direct myocardial injury and supporting our choice for prolonged aggressive antibiotic treatment. This myocarditis seemed to have a

biphasic course. One possible explanation could be an immune mediated relapse and although immunoglobulin treatment was not implicated in other cases of bacterial myocarditis and their benefit is not proven in viral myocarditis, our patient recovered gradually after administration. Natural disease course cannot be excluded. Despite prolonged inotropic support, complete recovery occurred.

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Maternal derived anti SS-A antibody as a possible etiology of secondary dilated cardiomyopathy associated with congenital left bundle branch block

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Introduction: Maternal derived anti SS-A antibody affects the fetal heart by initiating a series of events leading to cardiac inflammation, fibrosis and calcification, and eventual blockage of signal conduction at the atrio-ventricular node. Herein we report two infants with dilated cardiomyopathy with left bundle branch block (LBBB) associated with maternal derived anti SS-A antibody in whom clinical status dramatically improved after cardiac re-synchronization therapy (CRT).

Methods: We present two cases of infantile cardiomyopathy with LBBB.

Results: Case 1; A 5 months-old baby was addressed with severe heart failure and a 1 month history of poor feeding and failure to thrive (body weight; 4 kg). He was referred to our hospital after the medical treatment of acute heart failure for further management of cardiomyopathy. The electrocardiography (ECG) revealed LBBB pattern with QRS duration of 140 ms and the echocardiography showed a typical septal flash (Fig. 1). He underwent CRT pacemaker implantation by thoracotomy. 7 weeks after the CRT, B-type natriuretic peptide decreased to 38 from 2,200 pg/ml and left ventricular ejection fraction improved to 55 from 19%. His ECG at the age of 7 days of life also revealed LBBB pattern with QRS duration of 100 ms. The titer of the anti SS-A antibody was 63 U/ml, and his mother's titer was over 10,000 U/ml. Case 2; A 3 months baby was referred to our hospital after initial therapy for cardiogenic shock. A laboratory data revealed metabolic acidemia and disseminated intravascular coagulation (DIC). The ECG revealed LBBB pattern with QRS duration of 120 ms and the echocardiography showed a typical septal flash (Fig 1). She underwent CRT pacemaker implantation by thoracotomy after break away from DIC. After CRT, we successfully weaned off the

mechanical ventilation. And now we started medical treatment for chronic heart failure. The titer of the anti SS-A antibody was 299 U/ml, and her mother's titer was over 10,000 U/ml.

Conclusions: Congenital LBBB associated with maternal derived anti SS-A antibody may cause mechanical ventricular dyssynchrony and heart failure. That means the electrical and mechanical re-synchronization of the ventricle improves clinical status as well as cardiac function in these infants.

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Dilated Cardiomyopathy in hospitalized children - a single center experience from Poland

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Background: Dilated cardiomyopathy (DCM) is one of the most common causes of the heart failure in children and one of leading reasons for the heart transplantation in this population. The prevalence of DCM in Polish children remains unknown and a national-based registry of DCM does not exist so far.

Methods: A retrospective study of 88 children and youth (41% of girls and 59% of boys) admitted with confirmed DCM from 2008 to 2014 to a single center was performed. Patients' median age on the first admission was 2 years (2 months - 17 years). The diagnosis was made on a basis of echocardiographical findings in all patients. In a further evaluation a computed tomography was performed in 8 patients and a magnetic resonance imaging in 5. Etiology, administered treatment and mortality were evaluated.

Results: Idiopathic DCM was diagnosed in the majority of cases (28 patients; 31,82%). Among identified causes of DCM there were left ventricular noncompaction (NCLV) (confirmed in 21 patients, including 3 with Barth syndrome - 23,86% and under evaluation in another 13 - 14,77%), myocarditis (18 patients; 20,45%), myocardial infarction (4 patients; 4,55%), arrhythmia (3 patients; 3,41%) and drugs (1 patient; 1,14%). 87 patients were treated with at least one pharmacological agent. ACE inhibitor was administered in 75 patients (85,22%), β -blocker in 77 (87,50%), steroidal antimetabolocorticoid in 79 (89,72%), diuretic in 47 (53,40%). A group of 70 patients (79,55%) received combined treatment with ACE inhibitor and β -blocker. 30 patients (34,09%) were listed for the transplantation, including 8 with NCLV (26,66%). 15 children (17,04%) underwent heart transplantation, 7 of them (46,66%) required mechanical circulatory support as a bridge. Overall mortality was 9,09% (8 patients, including 1 after heart transplantation). Our research as a retrospective and single-centre has some biases.

Conclusion: The number of patients with NCLV as a cause of DCM in presented data is considerable (over 20%). Despite the optimal pharmacological treatment the quick progression to the end-stage heart failure was often observed in the studied group. Further research to assess morbidity and prognostic factors in DCM is necessary.

P-259

The perception of self-efficacy in a collective culture: adolescents and young adults with corrected congenital heart defects choosing a career in Bolivia

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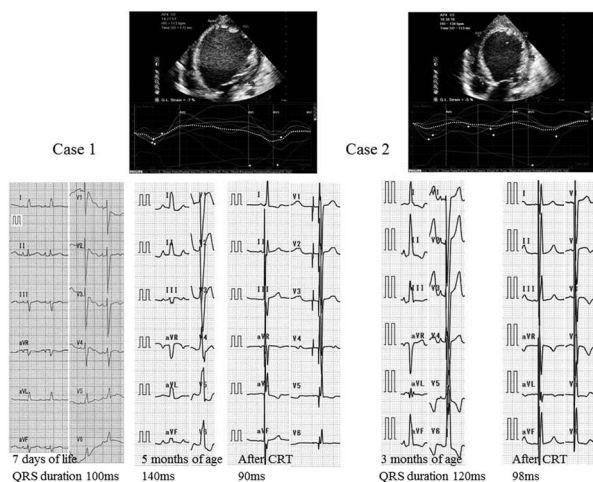


Figure 1.

Introduction: The aim of this study is to identify specific cultural beliefs in young Bolivian patients with congenital heart defects (CHD), which influence the perceived self-efficacy to cope with their condition, as they choose a career.

Bolivia is inhabited mostly by Quechua (45.6%) and Aymara (42.4%) populations, who practice collective traditions and whose beliefs include expressing external locus of control, magical thoughts, ritualistic offerings, and faith in the devil as well in Mother Earth. Aymaras developed a hybrid identity and culture as a response to domination by the Incas, then by the Spaniards, and later by authoritarianism. Nowadays globalization, economic growth, media acculturation and migratory processes are transforming values, habits and identities of the natives. The hegemony of individualism over collectivism seems to be developing in the cities.

Methods: Follow-up study with three testing phases, using descriptive and non-parametric methods in a qualitative and quantitative design. The sample is intentional and includes 21 patients (16–31 years of age) from Kardiozentrum in La Paz.

Results: The study reveals that 38% of the patients show discontentment with their health and career development, including shortages in self-regulation, coping and self-efficacy or the sense of personal competence to deal effectively with a variety of stressful situations. Almost 28% of patients denote a strong goal-orientation and an active attitude in their self-management. The IQ-score media from 16 CHD patients was 85 points, but this lack of thought fluency does not seem to be notoriety in a collective society.

Conclusions: Observed patients in a collectivist society show lower scores in perceived self-efficacy than patients socialized in individualist societies. Cultural beliefs, traditions and practices should be recognized and their value considered for the purpose of activating self-efficacy and self-management in the development of a tailored-made career program.

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Health-Related Quality of Life in Children with Surgical Therapy of Congenital Heart Disease

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Background: Although survival of children with congenital heart disease (CHD) has improved significantly, their health-related quality of life (HRQoL) is less inquired, particularly in relation to surgical therapy. Since 2009, HRQoL has been assessed in part with DISABKIDS questionnaire (DCMG12), and recorded in the Swedish national registry of congenital heart disease (SWEDCON).

Methods: A retrospective SWEDCON survey of 365 children (147 girls and 218 boys; age 9–18 years). The majority (n = 345) had biventricular heart (BVH), while the remaining (n = 21) had univentricular heart (UVH). Cardiac surgery ± catheter therapy was pursued in 218 children. NYHA class and indices of cognitive function (learning ability) were available in 359 children. HRQoL from DISABKIDS was expressed as total score (TS; range from 0 to 100 highest). TS data are given as median/interquartile range (25–75 percentiles). Kruskal Wallis or Mann-Whitney test, when appropriate, were used as statistical analyses.

Results: DISABKIDS was answered by 340 children. In the whole cohort, TS was high (95/88–100) without any difference in gender (p = 0.78) or age (p = 0.1). There was no difference in TS between BVH and UVH children (p = 0.57). In the BVH cohort, TS was lower in those with previous surgery than in those without (95/83–100 and 98/90–100 respectively; p = 0.008). Children with ≥ 3 surgeries (n = 32) had the lowest TS (81/69–93; p < 0.001), needed more often aid in school (p = 0.024) and had more

frequently NYHA ≥ 2 (p = 0.006 vs NYHA 1). As much as 96% children without surgery (n = 138) attended regular school and were classified in NYHA1. In those with surgery (n = 154), 79% attended regular school and 81% were in NYHA 1 (p < 0.001 versus nonsurgical cohort).

Conclusion: Overall, children with biventricular CHD have high HRQoL. Cardiac surgeries appear to have cumulative adverse effect on HRQoL, learning ability and NYHA. Further larger studies are warranted to validate these findings and to assess the efficacy of putative interventions in improving HRQoL in this cohort.

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Increase of Frequency in Attention Deficit/Hyperactivity Disorder in Children with Rheumatic Fever:

Preliminary Study

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Introduction: Genetically, susceptible children who are infected by Group A β-hemolytic streptococci may develop a systemic autoimmune disorder known as rheumatic fever (RF). Several psychiatric disorders, in particular obsessive-compulsive spectrum disorders (OCD), attention deficit/hyperactivity disorder (ADHD), and tic disorders have been described in RF. This study aims to examine neuropsychiatric symptoms which are determined in pediatric patients with RF.

Methods: Thirty consecutive patients with RF (mean age 12.8 ± 1.9 (9–16) years; M/F; 16/14) were studied for a structured psychiatric evaluation according to DSM-IV-TR by a child and adolescent psychiatrist. Also, Yale-Brown obsessive compulsive scale (Y-BOCS), Yale global tic severity scale (YGTSS), and Turgay Attention-Deficit/Hyperactivity Disorder (ADD/ADHD) DSM-IV-Based Diagnostic Screening and Rating Scale were performed to the patients.

Results: According to DSM-IV-TR criteria, 20 (66.6%) cases had at least one psychiatric disorder. The most frequent diagnosis by the cases is found to be ADHD (n = 16). Other diagnoses were OCD (n = 4), anxiety disorder (n = 4), tic disorder (n = 3), and enuresis nocturna (n = 1). Parents have declared fewer ADHD symptoms (n = 6) by using the ADD/ADHD DSM-IV-Based Diagnostic Screening and Rating Scale than the clinician as usual in the literature. The mean total score of Y-BOCS was 2.3 ± 6.2; the mean obsession subscale score was 1.3 ± 3.6 and the mean compulsion subscale score was 0.96 ± 3.6. The mean score of YGTSS was 0.5 ± 2.2 in our study group.

Conclusions: On the basis of our results, especially ADHD symptoms seem to be more frequent in patients with RF. Thus, clinicians should not only deal with the treatment of RF but also be aware of the neuropsychiatric manifestations for early diagnosis.

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Quality of life of children with congenital heart diseases: a multi-center controlled cross-sectional study

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Objectives: To assess health-related quality of life (QoL) in children with congenital heart diseases (CHD) with a validated questionnaire in comparison with control children.

Methods and Results: we prospectively recruited 282 children with CHD aged 8 to 18 in two tertiary care centers (France and Belgium) and 180 same-age controls in randomly selected French schools. Children's QoL was self-reported with the Kidscreen-52 questionnaire and reported by parents with the Kidscreen-27. QoL scores of each dimension were compared between CHD and controls and between the classes of disease severity defined by Uzark et al.

Both centers were comparable for most demographic and clinical data. Age- and gender-adjusted self-reported QoL scores did not differ between CHD children and controls except for physical well-being (mean \pm SEM: 45.97 \pm 0.57 vs 50.16 \pm 0.71, $p < 0.0001$), financial resources (45.72 \pm 0.70 vs 48.85 \pm 0.87, $p = 0.01$) and peers/social support (48.01 \pm 0.72 vs 51.02 \pm 0.88, $p = 0.01$). Parent-reported scores were lower in CHD children for physical ($p < 0.0001$), psychological well-being ($p = 0.04$), peers/social support ($p < 0.0001$) and school environment ($p < 0.0001$). Similarly, the severity of the disease had an impact on physical well-being ($p < 0.001$), financial resources ($p = 0.05$) and peers/social support ($p = 0.01$) for the self-reported dimensions, and on physical well-being ($p < 0.001$), psychological well-being ($p < 0.01$), peers/social support ($p < 0.001$) and school environment ($p < 0.001$) for the parent-reported dimensions. However, in multivariate analysis, disease severity was not significantly associated with self-reported QoL.

Conclusions: Self-reported QoL of children with CHD was similar to that of same-age healthy children in 7 out of 10 dimensions but parents-reported QoL was impaired in 4 out of 5 dimensions.

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What are Psychosocial Needs of Children and Adolescents undergoing an Invasive Procedure for a Congenital Heart Defect and their Parents? A Cross-sectional Cohort study

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Objective: The aim of the study was to investigate the psychosocial needs of both parents of children with congenital heart disease (age range children 0-18 years) and the patients themselves (age range 8-18 years) in the week before cardiac surgery or a catheter intervention.

Methods: Inclusion: eligible were all consecutive patients (0-18 years), scheduled to undergo cardiac surgery or a catheter intervention in our hospital between March 2012 and July 2013. Psychosocial needs considering five specific domains (physical/medical, emotional, social, educational/occupational, health behaviour) were measured by a disease specific questionnaire. Additionally, parents and patients were asked from whom and in what format they would prefer to receive psychosocial care. Quality of Life (QoL) was also assessed.

Interventions: if parents and/or patients reported a need for psychosocial care, referral to health care professionals in or beyond our hospital was performed.

Results: More than 40% of participating parents and more than 50% of participating children reported a need for psychosocial care on each of the five domains. Needs for psychosocial care for parents themselves were highest for those with children aged 0-12 years. Parents and patients report clear preferences when asked from whom and in what format they would like to receive psychosocial care. QoL was relatively high for both parents and patients. Psychosocial care interventions in our hospital increased significantly due to the implementation of this study.

Conclusions: Results show that psychosocial care is rated as (very) important by both parents and children during an extremely stressful period of their life.

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Uhl's anomaly associated with pulmonary atresia intact ventricular septum: Report of two cases

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Introduction: Uhl's anomaly is rarely encountered anomaly. Absence of right ventricular myocardium may be the result of primary non-development of myocytes or may be due to selective apoptosis. Uhl considered the disease to be congenital in origin, since then, there have been numerous similar case reports of apparently congenital hypoplasia of the entire or near-entire right ventricle, with or without other associated congenital heart defects.

Case 1: The premature neonate was born at 35 4/7 weeks gestation and weighed 1510 g. His initial oxygen saturation was 65%. Echocardiography revealed enlargement of right atrium and ventricle, severe tricuspid insufficiency, and thin musculature of right ventricle and intact ventricular septum. Pulmonary antegrade blood flow could not be detected. It was provided by ductus arteriosus (Figure 1). The condition of the infant continued to deteriorate. Recurrent hypotension and severe cyanosis required volume and dopamine, prostaglandin infusions. The patient died at the two hours of life due to intractable heart failure.

Case 2: The second term neonate was born 3120 g. with oxygen saturation of 75%. Echocardiography revealed intact ventricular septum with pulmonary atresia associated with enlargement of right atrium and ventricle, severe tricuspid insufficiency with 2.2 m/sec velocity, and thin musculature of right ventricle.

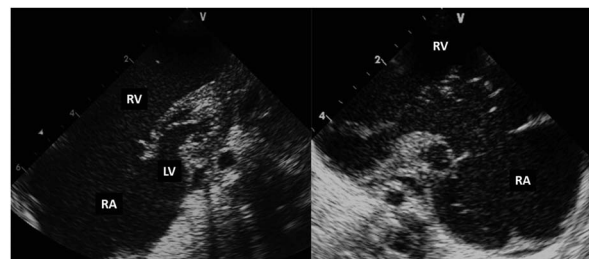


Figure 1. Echocardiographic examination of patient. Massive right ventricular and atrial dilatation, thin muscular layer of right ventricle. (RV; right ventricle, RA; right atrium)

Anjiography was performed for ductal stenting but unfortunately could not be performed due to ductal position.

Conclusion: Dysplasia of tricuspid valve, pulmonary atresia with intact ventricular septum have been described in association with Uhl's anomaly. The necropsy findings of a case of Uhl's anomaly associated with pulmonary atresia in a newborn was reported in the literature as an important reduction in the number of muscular fibers and a very thin right ventricular wall as described by Uhl. The prognosis of the complete form of Uhl's anomaly associated with pulmonary atresia with intact ventricular septum is poor as in these cases.

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Growth-differentiation factor-15 and tissue Doppler imaging in detection of anthracycline induced cardiomyopathy in during therapy of childhood cancers

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Objectives: The aim of this study was to evaluate the importance of growth-differentiation factor-15 (GDF-15) levels and tissue Doppler imaging (TDI) in the early detection of anthracycline-induced cardiomyopathy during the treatment of childhood cancers.

Patients and Methods: Twenty patients (13 males and 7 females) newly diagnosed with childhood cancer whose treatment protocol included anthracycline were included in the study. Echocardiography, including M-mode, pulse Doppler and TDI, was performed after the first anthracycline treatment at cumulative doses of 100, 200 and 300 mg/m² and at least 6 months after the last treatment. Growth-differentiation factor (GDF-15) and troponin-I were also measured at these time points.

Results: The median age of the patients was 14 years (range, 3–18 years). The median cumulative anthracycline dose was 220 mg/m² (range, 60–400 mg/m²). Conventional pulse wave and pulse wave tissue Doppler methods revealed significant differences in the right ventricular myocardial performance indices of the patients who received cumulative anthracycline doses of 300 mg/m² compared to their indices at least 6 months after the last treatment. The serum GDF-15 levels after the cumulative anthracycline dose of 200 mg/m² were also higher than the patients' pre-treatment levels.

Conclusion: Doppler/TDI and GDF-15 levels may be used in the early determination of anthracycline-induced cardiomyopathy during the treatment of childhood cancers.

Key words: anthracycline cardiotoxicity; tissue Doppler imaging; growth-differentiation factor-15; children.

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Cardiovascular risk factors in portuguese children - where did Mediterranean lifestyle go?

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Introduction: It is currently now well known that Mediterranean diet and lifestyle (recognized by UNESCO since last December 2013) are both protective from dyslipidemia, overweight, obesity and high blood pressure. However, in Portugal cardiovascular risk factors seem to be an emerging public health problem.

Aim: To study the pediatric population in a tertiary pediatric cardiology centre, in order to assess current antropometry, lifestyle and cardiovascular risk factors in patients. Also we assessed cardiovascular risk in first degree relatives.

Methods: Transversal study was conducted, through chart review from the outpatient clinic of Cardiovascular Risk.

Results: We have classified the population in younger or older than 12 years-old. The total number of patients in the last 9 months were 61, 36 females, age from 5 to 20 years-old. Of these, 31 had a first degree relative with at least one cardiovascular risk factor. None had congenital heart disease, 2 were smokers, only 2 had exercise as a hobby. 21 patients had BMI > 25, 10 had total cholesterol above 200 mg/dL, 13 had high levels on fasting glucose (>100 mg/dL), 14 patients had hypertension (class I or II on ambulatory blood pressure measurement) and 4 had diffuse liver steatosis on abdominal ultrasound. At least on one medication was taken in 17 patients (blood pressure lowering agent, statins, metformin). Statistical analysis between these 2 groups will be presented when a larger population is reached.

Discussion: We found that childhood overweight, obesity, high blood pressure, dyslipidemia and diffuse liver steatosis were present in our sample. We also point out the significative amount of first degree relatives with cardiovascular risk factors and the the need for further studies in congenital heart disease patients.

P-267

Management of children and adolescents with familial hypercholesterolaemia in a specialist out-patients clinic

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Introduction: Heterozygous familial hypercholesterolaemia (FH) is a common autosomal-dominant genetic disorder (1:200) with an increased risk of premature coronary artery disease even in the young. Published criteria exist for diagnosis and management of FH¹. We report our findings in paediatric and adolescent patients referred to and treated in the FH Outpatients Clinic.

Methods: We retrospectively reviewed hospital records of patients diagnosed with genetically confirmed FH under 18 years of age in a single tertiary cardiac centre between January 2006 and June 2014.

Results: Forty eight patients (25 male) with FH were identified. Median age at diagnosis was 10.1 (range 2.3 - 17.3) years. Reasons for patients' referral was mainly diagnosis of FH in a relative (79%) and abnormal serum lipid profile in referred patient (8%). Medical records and follow-up data were available in 39 patients. Dietary intervention was commenced in all patients and 29 patients (67%) were started on treatment with a statin at a median age of 12.45 (range 7.6 - 18.6) years. Decrease in total cholesterol (TC) levels was achieved from initial TC (mean (SD)) 7.05 (1.47)mmol/L to 5.45 (1.19)mmol/L at latest review (p=0.000001). Similar decrease in low-density lipoprotein (LDL) values was encountered (initial LDL 5.44 (1.55)mmol/L, at latest review 3.63 (1.08)mmol/L; p<0.0001). No significant changes were seen in high-density lipoprotein (HDL) levels (initial HDL 1.32 (0.35)mmol/L, at latest review 1.34 (0.29)mmol/L; p=0.75).

Conclusions: Dietary and medical treatment of children and adolescents with FH has beneficial effects on lipid profile. As early diagnosis and treatment are essential for long-term prognosis², a detailed family history review focused on hypercholesterolaemia and timely referral to a specialist Outpatients Clinic are mandatory in general paediatric and paediatric cardiology clinics.

References

¹www.nice.org.uk/guidance/cg71 (last review on 10th December 2014).

²Cuchel, M., et al., Homozygous familial hypercholesterolaemia: new insights and guidance for clinicians to improve detection and clinical management. A position paper from the Consensus Panel on Familial Hypercholesterolaemia of the European Atherosclerosis Society. *Eur Heart J*, 2014. 35 (32): 2146-57

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Early Identification of at Risk Population through Service Quality Improvement Project in Paediatric Cardiology Out-patient's Clinic

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Introduction: As part of a wider programme to early identify children at risk of familial hypercholesterolaemia and/or sudden cardiac death we conducted a pilot qualitative study using adapted version of a previously validated questionnaire¹ in the setting of paediatric cardiology out-patient's clinic.

Methods: Parents of 120 patients attending general paediatric out-patient's clinic in a tertiary referral centre between October and December 2014 were asked to fill in a questionnaire.

Results: We received 89 (74%) fully filled questionnaires from 31 newly referred patients (palpitations, dizziness, chest pain, murmur) and 58 follow-up patients (structural congenital heart disease). Clinically important information was received from parents of 19 (61%) new referrals related to family history of high blood cholesterol levels, sudden or premature death and/or cardiomyopathy. Information of similar importance was obtained from 30 (52%) families of follow-up patients. Out of 34 questionnaires (13 from new referrals and 21 from follow-up patients) where family history of hypercholesterolaemia was mentioned, 26 (76%) families satisfied Simon Broome diagnostic criteria² for further evaluation to exclude familial hypercholesterolaemia as per NICE (United Kingdom) guidelines³. New information about family history of sudden death was received in 10 cases (11% of all questionnaires returned).

Conclusions: Our pilot study using an adapted version of a previously validated questionnaire brought up additional potentially important clinical information in the majority of families seen in the setting of general paediatric cardiology out-patient's clinic. Surprisingly high number of families fulfilling criteria published by NICE (United Kingdom)³ for familial hypercholesterolaemia screening were identified. Further evaluation of the suspected at risk families by specialised familial hypercholesterolaemia and sudden cardiac death services is mandatory.

References

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Evaluation of the prevalence of overweight and obesity in teenagers of the metropolitan area of Warsaw

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Objectives: Evaluation of the prevalence of overweight and obesity in adolescents aged 15–17 years in the metropolitan area of Warsaw, Poland.

Methods: The study group consisted of 690 middle and high school students, 366 boys and 324 girls aged 15–17 years, from the metropolitan area of Warsaw, who underwent screening.

Anthropometric measurements included: body weight, height, arm, hip and abdominal circumference. On the basis of taken measurements following indexes were determined: body mass index–BMI, Waist to Hip Ratio–WHR, Waist to Height Ratio–WHtR, Hip to Height Ratio–BAI (Body Adiposity Index). Skin-fold thickness was measured on the rear surface of arm, below the inferior angle of the scapula, and at the belly.

Results: We found that 23.6% of children were overweight or obese, based on BMI. Obesity was diagnosed in 11.3% and overweight in 12.3% of children. These percentages were comparable among boys and girls also in different age groups. Based on the growth charts, obesity was diagnosed in 8.6% (n = 59) and overweight in 9.3% (n = 64) of the study group.

A discrepancy has been noticed between the assessment based on the body weight growth charts and the calculation based on BMI. In 29 (4.2%) patients body weight was assessed as correct based on growth charts, while the assessment on the basis of BMI showed overweight or obesity. Only in 11 (1.6%) patients method based on the growth charts overestimated the assessment.

Very strong correlations have been found for BMI and waist, hip and arm circumference, subscapular and abdominal skinfold thickness, WHtR and BAI – (properly: r = 0.86; r = 0.84; r = 0.88; r = 0.81; r = 0.75; r = 0.88; r = 0.81; p < 0.05).

Conclusions:

1. The prevalence of overweight and obesity in middle and high school students in the analyzed region is alarmingly high.
2. Research methods and standards for body weight categorization on the basis of growth charts and BMI, lead to slightly different epidemiological results.
3. BMI strongly correlates with anthropometric measurements, which justifies its use for the assessment of overweight and obesity in daily practice.

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Prediction of the hypertension risk in teenagers

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Objectives: Creation of a hypertension risk stratification model and development of an algorithm to detect hypertension in teenagers.

Methods: The study group consisted of 690 middle and high school students, aged 15–17 years, from the metropolitan area of Warsaw, Poland.

Information concerning family history and presence of risk factors for cardiovascular disease was gathered.

Three – time blood pressure measurements were taken during at least two separate visits, at least a week apart, using the auscultatory method according to standard procedures. Anthropometric measurements included: body weight, height, arm, hip and abdominal circumference. On the basis of taken measurements: body mass index–BMI, Waist to Hip Ratio–WHR, Waist to Height Ratio–WHtR, Hip to Height Ratio were determined. Skin-fold thickness was measured on the rear surface of arm, below the inferior angle of the scapula, and at the belly.

Results: A logistic regression model describing the risk of hypertension in adolescents aged 15–17 has been invented. The formula has been created, allowing the pre-selection of adolescents at risk of hypertension during screening:

$$\hat{\pi}(x) = \frac{e^{\hat{\beta}(x)}}{1 + e^{\hat{\beta}(x)}}$$

where: $g(x) = -0,09711 * \text{height} + 0,08487 * \text{weight} + 7,76428 * \text{WHR} + 1,31222 * \text{family_hypertens} = \text{yes}$

Based on the created risk model an algorithm for the detection of hypertension for practical use has been proposed:

1. Information campaign at schools.
2. Completion of website form using the developed model to estimate a risk of hypertension.
3. In the case of estimated risk >50% - three-time blood pressure measurements at weekly intervals should be taken.
4. If the arithmetic mean of the second and third measurement is between 90 and 95 percentile, non pharmacological treatment and periodic blood pressure controls are recommended.
5. If it is >95th percentile, further specialized diagnostics is recommended.
6. If the risk is >75% - further evaluation and periodic check up are recommended even if blood pressure values are normal.

Conclusions: The body weight, WHR and incidence of hypertension in the family are the strongest predictors of hypertension in teenagers.

The proposed screening algorithm can be a useful tool for selecting teenagers at risk of hypertension and in need of specialized diagnostics.

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The Relationship between Blood Pressure Variability and Left Ventricular Mass Index in Children with Primary Hypertension

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Introduction: Increased blood pressure variability (BPV) is related to subclinical target organ damage and greater incidence of cardiovascular events in adults. There has been limited data on its influence in children. The aim of this study was to investigate the relationship between 24-hour ambulatory blood pressure monitoring (ABPM) indices of BPV and the presence of left ventricular hypertrophy (LVH) in children and adolescents with primary hypertension.

Methods: A total of 85 children and adolescents aged 7-20 years (mean 14.99 ± 2.4) with office hypertension were evaluated. All patients underwent ABPM (Schiller MT-300) as well as standard echocardiographic examination (Vivid 9, GE). BPV was measured as weighted standard deviation (wBPSD) and average real variability (ARV). We also calculated average 24-hour systolic and diastolic blood pressure (aSBP, aDBP) and 24-hour SBP load. Left ventricular mass index (LVMI) was assessed as an indicator of LVH. Data were analyzed using SPSS 17.0 statistical package.

Results: Of the total number of patients 18% was overweight, 39% obese. Ambulatory prehypertension was found in 30.6% of patients, ambulatory hypertension in 54.1%, white coat hypertension in 15.3%. LVH was found in 36 patients (42.4%), and 7 (8.2%) of them had severe LVH. LVMI significantly correlates with body mass index (BMI) ($r = 0.359$; $p = 0.001$). There was no significant correlation between BMI and 24-hour systolic wBPSD ($r = 0.025$; $p = 0.821$), and 24-hour systolic ARV ($r = 0.024$; $p = 0.83$). We found significant correlation between LVMI and BPV indices: 24-hour systolic ARV ($r = 0.356$; $p = 0.001$) and 24-hour systolic wBPSD ($r = 0.391$; $p < 0.001$). LVMI significantly correlated with aSBP ($r = 0.275$; $p = 0.001$) and 24-hour SBP load ($r = 0.264$; $p = 0.015$). By using multiple regression analysis with LVMI as dependent variable and aSBP, 24-hour SBP load, 24-hour systolic wBPSD, 24-hour systolic ARV and BMI as independent variables, only BMI represented an independent predictor of LVMI ($p < 0.001$).

Conclusions: Blood pressure variability indices significantly correlate with LVMI, but they do not represent reliable parameters for identification of patients with LVH. The main independent predictor of LVH in children and adolescents with primary hypertension was BMI. Overweight children with greater blood pressure variability need closer evaluation for LVH.

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A mid-term results of nurse-aid self-educational program for the child and early adolescents with congenital heart disease in outpatient clinic

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Background: The transitional program from the child to adult with congenital heart disease (CHD) has become an more important issue, to avoid a drop out from medical center and to keep their life in good medical care along their life. We report the impact of a self-educational program on understanding of their own disease and treatment to facilitate the smooth transition to the adult CHD medical center, no matter where the ACHD patients live.

Aim: We evaluate the nurse-aid self-educational program for young adolescents with CHD guided by questionnaire in outpatient clinic.

The subjects were 6214 patients with CHD > 15 years old visited to our outpatient clinic between May in 2011 to November in 2014.

Methods: All patients were once informed from the age of 6 years old about their diagnosis and the content of treatment including the surgery and medications with figures in outpatient clinic. The status of the ACHD patients was assessed by questionnaires regarding to their diagnosis, surgical procedure, hemodynamics, medications, exercise range, the prevention of infective endocarditis, and the issues of pregnancy.

Results: The patients were also requested to draw the figures of their post-surgical state which had been taught by the charged doctor from the elementary school age.

Results: the rate of the correct answers for all major questions was 13.6% (<20 y) and 36.7% (20 y <), while that of no correct answers for all major questions was 14.7% (<20 y) and 16.3% (20 y <). The rate to need further follow-up to ascertain the self-esteem for their medical and social issues changed from 71.5% (<20 y) to 47% (20 y <). After receiving the nurse-aid program, 67% of patients could improve their achievement scores, however there still remained high percentage of those who could not draw their post-surgical figures.

In conclusion, the nurse-aid self-educational program for children and early adolescents with CHD is indispensable to train them to be independent in adult as a transition, however the educational approach needs to improve the self-confident understanding of their own disease.

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Learning through play in a congenital cardiac unit: it's child's play

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Introduction: In our paediatric cardiac surgery unit, anxiety generated by the perspective of surgical act or invasive intervention is a pervasive problem. In order to reduce this anxiety for the child and his family, we have conceived a preoperative learning-through-play protocol.



Figure.

Material and Methods: During dedicated training courses, the paramedical team chose two specific tools to explain to the hospitalised child the process of surgery and heart catheterisation: Playmobil® medical play kits and a doll equipped with perioperative supplies.

Playmobil® kits are used by the childcare assistants at the admissions day. The description of the entire preoperative and post-operative course, from the departure from the unit to the operating room, and then to the intensive care unit (ICU) and afterwards, allows the child to situate her/himself in time and space, to identify the specific roles of each member of the team and to delimit the time between parental separation and reunion.

The equipped doll is used by the nurses the day before surgery in order to show the different medical devices, which will be implanted in the operating room. The intravenous lines and chest tubes are presented in real size, their utility and their temporary nature are emphasised. The child can handle the devices, play with them, checking for instance that there is no needle in the venous lines. The doll also helps the nurse to explain the surgical incision and the management of pain.

Results: Since February 2012, 263 children (3 - 11 years old) have been informed with Playmobil® kits and 75 with the equipped doll. The feedbacks of the families, who were sometimes initially sceptical, were globally positive, as assessed by satisfaction questionnaires. Medical and paramedical teams in the operating room and in ICU have noticed a dramatic decrease in anxiety in informed children.

Conclusion: The systematic use of these tools enabled us to place the child at the center of the perioperative care. This learning-through-play approach allows us to create a trust relationship with the child and his family, and reinforces our determination to improve the quality of care in young patients with congenital heart defects.

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Improving breastfeeding services for neonates with Congenital Heart Defect using the Experience-Based Co-Design approach

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Rationale: Previous studies have shown that breastfeeding benefits infants with Congenital Heart Defect (CHD). This thesis studies

the trajectory families undergo to breastfeed their infants. It aims to understand the challenges families and health professionals experience to deliver high quality breastfeeding services, alongside the improvements required to make the breastfeeding journey successful. Additionally, this project also aims to understand if the Experience-Based Co-Design (EBCD) methodology was a suitable approach to sustain quality improvement developments within the paediatric setting.

Methodology: The study was performed using the EBCD methodology. Fieldwork involved 25 filmed interviews of families and staff members. During the course of the project three events were organised where participants gave feedback on their experiences. Necessary improvements to the breastfeeding services provided were also discussed by participants. The project setting was at The Royal Brompton Hospital (RBH), a Paediatric cardiac unit in London, England.

Results: The experiences reported confirm that breastfeeding a cardiac infant is challenging and tailored support is required. "Education for staff", "Family support & well-being" alongside "Communication" are the main themes needed to improve breastfeeding journeys at the RBH. Furthermore, respondents described EBCD as a successful and sustainable method of improving the individual family experience and the overall quality of the health service.

Conclusion: A policy/guideline for breastfeeding cardiac infants needs to be developed for high quality care to be attained. For this document to be developed improvements, primordially around efficient education and investment in human resources, in cardiac units is required. Regarding the EBCD methodology, the primary strength noted was the improvements immediately reflected on the unit. Nevertheless, time constrains and lack of human resources to adequately develop the methodology can portray a challenge.

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First UK multicenter study to review the Efficacy of Nutritional Treatment Strategies in the Management of Post-cardiothoracic surgical chylothorax

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Background:

- Incidence of chylothorax has increased 0.9%-1.5% to 6.6%
- No agreement regarding efficacy of Long Chain Triglyceride (LCT) reduction in reducing chyloous output

Aim:

1 - Primary aim:

Investigate daily LCT intakes throughout the entire course of diet therapy and compare against the nationally accepted guidelines.

2 - Secondary Aims:

To investigate the efficacy of diet therapy changes and reducing LCT intakes upon drain output.

To compare participating centre guidelines

Methods: A multi-centre prospective observational study conducted between January 2013 and January 2014 reviewing routine diet therapy of paediatric congenital heart patients diagnosed with chylothorax. Daily LCT intakes and drain outputs were recorded. Regression analysis investigated relationships between feed type and drain output, the effect of age and LCT intakes on the relationship between feed and output and the effect of LCT intake on output.

Results: 51 children enrolled <18 months of age. 5 children died. 68% of chylothoraces resolved with standard first line diet therapy. 20% resolved with > 2 changes to diet therapy.

Feeds: Feed 1: Reduced LCT formula (Monogen), Feed 2: Fat free modular feed with IV intralipids, Feed 3: TPN and Feed 4 = Reduced LCT (solid diet)

- When feed 1 and 4 were compared: Feed 4 had a significant effect ($p=0.005$) on output. When correcting for the effect of LCT and age, feed 4 gives a higher output than feed 1 by 0.58mls for every unit increase in age, therefore the effects of LCT are marginally significant.
- In patients who received > 2 types of diet therapy but only received feeds 1,3 and 4 there was a significant ($p < 0.001$) decrease in drain output with feed 3 after correcting for the effect of LCT intake
- In patients that received > 2 types of diet therapy and received feed type 2 there was a decrease in drain output but this was not significant ($p=0.13$) however the small sample size was 8 children in the group that received feed 2
- Logistic regression showed there was no effect of output on survival after correcting for LCT and age
- This investigation identified that the Royal Brompton Hospital was the only hospital to use a three staged diet therapy approach thereby avoiding direct transition to TPN. All other centres proceeded to TPN if standard diet therapy failed.

Conclusion:

1. Children who respond to standard diet therapy all exceeded the nationally accepted guideline with no reoccurrence of symptoms.
2. LCT free enteral feeds can reduce drain output and avoid the use of TPN as per step 2 of the Royal Brompton guideline.
3. The duration and timing of diet therapy could potentially be reduced to 4 weeks in patients that respond to standard diet therapy treatment.
4. Further investigation is required to establish evidence based effective guidelines for the management of this high risk post operative complication

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Young people with Congenital Heart Disease – how to prepare for adult care—their own perspective

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Introduction: For many adolescents with Congenital Heart Disease (CHD) medical and surgical progress has resulted in an increased life expectancy. Lifetime care should be provided in order to maximize their potential and lifetime functioning. Paediatric-to-adult transfer of care should be preceded by a preparatory transitional phase. Transition in this respect is defined as a process by which adolescents and young adults with chronic childhood illnesses are prepared to take charge of their lives and their health in adulthood. During adolescence they need to get prepared to take over the responsibility of their health and to adopt good health behaviour in order to prevent late complications.

Objective: To describe young people's perception on what is essential for the transfer to adult care and what they consider to be important to know.

Method: Four group interviews were performed with adolescents aged 14–18 years ($n = 18$), they had moderate to complex CHD. The participants came from 4 different paediatric cardiology

centres in Sweden. The transcribed interviews was analysed with qualitative content analysis.

Result: The analysis revealed in two categories: “an on-going process” and “not being distressed”. The adolescents described the transition as a natural process that has to go stepwise. Further, the adolescents expressed that they had no specific concerns about the transfer while; it was difficult to have an opinion when they did not know what the difference would be. Additional, the responsibility for the process was shared between the cardiology team and the parents.

Conclusion: Young people with CHD have poor knowledge about their diagnosis and what they need to know about the transfer. The transfer procedure is unfamiliar and they are unaware of where their medical check up will continue after leaving paediatric care. The cardiology team should successively introduce and teach young people with CHD how to handle their health and treatment. The parents need to give more responsibility to their child and promote their independence.

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Adolescents with Congenital Heart Disease and their parents—needs before transfer to adult care

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Introduction: Young people with Congenital Heart Disease (CHD) undergo different stages during their life. One of the many important phenomena that occur includes a developmental transition into adulthood, a phase during which young people evolve from being a dependent child to becoming an independent adult. Another important phenomenon is that their setting of care is transferring from a paediatric context to an adult-focused environment. Indeed, a timely and well-prepared transfer to adult-centred care is advocated. For adolescents with CHD and their parents the transfer from paediatric health care to an adult care can be difficult.

Objective: To identify and describe the needs of adolescents with CHD and their parents during the transition before transfer from paediatric cardiology to adult cardiology health care.

Methods: This study has an exploratory design with a qualitative approach, where 13 adolescents with moderate to complex CHD and their parents (12) were individually interviewed. The interviews were analysed with qualitative content analysis.

Results: The analysis of the adolescents' interviews resulted in three categories: Change of relations, Knowledge and information and Daily living. The theme that emerged depicting the meaning of the categories found in the responses was Safety and Control, indicating needs of continuity, knowledge and taking responsibility. Analysis of the parents' answers gave two categories: Change of relations and Daily living. The theme that emerged was Safety and Trust indicating needs of continuity and shifting responsibility.

Conclusions: The transition is a process that must be carefully planned to ensure that adolescents with CHD and their parents can master new skills to manage the transfer to ACHD (Adult with Congenital Heart Disease). A structured programme may facilitate and fulfill the needs of the adolescents and their parents, taking into consideration the aspects of trust, safety and control. However the content and performance of such a program must also be examined. The results from present study will serve as baseline for further intervention studies.

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A Narrative Review of Early Nutrition in Infant after Cardiovascular Surgery

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Introduction: Early nutrition is important for neonate or infant with congenital heart disease. However, few have been known about the knowledge of nutrition in neonate or infant after their complex heart surgery in critical care.

Methods: This study was conducted in a narrative literature review method. A comprehensive search of four electronic databases (PUBMED, MEDLINE, CIHNAL, Cochrane Library) was undertaken between 1990 to 2014. Inclusion criteria were research papers related to outcomes in pediatric patients after cardiovascular surgery, incorporating enteral feeding or nutrition in critic care.

Results: Eighteen research articles and two review articles met the criteria were selected to be discussed. Three themes were emerged from the literature review: (1) Variance affects the weight in congenital heart infants. (2) Implementing advancement and progressive feeding protocol after surgery could be benefit on decreasing hospital or ICU length of stay, gaining weight at discharge, avoiding on incidence of necrotizing enterocolitis and better life of quality on sleep. (3) Addressing on home-monitoring or project after discharge to Norwood stage II.

Conclusions: Most research has been addressed on the nutrition in infant with congenital heart disease in their recovery period after cardiovascular surgery except for patients in critical care. The future research could be focused on approaching the knowledge of factors associated successful enteral feeding or initial feeding time in neonate or infant after cardiovascular surgery in critical care.

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Nursing management of Necrotizing enterocolitis in hypoplastic left heart syndrome after Hybrid Procedure

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Introduction: Hypoplastic left heart syndrome (HLHS), accounts for 2-3% of all congenital heart defects and is a fatal congenital heart defect if untreated. Early diagnosis and treatment of these neonates is priorital to achieving an optimal outcome. Diagnosis, stabilization, medical treatment, surgical repair and recovery are required within the first few days of life. Nursing care of the neonate with CHD begins with the nurse in the labor and delivery room and continues with the nurse in the neonatal intensive care unit. Necrotizing enterocolitis (NEC) is the most serious and frequently acquired gastrointestinal disorder in neonates. According to available literature the incidence of NEC, is higher in patient with HLHS than general population. It is due to the presence of low flow at splanchnic level. The proposed mechanism for the development of NEC in case of HLHS is the combination of a widened pulse pressure and low diastolic pressure as demonstrated by retrograde diastolic flow in the descending aorta, which may contribute to mesenteric ischemia. We report our experience about nurse practitioner in HLHS patients after early hybrid approach and early enteral feeding introduction.

Methods: From October 2011 to November 2014, 43 consecutive neonates underwent single stage hybrid procedure for palliation of HLHS and variants at our Institution. Median age and weight at time of procedure was respectively 1 day and 3 Kg. Hybrid procedure was performed during prostaglandines infusion and

consisted in bilateral pulmonary artery banding, stenting of ductus arteriosus. All patients survived to the procedure but two neonates died respectively at 9 and 15 days of life for myocardial ischemia. Median ICU and hospital stay was respectively 7 and 16 days. All patients were fed according to our dedicated feeding protocol that is based on initial enteral nutrition with low volume oral feedings. After starting feeding nurse provided to the surveillance of each patients according to Bell's criteria.

Results: No episodes of NEC occurred in our population.

Conclusions: Our experience indicates that combination of early hybrid approach, dedicated feeding protocol adherence and dedicated nurse surveillance could reduce the incidence of gastrointestinal complication in this group of neonates.

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...and the anxiety has gone away...**Observational study of preoperative anxiety assessment in children undergoing cardiac catheterization**

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Background: Cardiac catheterization in sedation is a reason of fear and concern for children because it is dreaded what is not known. Children develop anxiety and fear for different reasons such as lack of knowledge of operation, separation from family, little familiarity with hospital environment and carrying out of invasive procedures. It is estimated that 60% of children suffers from anxiety in the pre-operative period. Information and preparation can considerably reduce anxiety if they are made in regard of child's understanding skills. An anxious parent induce a higher anxiety state in child.

The aim of the study is to measure anxiety in preschool and school children undergoing cardiac catheterization comparing the result to parents' anxiety and use of techniques to reduce anxiety.

Methods: The anxiety is measured in children by Modified Yale Preoperative Anxiety Scale (m-YPAS). In the parent, anxiety is measured by State Trait Anxiety Inventory (STAI). Measuring moments are the preoperative day and the day of catheterization. The information/preparation techniques are: delivery of booklet, involvement in therapeutic play performed by child life specialists through the use of books that look like theatres, explanation of the different phases of the procedure by health workers to parents and child, constant presence of parents, visit to the ward during the preoperative day, use of anaesthetic cream before venous cannulation and administration of pre-anesthesia the day of cardiac catheterization, It has also been evaluated the waiting time.

Results: Preschool and school children have been involved. There is no presence of anxiety in children in the preoperative day after information/preparation, presence of maximum anxiety in the day of cardiac catheterization in the moment of the separation from parents in the hemodynamic room. In parents anxiety is always present, it is maximum in the preoperative moment. The major part of the provided reduction techniques has been used but the waiting times between an activity and the other are very long.

Conclusion: Anxiety can be reduced if the approach to child and family is based on multiprofessional actions, if methods are integrated and if parents are involved in the care pathway.

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A New Challenge For Nursing Staff - Spontaneous Breathing Infants with VV ECMO

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Introduction: Extracorporeal Membrane Oxygenation (ECMO) therapy has been available in our Children's Hospital for children with organ failure treated with or without cardiac surgery. Analgo-sedation and ventilation is used during ECMO therapy. Presently we are attempting to reduce the level of analgo-sedation and, for patients with good lung function, early extubation. This change in procedures has created new challenges in caring for these patients.

Methods and Patient Sample: A literature review highlighted the newest evidence in ECMO therapy for children. We tried early analgo-sedation reduction with two infants. A 2-month old boy born with a hypoplastic left heart syndrome was admitted to the pediatric intensive care unit (PICU) after a Glenn Procedure. An oxygenation problem developed due to small pulmonary vessels and ECMO therapy was started. His condition stabilized and he was extubated during ongoing VV-ECMO therapy.

A 5-month old boy with Kawasaki Syndrome was admitted to the PICU following cardiac arrest and resuscitation. ECMO therapy was initiated. Following hemodynamic stabilisation, an LVAD was installed using the ECMO circuit. The child was extubated soon after.

Results: Extubation required a rapid reduction in analgo-sedation, but resulted in agitation in both infants. The 2-month old had periods of uncontrollable agitation unresponsive to verbal or physical comforting. He randomly grabbed catheters and cannulae, negatively affecting the ECMO flow. One-on-one care was initiated, but atelectasis of the right upper lobe developed due to the difficulty with mobilization caused by ECMO cannula placement. Mobilization of the 5-month old was easier due to the type and placement of the cannulae. He was alert, active and could be mobilized, therefore muscle wasting was reduced and lung volume increased. Extubation had a positive effect on the circulation. Less catecholamine was used, nutrition optimized, and elimination normalized.

Conclusion: Early extubation positively influences the circulatory status and is recommended. To achieve this in infants who are breathing spontaneously, analgo-sedation must be adequate and optimal cannulae placement critical.

Enabling the provision of one-on-one care is advised and parents can be supported to help care for their infant.