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Cardiology in the Young

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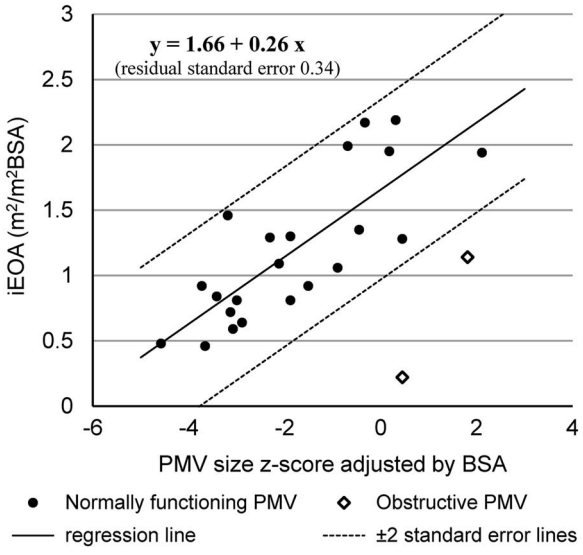
50th Annual Meeting of the Association for European Paediatric and Congenital Cardiology, Rome, Italy, June 1–4, 2016

YIA-1

Echocardiographic assessment of prosthetic mitral valve in children

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Introduction: Although the diagnostic utility of Doppler-derived hemodynamic parameters of prosthetic mitral valve (PMV) for detecting prosthetic dysfunction has been demonstrated in adults, interpreting these parameters in children is difficult, as the relative size of PMV varies more greatly in them than in adults. We aimed to elucidate how the Doppler-derived hemodynamic parameters change as the PMV size decreases relative to the somatic growth of children. We also evaluated the diagnostic utility of the parameters for detecting prosthetic obstruction.
Methods: We reviewed 26 echocardiographic examinations of 15 mechanical PMVs in 12 children. The median age at echocardiographic examination was 6.6 (0.6–18.1) years. The PMV functioned normally in 24 examinations, but was obstructed due to intraoperatively confirmed PMV thrombosis in 2 examinations. PMV size ranged between 16 and 25 mm, which was standardized to body surface area (BSA) at the examination by calculating z-scores based on previously published normative data of mitral valve diameter. We assessed the Doppler-derived hemodynamic parameters, namely peak E velocity, mean pressure gradient (PG), and pressure half time (PHT) of the transprosthetic flow, the velocity-time integral (VTI) ratio of the PMV inflow to the left ventricular outflow, and BSA-indexed effective orifice area (iEOA) of the PMV calculated by using the continuity equation. The correlation between the PMV size z-score and Doppler-derived hemodynamic parameters of the normally functioning PMVs was evaluated by using the linear regoression analysis. The measurements in the obstructive PMVs were compared with the predictive values derived from the regression equations. A p-value of < 0.05 was considered statistically significant.
Results: All parameters of the normally functioning PMVs had a statistically significant correlation with PMV size z-score (Pearson correlation coefficients: peak E velocity, −0.68; mean PG, −0.71; PHT, −0.82; VTI ratio, −0.76; and iEOA, 0.79). Multivariate analysis revealed that PHT and iEOA were independently correlated with PMV size z-score. iEOA was the sole parameter with which both cases of obstructive PMV exceeded ± 2 standard errors of the predictive value.
Conclusions: iEOA strongly reflects BSA-indexed PMV size. Deviation from the expected iEOA, based on relative PMV size, may indicate prosthetic obstruction in children with PMV.



YIA-2

Postnatal developmental changes in sensitivity of L-type Ca2+ channel to inhibition by verapamil

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Background: L-type Ca2+ channel (ICa,L) blockers can be divided in three groups on the basis of their structures; namely, dihydropyridines, benzodiazepines and phenylalkylamines. They are widely used for the treatment of hypertension and tachyarrhythmias in clinical settings. However, intravenous administration of the phenylalkylamine verapamil for tachyarrhythmias is considered to be contraindicated in neonates and infants, due to

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the perceived risk of hypotension or bradycardia. However, its ionic basis has yet to be fully elucidated. In the present investigation, we examined the postnatal developmental changes in the sensitivity of ICa,L to the three classes of its blockers using mouse heart model.
Methods: Ventricular myocytes were enzymatically digested from the heart of postnatal days 0, 7, 14, 21, 28 and adult (10-15 weeks) mice using similar Langendorff-perfusion methods. Whole-cell patch-clamp technique was used to record ICa, L in ventricular myocytes of various postnatal ages in the absence of presence of nifedipine, diltiazem and verapamil at concentrations of 1 nM to 1 μ M. Concentration-relationship was constructed by plotting the percentage inhibition of ICa,L as a function of drug concentrations.
Results: There is a postnatal developmental increase in the amplitude of ICa,L. ICa,L in day-28 and adult ventricular myocytes are larger than that in day-0, day-7, day-14 and day-21 myocytes. The half-maximally inhibitory concentration (IC50) for the inhibition of ICa,L by verapamil was significantly smaller in day-0, day-7, day-14 and day-21, compared with day-28 and adult ventricular myocytes. In contrast, there were no significant differences in IC50 for the inhibitory action of nifedipine or diltizem in all postnatal developmental ages.
Conclusions: ICa, L in neonates and infants exhibits a higher sensitivity to inhibition by verapamil compared with that in child and adult stages in the mouse model, which may explain at least partly severity of the verapamil-induced hypotension in neonates and infants.

YIA-3
History of open heart surgeries strongly predicts a type of restrictive ventricular characteristics (constrictive or restrictive) in repaired adult patients with congenital heart disease

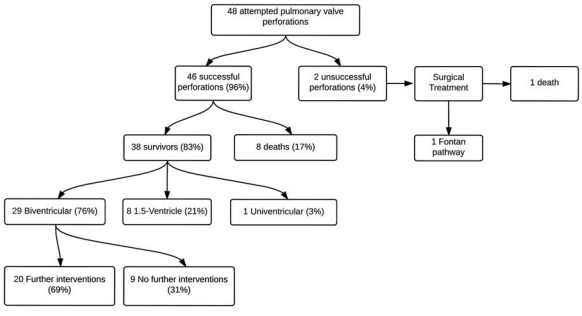
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Background: Adults with congenital heart disease (ACHD) often have histoies of multiple open heart surgeries (OHS) that may cause restrictive hemodynamics characterized by high central venous and ventricular end-diastolic pressures (CVP, EDP). Constrictive change due to pericardial adhesion and restrictive remodeling of myocardium after OHS might coexist. Their characteristics and clinical features have not been well described.
Methods: We studied 64 consecutive postoperative ACHD patients with biventricular physiology (29 ± 10 years old, tetralogy of Fallot's in 37, 29 males). We obtained intracardiac pressure waveforms by cardiac catheterization and calculated the ratio of the right ventricular to left ventricular systolic pressure-time area during inspiration versus expiration (systolic area index; SAI, JACC 2008) that is the gold standard for differentiating ventricular constriction ($SAI > 1.1$) from restrictive physiology ($SAI < 1.1$). We compared the SAI with clinical profiles, including hemodynamic parameters and peak oxygen uptake (peak VO2).
Results: Of clinical variables, the number of OHS was the only determinant of SAI ($r = 0.45$, $p < 0.001$). We divided our patients into four subgroups based on the number of OHS; ACHD who had underwent OHS never ($n = 3$), once ($n = 33$), twice or three times ($n = 21$), and more than three times ($n = 7$). The percentage of SAI >1.1 for each groups was 0%, 36%, 62% and 0%, respectively, suggesting their ventricular characteristics would shift from constrictive to restrictive as the number of OHS increased. Age, sex, ejection fraction of both ventricles, and cardiac index were not significantly different between the groups ($p > 0.1$). However, ACHD with greater number of OHS showed smaller left ventricular end-diastolic volume (80, 87, 90, 71 mL/m^2, respectively, $p = 0.024$), lower peak VO2 (27, 27, 24, 17 mL/kg/min, respectively, $p < 0.01$), higher CVP (4, 3, 7, 15 mmHg, respectively, $p < 0.001$), higher EDP of both ventricles, and higher levels of serum brain natriuretic peptide (9, 32, 49, 163 pg/mL, respectively, $p < 0.02$).
Conclusion: In postoperative ACHD patients with biventricular physiology, most patients show constrictive changes. And restrictive myocardial damages may add on the constrictive characteristics with the histories of multiple OHS, resulting in advanced ventricular diastolic dysfunction.

YIA-4
Long Term Outcome after Catheter Perforation of Pulmonary Valve in Pulmonary Atresia and Intact Ventricular Septum – a 25 year experience

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Introduction: Perforation of the pulmonary valve (PV) via interventional catheterisation for patients with pulmonary atresia and intact septum (PAIVS) was pioneered at our institution twenty-five years ago and is now an established procedure. This study examines the long-term outcome for our complete patient cohort, identifying late complications and outcome predictors.
Methods: Patients who underwent catheter based perforation with PAIVS from 01/05/1990 to 25/12/2015 were identified from the Departmental database. Demographic and clinical data were collected, with final outcome at last follow-up evaluated on 31/12/2015. One-way-ANOVA and Kruskal-Wallis tests were performed for data that was normally and non-normally distributed, respectively. Receiver operating characteristic (ROC) curve analysis was performed as predictor of biventricular (BV) versus non-biventricular (non-BV) outcome amongst survivors, with bimodal logistic regression to assess the relevance of risk factors.
Results: 48 patients were identified. The table and figure below demonstrate the key findings. The median follow-up period was 12.2 years (Range 0.3-25.9 years). All deaths were within 90 days of birth. One patient had an embolic stroke at age 4.3 years following TCPC completion, No other significant complications were identified. Five patients with BV outcome had surgical RV overhaul procedures at a mean age 1.6 years. No patient has as yet required interventional or surgical procedures beyond 11 years. One patient is currently listed for PV replacement. Saturations at one year (Sats1Yr) were a strong predictor of BV versus non-BV outcome (area under curve (AUC) 0.889) with Sats1Yr of 90% predicting BV outcome with sensitivity 75% and specificity 78%. AUC for tricuspid and PV z-score at birth were 0.715 and 0.744 respectively. Logistic regression modelling confirmed the prominent role of Sats1Yr in achievement of Biventricular repair.



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Conclusions: PV perforation for PAIVS is associated with good long-term outcomes, with few late complications. At up to 25 years follow-up, the incidence of arrhythmias and ventricular dysfunction secondary to PV regurgitation is low. The initial size and anatomy of the RV structures is clearly a strong predictor of final outcome, but the contribution of Sats1Yr suggests other possible factors such as diastolic dysfunction and ventricular fibrosis may play a significant role.

n = 48	Failed (2)	Died (8)	Non-Biventricular (9)	Biventricular (29)	p*
Birthweight	3.4 ± 0.75	2.69 ± 0.29	3.11 ± 0.15	3.07 ± 0.10	0.22
Procedure age(d)	18 ± 4	12.88 ± 8.53	5 ± 0.7	4.83 ± 0.8	0.80
Duct stent	0	3	7	13	0.12
zPV	-2.08 ± 0.65	-2.86 ± 0.17	-3.65 ± 0.44	-2.1 ± 0.3	0.012
zTV	-8.76 ± 2.54	-6.93 ± 1.98	-5.87 ± 0.69	-3.41 ± 0.52	0.019
Sats1Yr	-	-	84 ± 2	93 ± 1	<0.001

*'Failed' group excluded from statistical analysis, due to low numbers
zPV and zTV = Pulmonary valve and Tricuspid valve Z scores at birth (Daubeney et al.)

YIA-5

Pulmonary arterial stiffness indices assessed by intravascular ultrasound in children with early pulmonary vascular disease: prediction of disease progression and mortality during 20-year follow-up

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Introduction: Prognosis in children with pulmonary vascular disease (PVD) is closely linked to right ventricular (RV) failure due to increased RV afterload. Pulmonary arterial (PA) stiffening is known to occur early in the disease course of PVD and constitutes a main component of RV afterload. We aimed to evaluate the clinical value of indices of PA stiffness in children with early or advanced PVD, by determining the association of such indices with long-term disease progression and mortality.
Methods: Forty-one children with arterial PVD in early or more advanced stages, defined as mean PA pressure ≥20 mmHg and/or pulmonary to systemic flow ratio ≥1.2, and mean pulmonary capillary wedge pressure <15 mmHg, underwent cardiac catheterization with intravascular ultrasound (IVUS) imaging between 1994 and 1997 with follow-up until 2015. Indices of PA stiffness evaluated were compliance and distensibility. During long-term follow-up, transthoracic echocardiography and cardiac catheterization were performed to determine whether PVD had reversed or progressed.
Results: Following baseline cardiac catheterization, 27 (66%) patients underwent closure of a cardiac shunt defect. During a median (interquartile range) follow-up of 19 (18-20) years, 31 (76%) cases of PVD had reversed and 10 (34%) had progressed. Six patients died due to PVD. In addition to conventional hemodynamics, lower compliance and distensibility were significantly associated with PVD progression ($p = 0.007$ and $p = 0.011$) and mortality ($p = 0.007$ and $p = 0.009$), also after adjustment for age, sex and end-diastolic PA luminal area. Survival rates differed significantly between patients with high and low compliance ($p = 0.011$) and distensibility ($p = 0.009$, Fig. 1). Also in a subgroup of patients with favorable hemodynamic profiles at baseline, lower compliance and distensibility were associated with progression of PVD during follow up ($p = 0.002$ and $p = 0.030$).
Conclusions: PA stiffness indices assessed by IVUS are associated with long-term disease progression and mortality in children with PVD and are a valuable complement to conventional hemodynamic evaluation, especially in the early stages of the disease.

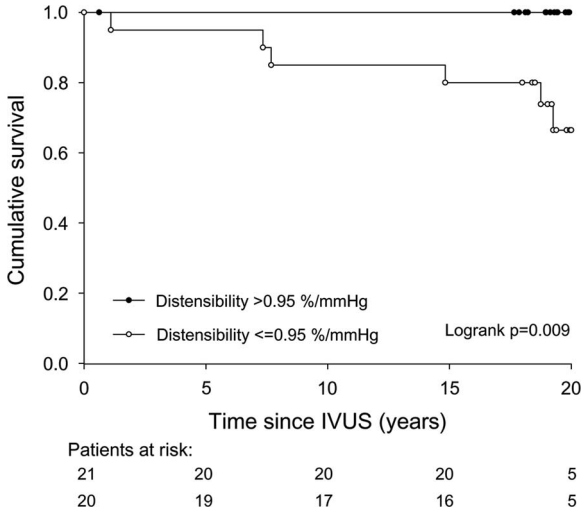


Figure 1. Comparison of survival of patients with high and low distensibility.

YIA-6

Late causes of death after congenital cardiac surgery - a population-based six-decade study with 98% follow-up

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Introduction (or Basis or Objectives): Survival after paediatric congenital cardiac surgery is lower than the general population, but has improved significantly during recent decades. Consequently, the number of adults with congenital heart defects (CHD) has surpassed paediatric patients. The majority of patients require lifelong follow-up. Information regarding causes of death late after surgery would benefit follow-up among these patients. We investigated whether late modes of death after surgery for CHD's have changed in recent years and how they compare to rates among the general population.
Methods: We obtained data retrospectively from a nation-wide CHD database, including paediatric patients who underwent cardiac surgery at <15 years of age at one of five university– or one district hospital in Finland 1953-–2009. The Finnish population registry supplied patient survival status. We categorised modes of death into CHD-related and non-CHD-related deaths using ICD diagnostic codes. Modes of death among the study population

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were compared to those among a sex-, age-, birth-time-, and hospital district-matched control population supplied by Statistics Finland.

Results: Between 1953 and 2009, 10,964 patients underwent 13,876 operations, with 98% follow-up (10,692). The longest follow-up was 60 years. Early (< 30 days) and late mortality were 5.7% (612 patients) and 11% (1,130 patients), respectively. The incidence of CHD-related death correlated with defect severity. Fatal heart failure (17% with pulmonary hypertension) was the most common mode of CHD-related death, but decreased among those undergoing surgery 1990–2009 (Table 1). The incidence of sudden death was zero after surgery for ASD, VSD, TOF, and TGA 1990–2009 (Table 1). Deaths due to respiratory, neurological, endocrine and metabolic disease were significantly more common among study patients than the general population. Pneumonia constituted the majority of non-CHD-related deaths among the study population.

Conclusions: Sudden death and fatal heart failure after congenital cardiac surgery decreased markedly among study patients, but remained a significant cause of death among patients operated for severe cardiac defects, warranting life-long surveillance. Pneumonia requires immediate diagnosis and treatment among these patients.

Table 1. Incidence of CHD-related deaths among different defect groups by era of operation.

	Cardiovascular incidence/1000 PY		***Heart failure incidence/1000 PY***	
Defect	1953–1989	1990–2010	1953–1989	1990–2010
PDA	0.07	0	0.31	0
ASD	0.08	0	0.11	0
COA	0.17	0	0.64	0.58
VSD	0.33	0.08★	1.42	0.84★
TOF	0.85	0.26	1.94	0.78
TGA	1.12	0.95	4.65	2.21★★
UVH	3.50	3.47	11.51	8.69
	Sudden death incidence/1000 PY		***Perioperative incidence/1000 PY***	
Defect	1953–1989	1990–2010	1953–1989	1990–2010
PDA	0.03	0.13	0.04	0
ASD	0.23	0	0.04	0
COA	0.64	0.14	0.20	0.29
VSD	0.61	0★★	1.13	0.34★★★
TOF	1.94	0	1.55	0.78★
TGA	3.53	0★★★	2.41	2.52
UVH	4.50	4.63	9.01	6.95

PY = Person years; ★p < 0.05; ★★p < 0.01, ★★★p < 0.0001 PDA patent ductus arteriosus; ASD atrial septal defect; COA coarctation of the aorta; VSD ventricular septal defect; TOF tetralogy of Fallot; TGA transposition of the great arteries; UVH univentricular heart

O1-1

Percutaneous PDA closure in Extremely Low Birth Weight Babies

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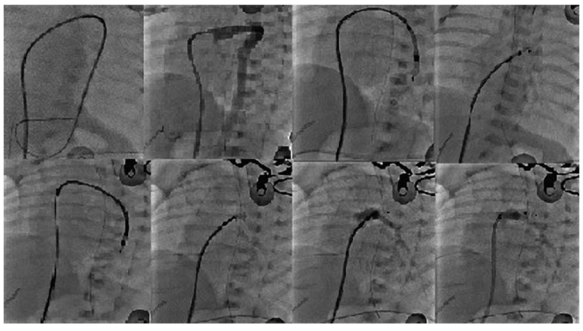
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Aim: Patent Ductus Arteriosus is an important cause of morbidity and mortality in preterms. As birthweight decrease, risks increase. Main aim of our study is to emphasize the effectiveness and safety of percutaneous PDA closure even in extremely low birth infants (less than 1000 gr).

Material Method: In our center between the dates June 2014-December 2015, PDA of eight patients less than 1000gr were closed percutaneously. To our knowledge this study includes the largest cohort of infants less than 1000g in the literature, whose PDA were closed percutaneously.

Results: Symptomatic patients, less than 1000 gr having PDA were included in the study. All have 3 times medical therapy for PDA closure but it did not work. PDA was decided to be contributor of this medical state of them. The mean patient age 16 ± 5.9 days. The mean weight of patients was 923 ± 75.9 gr. Mean gestational age was 27.2 ± 1.28 weeks. Mean PDA diameter was 2.48 ± 0.5 mm. Mean Qp/Qs was 1.7 ± 0.2. Morphology of PDA: 5 of them were conical and 3 of them were tubular. In all patients ADOII-AS device were used for PDA closure (Table1). Steps of percutaneous PDA closure procedure was shown by Figure 1. In all patients, we have done closure by venous route. We did not ever used arterial route in 4 patients. There were no major complications reported. Left pulmonary arterial stenosis was detected in 2 patients which were all resolved in 6 months duration.

Conclusion: Interventional catheterization procedures are more commonly used, in the recent years. The advantages of percutaneous PDA closure include a high success rate, shorter length of hospital stay, reduced blood loss, low morbidity rate, and no traumatic scars. Since the length of hospital stay decreases with catheterization, it is much more cost-effective than surgery. We want to emphasize that in experienced centers percutaneous closure of PDA can be an alternative to surgery even in the extremely low birth weight babies.



	Gestational week	**Gender**	**Age (day)**	**Weight (gram)**	**PDA Type**
1	28	Female	15	980	conical
2	29	Male	24	910	conical
3	27	Male	10	978	Tubular
4	28	Female	21	930	Conical
5	28	Female	10	970	Tubular
6	26	Female	10	750	Conical
7	27	Female	23	820	Conical
8	24	Female	15	960	Tubular

	PDA Size (mm)	**Qp/Qs**	**Device type**	**Device diameter**	**Floroscopy Time(min)**
1	3.5	1.9	ADOII-AS	5 × 2	21
2	2.4	1.5	ADOII-AS	3 × 2	8
3	1.7	1.66	ADOII-AS	3 × 2	12
4	2.5	2.1	ADOII-AS	4 × 5	20
5	2.5	1.6	ADOII-AS	4 × 2	8
6	2	1.7	ADOII-AS	3 × 4	19
7	3	2	ADOII-AS	4 × 2	10
8	2.2	1.7	ADOII-AS	3 × 2	13

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O1-2

Percutaneous Pulmonary Valve Implantation with Edwards–Sapien XT in Patients with Native and Large Right Ventricular Outflow Tract; Early Results

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Introduction: Percutaneous pulmonary valve implantation (PPVI) has been used mainly for conduit dysfunction in right ventricular outflow tract (RVOT). Until recently, native RVOT without stenosis used to be considered a relative contraindication to transcatheter valvulation. We present early results of PPVI with Edwards–Sapien XT (ES-XT) in repaired tetralogy of Fallot (TOF) patients with native-large RVOTs.
Method: 34 s/p repaired TOF patients who had native RVOT with free pulmonary regurgitation and right ventricular dilatation without significant stenosis. Balloon sizing was performed with compliant (34 mm Amplatzer sizing) and semi-compliant balloons for interrogation. The size of the Z-Med balloons and BIB catheters that the Andra Stents XXL would be mounted on was decided up to the indentation diameter occurred during interrogation; as at least 1 mm larger than the indentation diameter.
Results: Mean age and weight of the patients were 18 9.2 (7-50) years and 48 15 (22-84) kg, respectively. Before presenting pressure gradient between right ventricle and pulmonary artery was 5.6 4.3 (0-14) mmHg. Indentation diameter with balloon interrogation was 25.6 2.2 (22-28) mm. Balloon size used for prestenting was 27.72.2 (24-30) mm. Successful valve implantation was achieved in all patients; 29 mm in 27 and 26 mm in seven. Valvulation was performed in same session in four and 3-12 weeks after prestenting in 30. Valve function was good in all immediate after and at the last follow-up; a median of 4.5 months (1-15 months). Mild paravalvular leakage was observed only in two. Stent fracture has not been observed and no reintervention required yet.
Conclusion: PPVI with ES-XT valve, which has larger sizes as 26 and 29 mm, is feasible and safe in patients larger native RVOT without stenosis in adolescents and adults. Newer delivery system (Novaflex), which is used through 14-20 Fr smaller sheaths, gives us also an opportunity of early transcatheter valvulation in smaller patients with native RVOT. Prestenting for providing a secure landing zone is the most important part of the procedure. Only Andra XXL stents which has an expansion capacity up to 32 mm can be used in this purpose, currently.

O1-3

Performance of a self-expanding stent specifically designed for percutaneous arterial duct stenting for the "Gießen hybrid" procedure for primary treatment of complex congenital heart defects

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Objectives: To report on our 5-year single-center experience with a specifically designed self-expanding stent for the arterial duct for the "Gießen hybrid" procedure in complex congenital heart disease (CCHD).
Methods: Retrospective and prospective evaluation (December 2010 to December 2015) of implantation and performance of the self-expanding, Sinus Superflex-DS (Opti-Med).
Results: 52 newborns underwent the "Gießen hybrid" procedure with uneventful percutaneous arterial duct stenting for primary treatment of CCHD (hypoplastic left heart syndrome/complex (HLHS/HLHC), n = 30/16; others, n = 6) at a median age of 6 days (range 1 – 50) with a median weight of 3.0 kg (range 1.9–4.4; n = 9 ≤2500 g) and a median Aristotle score of 17.0 (range 14.5–21.5). Implantation of a second stent to completely cover the arterial duct was necessary in 8 patients. Duct restenosis treated by balloon dilation and/or implantation of a second stent occurred in 13 patients. 26 patients underwent comprehensive stage II palliation, 10 received biventricular repair, 13 are on inter-stage of which 3 are planned for biventricular repair. Heart transplantation was performed in two patients (pt #1 with severely hypoplastic ascending aorta and signs of cardiac ischemia, pt #2 with HLHC and non-compaction cardiomyopathy). Inter-stage mortality occurred in one patient with hypoplastic left heart syndrome and coarctation of the aorta due to cardiac ischemia.
Conclusions: The Sinus-Superflex-DS received a CE-mark for percutaneous arterial duct stenting in newborns. The low profile and open cell design allows usage with low risk and is easily positioned by arterial access through a 4 Fr sheath. However, in some cases the low radial power of the self-expanding stent requires balloon re-dilation.

O1-4

Nine years of experience with the Melody valve for PPVI Valve function remains preserved, incidence of endocarditis can be reduced

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Introduction: worldwide the experience with the Melody valve for PPVI is growing and also the indications for use have been broadened. Endocarditis appears to be a serious threat.
Patients and methods: national data are collected from the Belgian registry which started in 2006. Four implant centers serving 11 million people. Follow-up data prospectively and ongoing collection: leaflet function (echo), re-interventions, mortality, endocarditis, X-ray (stent fractures).
Results: 222 valves have been implanted 2006-2015. The indication was PS 40%, PR 35% and mixed PS/PR 25%, male/female ratio 1.8, mean age at implant 20.7 y (3.9-81.6), original lesion : Fallot 50.4%, PS 9%, Arterial Trunc 8.6%, DORV with PS 17.1% and other 11.9%. Adequate pre-stenting in 218 pts. Follow-up data : at last follow-up median RVOT gradient 22 mmHg (5-30), median PI 0/4 (0-2). Endocarditis occurred in 14/206 (6.8%), freedom from endocarditis is 85% at 8 y; incidence dropped after firm recommendations of life-style (skin and dental care) and prophylaxis from 2.8%/y to <0.6%/y. Overall survival was 98% at 9 years, no procedure or valve related deaths. Freedom from stent fractures was 80% at 9 y, but none was hemodynamically important and none did require intervention. Overall freedom from re-intervention was 96% at 9y, mostly balloon dilation for somatic growth. Five valves were explanted: endocarditis (n = 4), residual subPS (n = 1).
Conclusion: With 9 years of follow-up the leaflet function remains well preserved. Adequate pre-stenting the RVOT prior to PPVI has abolished the risk of hemodynamic important stent. Endocarditis incidence was reduced after firm recommendations of life-style and prophylaxis.

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O1-5

Pulmonary valve replacement: is there an interest for the "folded Melody valve" in short RVOT with early PA bifurcation and/or abnormal course of coronary artery?

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Transcatheter pulmonary valve replacement with the Melody valve may be challenging in patients with short RVOT and early PA bifurcation because of the risk of jailing one PA or in patients with abnormal course of coronary artery. Folding of the Melody valve may be a solution.
Since March 2015, 5 patients were included in this study, aged from 10 to 45 years. Initial pathology included transposition and coarctation (n = 1), pulmonary stenosis and VSD (n = 1) or ASD (n = 1), tetralogy of Fallot (n = 1) and with pulmonary atresia (n = 1). These patients had undergone from 2 to 5 previous surgical repairs. The indications for pulmonary valve replacement were: mixed lesion (n = 4) and pulmonary leak (n = 1). All patients had before the procedure, MRI study and CT scan to delineate the exact morphology of the RVOT. Before implantation, balloon dilatation of the RVOT with control aortography to obviate any coronary artery compression was performed in all. Prestenting was realized in all with LD max stent (Ev3), usually placed under left ventricular pacing. Implantation of the Melody valve was realized in native RVOT (n = 2), in pulmonary homograft (diameter 26 and 27 mm)(n = 2), and in 18 mm Contegra conduit (n = 1). The indications for a folded melody valve were a short pulmonary artery trunk for 4 patients and abnormal course of the right coronary artery between the aorta and the pulmonary conduit in one. Folding of the Melody valve was performed on both extremities of the stent (n = 1) and only on distal end (n = 4).
During follow-up (1 to 21 months), no patient had reintervention. No myocardial ischemia was reported.
Folding of one or both extremities of the Melody valve is easy to perform and can be a good therapeutic option for patients with short RVOT and early PA bifurcation and/or abnormal course of coronary artery, especially in those with not too large native RVOT or conduit. Initial results are good but more experience and longer follow-up are mandatory.

O1-6

Percutaneous pulmonary Melody valve implantation in small conduits: early and long-term results

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Background: The Melody® transcatheter pulmonary valve received approval for treatment of dysfunctional right ventricular (RV) outflow tract conduit ≥16 mm. Limited data are available for the use of the device in small conduits.
Objective: To investigate technical and clinical outcomes of patients who underwent percutaneous pulmonary valve implantation (PPVI) in conduit ≤16-mm.
Methods and results: Eleven patients were included between 2009 and 2015. Primary underlying diagnosis was tetralogy of Fallot (n = 4), Ross Procedure (n = 4) and common arterial trunk (n = 3). RV outflow tract characteristics included: heterograft or homograft (n = 11, expandable in n = 10). Conduit diameter at time of surgical implant range from 12 to 16-mm. Indication of PPVI was stenosis or mixed lesions in 10 and regurgitation in one. All were prestented: 6 during a previous cardiac catheterization (median time between stenting-PPVI, 11-months, range 3 to 86 months) and 7 during the PPVI procedure (3 patients in addition with a previously placed stent). The median largest dilatation balloon diameter/implanted conduit diameter ratio was 1.25. All procedures were successful. Procedural hemodynamics showed a decrease in peak RV to PA gradient (-mmHg) from 45.5 ± 22.6 to 11.7 ± 6.8 ($p < 0.001$), in RV systolic pressure (-mmHg) from 67.8 ± 22.8 to 39.8 ± 8 ($p < 0.001$), and in the RV/Ao ratio from 0.74 ± 0.22 to 0.44 ± 0.09 ($p < 0.001$). No patient had significant pulmonary regurgitation. Early complications occurred in four patients. There were 2 confined conduit tears managed with placement of a covered stent; and 2 local vascular complications requiring prolongation of hospital stay but no transfusion. Mean follow-up after PPVI was 3,6-years (23days to 6,7years). Late complications occurred in 4 patients: 2 endocarditis requiring surgical removal (1 early, 1 late); 2 recurrent stenosis with one requiring surgical conduit replacement, and one a percutaneous Melody re-dilatation. Freedom from valve dysfunction or re-intervention was 64% at last follow up.
Conclusion: PPVI is feasible in small conduit ≤16mm with good procedural and early hemodynamic result postponing the need for surgery. However, rates of reintervention and complications are higher as compared with patients with larger conduits.

O2-1

Indications for Aortic Valve Interventions in Pediatric Patients with Bicuspid Aortic Valve

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Introduction: Bicuspid Aortic Valve (BAV) is one of the most common causes of aortic valve (AV) intervention in pediatric patients. There is limited data available regarding the indications used by cardiologists and surgeons for deciding mode of intervention in pediatric patients with BAV.
Methods: Mayo Clinic Echocardiography Database was retrospectively reviewed to identify 1010 patients (<22 years) diagnosed with BAV from 1990-2015. Indications for aortic valve interventions were interpreted from the clinical and surgical notes.
Results: Out of 1010 patients with BAV, 109 (10.7%) young patients underwent interventions of the AV. In patients with BAV, critical aortic stenosis (CAS) (100%) and left ventricular hypertrophy (LVH) (43%) were the most common indications for balloon valvuloplasty. Similarly, CAS was the primary indication for surgical valvotomy, but in slightly older patients (mean age 4.7 years). Surgical repair/valvuloplasty was performed mainly for aortic regurgitation (AR) (69%), in yet older patients (mean age 10 years). AV replacement (AVR) was a fourth modality utilized, and was performed mainly for AR (72%), at a mean age of 17 years. AVR was the intervention most likely to take into account clinical presentation (38%). Echocardiographic data is mentioned in the table.
Conclusions: 10.7% young patients with BAV had severe enough disease that warranted aortic valve intervention. Type of valvular dysfunction, as well as age, were important factors in decision-making. Symptomatic presentation was considered more important for AVR.

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	Balloon Valvuloplasty	Surgical Valvotomy	Surgical Repair	Aortic Valve Replacement
Total No.	51	29	18	39
Right-left fusion	29(57%)	16(55%)	12(67%)	18(46%)
Right-non fusion	22(43%)	13(45%)	6(33%)	21(54%)
Mean Age (years)	2.7	4.7	10	17
Male/Female				
Symptomatic	35%	38%	40%	64%
	Factors in Deciding Mode of Intervention			
Aortic Stenosis	**100%**	100%	37%	48%
Aortic Regurgitation	0%	0%	**69%**	**72%**
CAVD	0%	0%	6%	21%
LVH	**43%**	24%	12%	3%
Clinical Presentation	**18%**	**14%**	**19%**	**38%**
	Imaging Data			
AS gradient (mmHg)	62 ± 22	49 ± 22	42 ± 35	35 ± 26
AR grade	1	2	4	4
LV Mass Index (g/m^2)	82 ± 38	103 ± 28	119 ± 36	129 ± 45
PWT (z-score)	1.6 ± 2	1.6 ± 1.4	2.2 ± 1.9	1.1 ± .5

CAVD, combined aortic valve disease; LVH, left ventricular hypertrophy; PWT, posterior wall thickness; AS, aortic stenosis; AR, aortic regurgitation

O2-2
The changing epidemiology of Acute Rheumatic Fever in Manitoba's paediatric population: results of a 10-year retrospective study

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Objectives: Thirty years ago, the incidence of acute rheumatic fever (ARF) in Manitoba was found to be significantly greater in Aboriginal compared to non-Aboriginal children. Although Aboriginal children now make up 25% of Manitoba's paediatric population, ARF epidemiology in Manitoba's paediatric population has not been recently reviewed. In this study we determine incidence rates of ARF in Aboriginal and non-Aboriginal children in Manitoba from 2000-2010, and compare these to rates from 30 years prior.
Methods: We examined ARF incidence rates in Manitoba, Canada from 2000-2010 for children aged 0-17 years, residing in Manitoba or the Kivalliq region of Nunavut. Study participants were identified from the Variety Heart Centre patient database and the diagnosis of ARF was based on the revised Jones criteria. Incident rates were calculated by ethnicity, gender and location of residence. Rates were compared using rate ratios and 95% confidence intervals.
Results: 59 patients were eligible for this study. From 2000-2010, the incidence rate of ARF in Manitoba's total paediatric population was 1.71 per 100 000 person years. The incidence rate in the Aboriginal population was significantly greater than that in the non-Aboriginal population ($p < 0{\cdot}0001$) at 4.98 per 100 000 person years and 0·618 per 100 000 person years respectively. Moreover, the Aboriginal population had an 8·07 (95% CI 4.54-14.32) times greater relative risk of acquiring ARF compared to the non-Aboriginal population. While the absolute incidence rates across all population groups fell over 30 years, the relative risk of acquiring ARF as an Aboriginal child compared to a non-Aboriginal child almost doubled in that time. Lack of safe water sources, overcrowding, and poor access to health care were identified as socioeconomic factors that may contribute to the higher incidence of ARF found in Aboriginal children.
Conclusions: While ARF incidence rates in Manitoba's paediatric population as a whole have decreased over the last 30 years, incidence rates in the Aboriginal population are still unacceptably higher than the non-Aboriginal population. Targeted primary and secondary prevention strategies are needed in order to decrease the direct and indirect burden of ARF in this population.

Table 1. Incident rates of ARF in Manitoba by year of study (per 100 000 person years)

	1970–1979	2000–2010	p-value
Entire population (95% CI)	35·89 (32·87-39·20)	1·71 (1·32-2·20)	<0·0001
Aboriginal population (95% CI)	125·75 (105·39-150·04)	4·98 (3·69-6·72)	<0·0001
Non-Aboriginal population (95% CI)	29·02 (26·21-32·13)	0·62 (0·38-1·01)	<0·0001
Relative Risk (95% CI)*	4·33 (3·53-5·31)	8·07 (4·54-14·32)	-

*Relative risk of ARF in the Aboriginal compared to Non-Aboriginal population

O2-3
Congenital Heart Disease and Long-term Risk of Dialysis-requiring Chronic Kidney Disease: a Nationwide Population Study

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Background: Best evidence suggests that approximately 30-50% of adult patients with congenital heart disease (CHD) have impaired renal function. It is unknown to what extent this impairment is related to the development of dialysis-requiring chronic kidney disease (DRCKD) over a lifetime.
Aim: To examine the overall risk of DRCKD in the CHD population, with risk stratified by CHD severity, compared with the general population.
Methods: In this cohort study we used two nationwide population-based medical databases to identify individuals diagnosed with CHD in Denmark between 1963 and 1974 (before 15 years of age) and from 1977 to 2003 (at any age). Patients were followed from 1999 to 2012 for first episode of dialysis treatment for CKD using data from the Danish National Registry of Patients (DNRP: nationwide hospital discharge registry covering all Danish hospitals). DNRP dialysis coding provides a distinction between dialysis for CKD and dialysis for acute kidney failure. For each CHD subject we identified 10 controls from the general population, matched by sex and birth year. A unique personal identifier assigned at birth and used in all Danish public registries enabled virtually complete follow-up for migration, death or DRCKD. We computed cumulative incidences and hazard ratios (HR) of DRCKD.
Results: We identified 18,033 CHD subjects and 177,413 comparison cohort subjects. By 40 years of age, the cumulative incidence of DRCKD was 0.6% among CHD subjects and 0.1% among the control population. The HR of DRCKD among CHD subjects compared with controls was 7.2 (95% CI: 4.6-5.6). The HRs for mild, moderate and complex CHD were 2.3 (95% CI: 1.0-5.6), 6.6 (95% CI: 2.3-18.0), and 34 (95% CI: 11-105), respectively.
Conclusion: The incidence of DRCKD was increased in individuals with CHD compared with the general population. Although the absolute risk of DRCKD was low, this should be an important consideration for optimizing preventive and acute cardio-renal care for the adult CHD population.

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O2-4

Contemporary Outcomes and Factors associated with Mortality after a Fetal or Neonatal Diagnosis of Tricuspid Valve Disease

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Background: Ebstein's anomaly (EA) and tricuspid valve dysplasia (TVD) are rare anomalies and data on outcomes after a fetal or neonatal EA/TVD diagnosis is conflicting.
Methods: To examine the outcome and identify markers predictive of mortality, we reviewed our single center experience with 79 cases of EA/TVD from 2000-2014. Variables were analyzed separately for cases diagnosed in-utero without pregnancy termination (n = 5) and for all live-born patients.
Results: Of 47 fetal cases with an intention-to-treat, 8 (17%) died in-utero and 10 (21%) as neonates. Independent predictors of in-utero demise included severe tricuspid regurgitation with a Doppler gradient < 40 mmHg (odds ratio (OR) 1.22 per mmHg deduction; p = 0.003) and pulmonary regurgitation (OR 11.4; p = 0.03) at the baseline exam. A novel prognostic score (range 0-10) combining the severity of 5 echocardiographic findings shown in the Table was independently associated with overall mortality (hazard ratio (HR) 1.39 per point increase; p = 0.01).

	Weighting		
Variable	**0**	**1**	**2**
Cardiothoracic ratio	<0.65	0.65-0.75	>0.75
Right atrial area index	<0.75	0.75-1.0	>1.0
Pulmonary forward flow	Normal	Reduced	Absent
Tricuspid regurgitation	No or mild	Moderate or severe	Moderate or severe
-TR Doppler gradient	-	≥ 40 mmHg	<40 mmHg
Pulmonary regurgitation	No	Yes	Yes
-Umbilical artery flow	-	Antegrade	Absent or reversed

Survival rates of 66 live-births at 1-month, 1-year and 5-years were 86%, 82% and 80% respectively, while 75%, 60% and 55% remained free from surgery at the same points in time. Factors associated with postnatal death by multivariate analysis included a younger gestational age at birth (HR per week: 1.59; p < 0.001), tricuspid annulus diameter (HR per z-score increase: 1.76; p = 0.004), and no pulmonary forward flow (HR 4.63; p = 0.03).
Conclusion: Our experience with fetal and neonatal EA/TVD shows better survival rates than previously reported. Mortality after a fetal diagnosis was significantly associated with hemodynamic changes indicative of a circular shunt, including pulmonary and tricuspid regurgitation severe enough to cause diastolic umbilical arterial flow reversal.

O2-5

Risk factors for heart failure-related deaths in childhood hypertrophic cardiomyopathy

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Background: A number of researchers have studied risk-factors for sudden death in paediatric hypertrophic cardiomyopathy (paedHCM), but considerably less is known about risk-factors for heart-failure related death in paedHCM.
Patients and Methods: A Swedish national cohort consisting of patients with paedCM presenting at age <19 years was assembled from all Pediatric Cardiology Centres in Sweden. There were 152 patients, mean follow-up 10.8yrs, in 26% associated with Noonan-spectrum syndrome. 15 patients who died from heart failure were identified; nine in infancy and six died in their teens or later, often with dilating left ventricle. Univariate and multivariate Cox-hazard regression have been used to identify risk-factors for heart-failure related death (CCF-death).
Results: Patients with CCF-death were diagnosed at a young age, 0.10 yrs [median; IQR 0.006-6.0], had generalized left ventricular (LV) hypertrophy with a high LV wall-to-cavity ratio both early (0.46 [0.31-0.57]), and at latest follow-up (0.57 [0.33-0.65]). Left atrium-to aortic (LA:ao) ratio was increased already early 1.72 [1.55-1.84]. 83% had definite or probable Noonan-spectrum syndrome, over-represented both in early and late deaths. LV outflow obstruction at rest (LVOTr) was present in 83%, and in addition in the right ventricular outflow (RVOTr) in 55%. On ECG 12-lead amplitude-times-duration product and ECG risk score were high already at diagnosis 3.51 [2.11-3.71] mV.s and 7 [6-10] points respectively. Risk factors significant on univariate analysis were: diagnosis at young age (p = 0.010), co-existing Noonan-spectrum syndrome (p = 0.004), left-ventricular wall-to-cavity ratio (early: p = 0.003; late: p < 0.001), increased posterior LV-wall thickness Z-score (p < 0.001), LV outflow-tract obstruction at rest (LVOTOr; p < 0.001), RVOTOr (p < 0.001), and LA:ao ratio at late follow-up (p < 0.001), initial 12-lead product (p = 0.005), and initial (p = 0.025) and final (p = 0.016) ECG risk score. On multivariate analysis many, including Noonan-syndrome, are shown to be linked to LVOTOr, leaving as independent risk factors: RVOTr (p = 0.013), LVOTOr (p = 0.048) and late LA:ao ratio (p = 0.017). Furthermore the only therapy that significantly reduced the risk for CCF-death on multivariate analysis was beta-blocker therapy (p = 0.030), whereas calcium-blocker therapy had no significant effect (p = 0.313).
Conclusions: Patients at high risk of CCF-death can be identified early, and outflow obstruction should be treated vigorously. Treatment should include beta-blockers rather than calcium-blockers.

O2-6

Global Left Ventricular Relaxation Index: a tissue Doppler indicator of positive biopsy in children post heart transplantation

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Background: Cardiac catheterization with endomyocardial biopsy (EMBs) is the standard for evaluation after orthotopic heart transplantation (OHTx). We developed an echocardiographic index of global left ventricular relaxation (LVRI) which in a previously reported study by our group demonstrated 100% sensitivity and 90.9% specificity for detecting OHTx patients with Grade 1 R to 3 R EMBs. In this study we tested the utility of the LVRI in an additional group of OHTx patients from a different institution in order to further evaluate our initial results. Both patient groups were combined.

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Methods: LVRI was calculated as the sum of diastolic tissue Doppler imaging (TDI) velocities (E') of the left ventricular lateral, septal and posterior walls divided by the percentage of the left ventricular posterior wall (LVPW) thinning by M-mode. In this expanded study LVRI was measured in 70 OHTx patients and 50 patients with normal hearts. Of the 56 patients who underwent clinically indicated EMB, 22 had Grade 0 R EMB, 18 had Grade 1 R and 16 had Grade 2 R to 3 R biopsies. Sensitivity, specificity and predictive value of LVRI for discriminating Grade 1R to 3R EMB were calculated. LVRI was compared before and after OHTx rejection treatment and during the early and late post-transplant period. One-way analysis of variance was used to compare all groups.
Results: LVRI was measured in all patients although identification of the E' velocity on OHTx patients with bi-atrial anastomoses was challenging compared to those with bi-caval anastomoses. In this larger patient population we confirmed that: 1) LVRI < 0.8 selected OHTx patients with Grades 1R to 3 R EMB; 2) LVRI was lower in patients with Grade 0R EMBs compared with normal subjects; 3) patients with Grade 1R to 3R EMBs had lower LVRI than those with Grade 0R EMBs; 4) LVRI recovered after rejection treatment; 5) LVRI normalized (LVRI ≥ 0.8) between 90-120 days post-transplantation with patient variability.
Conclusion: LVRI is a TDI index of global left ventricular diastolic function. We demonstrated, in an expanded transplant patient population, its utility as an echocardiographic indicator of positive biopsy and for assessment of the natural recovery of diastolic function after transplantation.

O3-1
Pulmonary arterial hypertension associated with pneumonia in children living at high altitude in China

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Objectives: Pulmonary arterial hypertension (PAH) may be associated with pneumonia, but has been paid little attention due to its mild form in those at sea-level. We have recently reported an elevated pulmonary artery pressure and hypertrophied and dilated right ventricle (RV) with decreased systolic and diastolic function in healthy children born and living in Qinghai (average altitude >3000 m). The degree of PAH was hypothesized to be more prominent and associated with RV morphological and functional changes in pneumonic children at high altitude.
Methods: Echocardiography was assessed in 104 children (1 month-3.5 years, median 6 months, 28 Tibetans and 79 Han who lived at 3845 ± 490 m and 2732 ± 741 m respectively, $p < 0.0001$) with acute pneumonia at Women and Children's Hospital in Qinghai from 2010 to 2014. Systolic pulmonary arterial pressure (SPAP), RV outflow tract (RVOT), dimension of RV and right atrium (RA), RV anterior wall (RVAW), tricuspid E (VETV) and A (VATV) wave peak velocities, E/ATV ratio and myocardial enzymes were measured at admission and after 20 ± 8 days at discharge.
Results: PAH was in 55 patients (51.4%), being mild (36.7 ± 4.9 mmHg) in 40 (37.4%), moderate (62.7 ± 7.5 mmHg) in 7 (6.5%) and high (105 ± 25 mmHg) in 5 (4.7%), with the highest being 149 mmHg. RVAW were thicker (2.8 ± 1.2mm, high altitude reference value was 2.0 ± 1.0mm) and RV dilated (11.6 ± 4.5mm, high altitude reference value was 10.0 ± 2.0mm) respectively. E/ATV ratio was normal (1.4 ± 0.5). Myocardial enzymes were within normal range. SPAP was not significantly correlated with altitude or ethnics ($P > 0.05$) but increased with age ($p < 0.0001$). RVAW and RV significantly increased with SPAP ($p < 0.0001$). SPAP became normal or mild hypertensive at discharge.
Conclusions: PAH occurs in about 50% of children suffering from acute pneumonia at high altitude, and moderate to severe PAH in about 10%, with dilated right heart and RV hypertrophy. Myocardial function appeared normal. Treatment strategies should be targeted on PAH in these patients at high altitude.

O3-2
Isolated right aortic arch prenatally detected: prevalence and post-natal outcome

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Introduction: Right aortic arch (RAA) is usually associated with congenital heart disease (CHD). In the absence of cardiac defects, the significance of RAA has not been determined.
Methods: 925 cases referred to our Fetal Cardiology Centre between January 2008 and June 2015 were retrospectively examined for RAA whether associated with CHD or isolated.
Results: Out of 361 cases with structural heart anomalies, 51 (14%) with RAA were detected. In 25 (49%) cases RAA was associated with CHD: 11 tetralogy of Fallot (TF), 7 pulmonary atresia and ventricular septal defect (VSD), 3 double outlet right ventricle, 2 tricuspid atresia, 1 VSD, 1 truncus arteriosus, 1 absent pulmonary valve with intact ventricular septum. RAA was not associated with CHD in 26 (51%) cases. The AD was left-sided in 20 (77%), 13 of these had Kommerel diverticulum and aberrant left subclavian artery. In 6 (23%) cases aortic arch and AD were both right-sided. Postnatal outcome was obtained in all cases. During a mean follow-up of 4 years, we observed three complications. Among isolated RAA and left-sided AD, one patient had resection of the arterial ligament at 18 months of age because of progressive symptoms of upper airway obstruction. Another case had a transient and asymptomatic pulse loss of the left upper limb. Among cases with RAA and right AD, one patient had non-confluent pulmonary arteries (PAs) and the left one being perfused by of a second left-sided AD, missed at prenatal examination. After closure of this AD the newborn developed asymptomatic thrombosis of left PA. At late diagnosis the patient was considered unsuitable for surgical reconnection of the disconnected PA.
Conclusions: Prenatal diagnosis of RAA is constantly increased. Nearly half of cases are associated with CHD (most with PA obstruction) and half are isolated. The majority of isolated RAA have a left-sided AD with Kommerel diverticulum and aberrant left subclavian artery. Despite a potential vascular ring in this subgroup, symptoms are very rare and surgical resection is needed only in a minority of cases. In RAA and right-sided AD, fetal and post-natal examination must rule out bilateral AD and disconnection of PAs.

O3-3
Outcomes of newborns with prenatal ventricular asymmetry and not operated after birth

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Objective: The objective of this study was to assess the outcomes of the neonates prenatally diagnosed with ventricular asymmetry and to determine the risk factors for left outflow obstruction occurrence at follow-up.

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Material and methods: All neonates with prenatal asymmetry of the ventricles and dominance of right heart structures, diagnosed from August 1993 to July 2015, un-operated within the neonatal period, were retrospectively included in the study. Left heart echocardiographic measurements at birth and at last follow-up were collected and compared. Left heart anomaly included isthmus and/or aortic valve and/or mitral valve obstruction.
Results: Among 34 newborns included in the study, 12 (35%) had associated cardiac lesions (5 ventricular septal defects, 6 hypoplasia of the aortic arch and 1 mitral stenosis). Median follow-up was 2 years (from 7 days to 27 years). There was no death. Eleven patients were operated on (32.5%) at a median time of three months, 7 of them had an obstruction of the left outflow tract. Freedom of left heart surgery was 80% and 75% at respectively 6 months and 10 years. Risk factors for surgery of left heart were a small aortic annulus (z-score -5.44 vs -2.24, $p = 0.002$), mitral valve malformation (50% vs 14%, $p = 0.04$) and anomaly of the left heart (42% vs 9%, $p = 0.024$). The main risk factor for evolution to left heart anomaly was an hypoplasia of the aortic isthmus (z-score -6.9 vs -1.5, $p = 0.0003$), while the presence of a left superior vena cava tended to be at risk for further left obstruction (50% vs 15%).
Conclusion: Despite no early postnatal coarctation of the aorta, antenatal diagnosis of ventricular asymmetry requires further monitoring for detection of left heart obstruction. Significant isthmus hypoplasia and the presence of left superior vena cava may represent risk factors for mid-term the need of mid-term left heart surgery.

O3-4
Features and prognostic factors of Scimitar Syndrome in children

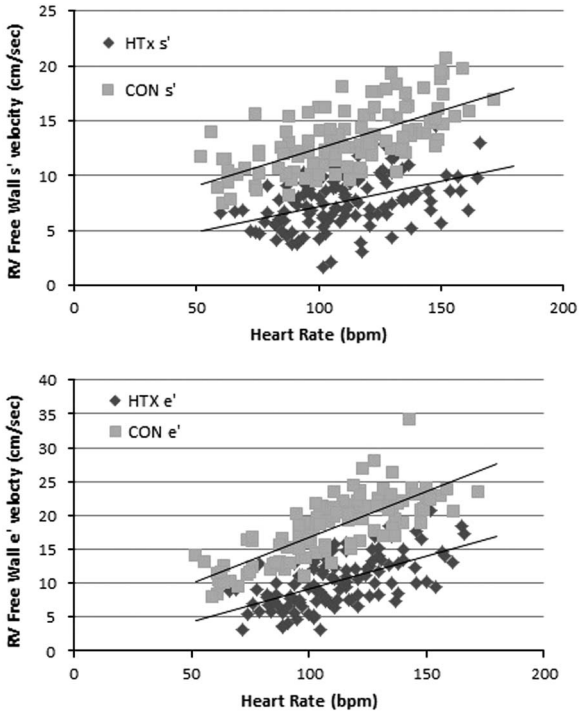
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Objective: the aim of this study was to assess the features of children with Scimitar syndrom and to determine factors for long term of outcome.
Methods: this is a retrospective two-centers study of all patients less than 18 years diagnosed with isolated SS. Demographic data were collected, echocardiographic measurements at diagnosis and last follow up, hemodynamic data when available. Prognostic factors for survival and bad outcome were analyzed.
Results: 52 patients (35 from Lyon center, France and 17 from Padova, Italy). Patients presented (median age 4.9 months) with respiratory symptoms in 48% or heart failure in 6% or were asymptomatic in 46%. Mean Z-scores for RV diameter, LV diameter, right PA and left PA branch were respectively +0.79, -0.6,-1.9 and +2.2. Mean pulmonary systolic pressure was 59 mmHg: 52.5% of cases had no or mild PHT and 47.5% moderate or severe PHT. Fifteen patients were operated on, 26 received medical treatment and 9 percutaneous embolization of systemic artery were performed at a median age of 0.6years. Nine deaths occurred (17%) at median age of 0.4years : mortality was 35.7% in neonates, 16.6% in infants and 5.5% in children. Median FU was 13.4years. Survival rates were 87% at 6months, 85% at 1 and 5years and 78% at 12years of FU.
Stenosis of the scimitar vein, neonatal onset, symptoms at onset, systemic artery and moderate/severe PHT were associated with worse survival ($p < 0.0001$). RV systolic pressure, PA systolic, dastolic and mean pressure were higher (respectively : 70 vs 39.6, 61.9 vs 33.3, 22.7 vs 13.5 and 38.1 vs 20.5 mmHg), and LV was lower (20.5 vs 29.9 mm) in deceased cases than in survivors ($p < 0.0001$).
Conclusion: Overall outcomes of children with Scimitar syndrome is favourable except in cases with very early onset symptoms and/or moderate/severe PHT and/or stenosis of the scimitar vein.

O3-5
Right ventricular systolic and diastolic response to exercise in children after heart transplant -a bicycle exercise study

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Introduction: Right ventricular (RV) dysfunction is a common problem after heart transplant (HTx). The mechanism of RV dysfunction after HTx is multifactorial, including donor and recipient factors. Several studies reported decreased RV function at rest in pediatric HTx recipients without acute rejection compared to healthy controls as studied by Tissue Doppler Imaging (TDI). However, information about RV function during exercise has not been well described in pediatric HTx recipients. The aim of the current study was to evaluate RV systolic and diastolic response to exercise in children after HTx using TDI during semi-supine cycle ergometry stress echocardiography (SSCE).
Methods: Twenty-five children after HTx and 23 controls were included. Median age at transplantation was 9 years (birth to 15 years) and median time since transplant was 4.9 years (0.5 to 15.4 years). A stepwise SSCE stress echocardiography protocol was used. TDI peak systolic and early diastolic velocities were measured in the RV free wall in all subjects at rest and at incremental heart rates (HR).



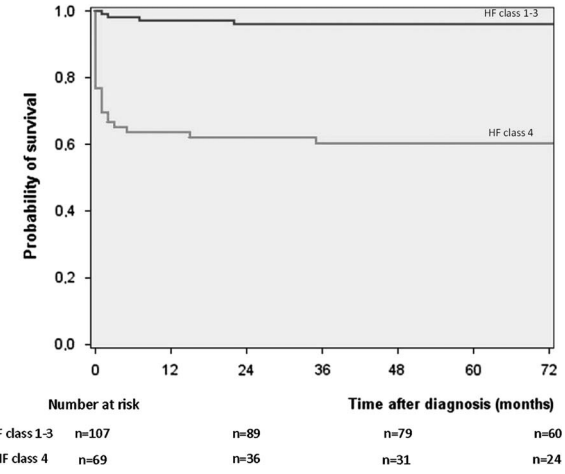
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Results: Resting HR (mean ± SD) was higher in the HTx recipients than in controls (88 ± 12 vs 70 ± 12 bpm, $p < 0.001$) but controls reached higher HR at peak exercise (142 ± 20 bpm vs 153 ± 11 bpm, $p = 0.02$). Systolic and diastolic RV free wall s' and e' velocities absolute values were significantly lower at rest and peak exercise in the HTx recipients compared to controls (Rest s': 5.4 ± 1.7 cm/s vs. 10.4 ± 1.8 cm/s, $p < 0.001$; rest e': 6.4 ± 2.2 cm/ s vs. 12 ± 2.4 cm/s, $p < 0.001$), (Peak s': 11.7 ± 9.3 cm/s vs. 16.2 ± 2.4 cm/s, $p < 0.001$; peak e': 13.3 ± 3.4 cm/s vs. 22.3 ± 3.3 cm/s, $p < 0.001$). When plotted versus HR the increase in TDI velocities showed a preserved response in children after HTx compared with controls (see figure).
Conclusions: Our data suggest that paediatric HTx recipients have preserved systolic and diastolic cardiac reserve during exercise compared with controls, despite lower systolic and diastolic RV TDI velocities at rest. Evaluation of cardiac function during exercise provides interesting additional information about cardiac function in these patients. The clinical implications of our findings need further investigation.

O3-6
Ebstein's Anomaly in Childhood: Factors at Time of Diagnosis associated with Death

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Objectives: To analyse long-term survival in paediatric patients (0-18 years) with Ebstein's anomaly and to assess the factors at time of diagnosis that are associated with death.
Methods: Records of all Dutch consecutive live-born patients diagnosed with Ebstein's anomaly between 1980 and 2013 were reviewed. Survival curves of the time between diagnosis and death were obtained using the Kaplan-Meier method. By using the Cox proportional hazard model we analysed the factors (at diagnosis) that are associated with death.
Results: We included 176 paediatric patients (89 males). The majority (112/176, 64%) was diagnosed in the neonatal period. The median time of follow-up after diagnosis was 70 months (range 0-216). Thirty-one patients (18%) died. The 1-year survival was 84%, from 35 months after diagnosis and onwards the survival rate remained stable at 82%. The 1-year survival for patients diagnosed in the neonatal period was 78% compared to 95% for patients diagnosed after the neonatal period. Nineteen patients (11%) with severe rhythm disturbances underwent catheter ablation of an accessory pathway, none of them died. Sixty-four patients (36%) underwent cardiac surgery; 10 of these patients (16%) died (all within 33 days after the surgery) compared to 21 patients (19%) in the non-surgical group. The median age of death was less than one month (range 0-49 months). The modified Ross Heart Failure class 4 at time of diagnosis was the most important factor associated with death (Hazard ratio, HR, 12.5; 95% Confidence Interval, 95% CI, 4.4-35.9). Furthermore, diagnosis in the neonatal period (HR 4.2; 95% CI 1.5-12.0), severe tricuspid valve regurgitation (HR 2.4, 95% CI 1.2-5.0), severe pulmonary valve stenosis or (functional) pulmonary valve atresia (HR 3.7, 95% CI 1.8-7.7) and a patent ductus arteriosus (HR 2. 8, 1.3-6.0) at time of diagnosis were univariately associated with death.



Conclusions: Heart Failure class 4 at time of diagnosis is the most important factor associated with death in paediatric patients with Ebstein's anomaly.

O4-1
Measuring invasive blood pressure by catheters guided solely by Cardiovascular Magnetic Resonance by using a new MRI-compatible guidewire without the need of a hybrid MRI-fluoroscopy suite

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Background: Blood pressure or blood pressure gradients cannot be evaluated accurately by routine cardiovascular magnetic resonance. However, blood pressure can be measured using invasive fluid-filled catheters guided by fluoroscopy in conventional catheter-laboratories. First clinical approaches have also been made using so-called hybrid cardiovascular magnetic resonance (CMR)-fluoroscopy suites. Therefore, the aim of this study was to test the feasibility of measuring blood pressure using fluid-filled catheters solely by CMR guidance without the need of a hybrid CMR-fluoroscopy suite.
Methods: Patients scheduled for routine clinical CMR and combined diagnostic and interventional catheterization by fluoroscopy were included into the study [10 patients with untreated or recurring coarctation of the aorta and 4 further patients with Fontan Circulation, RVOT-Conduit and pulmonary artery hypertension (2 female, median age: 23 years, range: 13 to 55 years)].
Results: Blood pressure was measured successfully by fluid-filled catheters guided solely by CMR using a guidewire (MRWire®, Nano4imaging, Aachen, Germany) CE certified for use in the MR environment. No guidewire-related adverse event occurred.
Conclusions: This study shows that invasive blood pressure can be measured relatively easily using fluid-filled catheters solely by CMR guidance without the need of a hybrid MR-fluoroscopy suite.

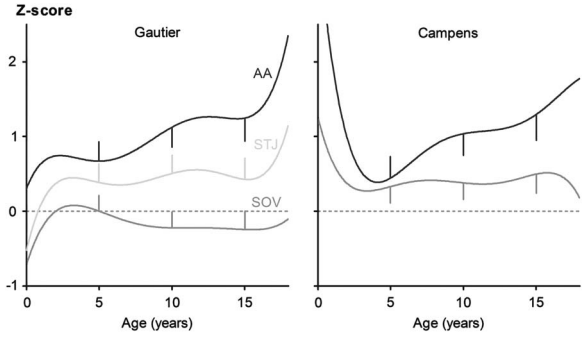
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O4-2

Aortic Diameter In Children With Bicuspid Aortic Valves

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Introduction: Prediction of aortic growth in patients with bicuspid aortic valve (BAV) is essential to identify patients at risk for dissection, but data in children are scarse.
Methods: We retrospectively identified all children with a BAV from an echocardiography database between 2005-2013. Medical records were reviewed and echocardiographic images re-measured. Aneurysm ($z > 2$) was based on two different z-score equation methods (Gautier et al. vs Campens et al.).
Results: In total 251 BAV patients were identified, mean age 5.8 years (64% males). Valve pathology (stenosis and/or regurgitation) occurred in 100% of Sievers' type 2, 62% of type 1 and 46% of type 0 ($p = 0.001$). In 234 patients aortic diameters were measured, mean follow-up was 4.7 years. The two z-score equation methods disagreed significantly for aortic sinus and, in patients below age 1.5 years for ascending aorta (AA). AA aneurysm was present in 24% (Gautier) vs 36% (Campens) at inclusion. Mean z-scores progressed significantly with age: 0.67 to 1.24 vs 0.45 to 1.29 at 5 and 15 years, respectively. Associations for AA growth were an initial z-score >2 ($p < 0.001$) and Sievers' type 2 ($p < 0.05$). Dissection or preventive aortic surgery did not occur. AA measurements on MRI and echocardiography agreed well (mean difference 1.1 mm, $p = 0.09$).
Conclusions: Aortic complications seem no major threat in paediatric BAV patients. Ascending aorta growth, based on existing z-score equations, is faster than expected. BAV morphology according to Sievers et al. seems predictive for valvular and aortic prognosis. MRI appears redundant for paediatric aortic diameter evaluation.

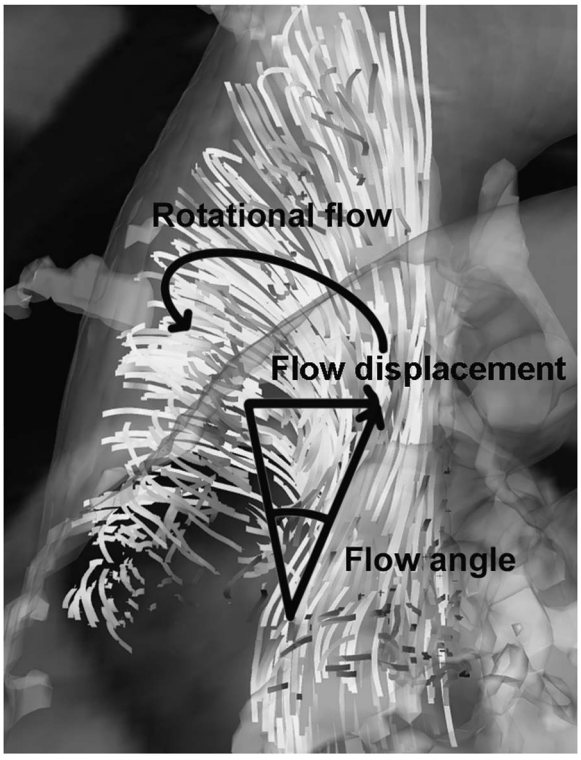


O4-3

Assessment of 4D flow imaging biomarkers in ascending aortic replacement

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Introduction: Bicuspid aortic valve disease (BAV) is associated with aortic dilatation and abnormal flow patterns, particularly increased helical flow and changes in the aortic wall shear stress. Aortic valve replacement is common and we sought to assess whether advanced imaging biomarkers can predict the need for additional ascending aortic replacement.
Methods: We prospectively enrolled 102 patients with BAV (age 8-72) who underwent 4D flow cardiovascular magnetic resonance. This included 59 BAV patients with a right-left coronary cusp fusion (RL-BAV), 33 with a right-non-coronary cusp fusion (RN-BAV) and 10 with other fusion patterns. We then followed this cohort with annual phone calls.
Results: During a 3 year follow-up period 20/102(19.6%) underwent aortic valve replacement (AVR); 10/59(16.9%) were in the RL-BAV group and 7/33(21%) in the RN-BAV group. Only 4/102(3.9%) underwent AVR with ascending aortic replacement (AVR+ AA). Final decision to include ascending aortic decision was made intraoperatively after visual assessment of the ascending aortic wall. Comparing patients in the AVR with the AVR+ AA group, at the initial study visit the AVR+ AA group had larger ascending aortic diameters/body surface area (22.9 ± 2.0 vs 18.8 ± 3.2 mm; $p = 0.014$), higher normalised displacement (0.205 ± 0.027 vs 0.1390.078 $p = 0.01$), absolute rotational flow (55.0 ± 9.2 33.7 ± 17.7 mm^2/ms; $p = 0.007$) and higher in-plane wall shear stress (0.89 ± 0.13 vs 0.54 ± 0.25 N/m^2; $p = 0.003$). Flow angle and overall wall shear stress changes did not reach statistical significance.
The patients in the AVR+ AA group had mainly stenotic disease. Comparing the AVR+ AA and only the stenotic patients in the AVR group the above changes remained significant.
Comparing the stenosis patients ($n = 14$) and regurgitant patients ($n = 6$) in the AVR group, only flow displacement was significantly lower in the regurgitant group (0.062 ± 0.032 vs 0.174 ± 0.065 vs; $p < 0.001$).
Conclusions: In BAV concomitant ascending aortic replacement is less frequently performed than AVR alone. Baseline flow abnormalities were comparable in the stenotic and regurgitant AVR group.
While ascending aortic diameters were significantly larger in the AVR+ AA group only one patient had a diameter >4.5 cm. The significantly increased haemodynamic flow abnormalities in the AVR+ AA group suggest that these may be useful parameters for risk assessment in the future.



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O4-4

Cerebral MRI findings and neurodevelopmental outcome in children before Fontan procedure at 2 years of age – Enlargement of liquor spaces influences outcome

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Objectives: Children with complex congenital heart disease undergoing neonatal cardiac surgery are at risk for neurodevelopmental impairment, especially in univentricular heart malformations. Neonatal cerebral MRI findings show pre- and postoperative cerebral injuries such as white matter injury and cerebral stroke. We focussed on the long term impact of cerebral injuries comparing cerebral MRI findings and neurodevelopmental outcome at two years of age.

Methods: In a prospective multicenter cross-sectional study we analysed consecutive patients with hypoplastic left heart syndrome (HLHS) or with UVH before Fontan procedure. Patients were palliated either by Hybrid or Norwood procedure. Neurodevelopmental outcome was investigated by Bayley Scales of Infant and Toddler Development III (Bayley III) and compared to findings of cerebral MRI scan before Fontan procedure.

Results: 48 patients (male 32; age mean 26.6 ± 3.8 months) with HLHS (n = 26) and non-HLHS (n = 22) were included. 44 patients were treated first by Hybrid procedure (n = 25), Norwood procedure (n = 7) and shunt or banding procedure without cardiopulmonary bypass (n = 12) before bidirectional cavopulmonary anastomosis (n = 48). In Bayley III Scales, median cognitive scale (CCS) 100 (range 65-120), language scale (LCS) 97 (68-124) and motor composite scale (MCS) 97 (55-124) were only inferior compared to norm data for LCS (Z = -2.2, p = .025). 36.2% (17/47) of patients showed disease-related intracranial lesions: isolated ventriculomegaly in 3, minimal (<2mm) white matter lesions in 5, cerebral stroke (less one third of vascular supply territory) in 9. The latter two findings where additionally associated with liquor spaces enlargement in 7 patients. Therefore, the enlargement of inner and/or outer liquor spaces was the most frequent pathologic MRI finding and was associated with lower scores in all subscales of Bayley III (CCS: p = .020; LCS: p = .002; MCS: p = .013).

Conclusions: Despite an overall good neurodevelopmental outcome before Fontan procedure 36.2% of patient exhibit intracranial lesions on cerebral MR Scan. Enlargement of inner or outer liquor spaces seems to be the major contributing factor of poor neurodevelopmental outcome. Improved perioperative management and neurodevelopmental follow up programs are needed in this patient group to detect neurological deficit early and to further improve quality of life.

O4-5

Identifying carriers of a mutation causing hypertrophic cardiomyopathy in childhood – M-mode echocardiography still the best simple technique

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Background: First-degree relatives of patients with hypertrophic cardiomyopathy (HCM) have a 50% risk of having inherited the same mutation and, as DNA analysis is expensive, simple screening measures that could estimate risk of mutation carriage at young age would aid determining clinical follow-up.

Patients and Methods: 60 individuals, mean age 8.4 years (SD = 6.0) from families with familial HCM with a known mutation and a confirmed DNA status were recruited. 30 mutation-positive (MP) and 30 mutation-negative (MN) children matched for gender and age, were compared in regards to ECG-features and echocardiographic measures, including tissue Doppler and 3D-left ventricular (LV) volume.

Results: ECG-findings: pathological Q-waves were present in 20% of MP, and 11% of MN individuals (p = n.s.), and there were no significant differences in 12-lead amplitude sums or 12-lead amplitude-times-duration products either. Echocardiography: There were no significant differences in diastolic function (E:A ratio, IVRT, E:e ratio, e:a ratio) or left atrial size, or in systolic function (fractional shortening). The LV cavity was however significantly smaller in MP than in MN (LV end-diastolic diameter Z-score p = 0.014; LV end-diastolic 3-D volume/m2BSA p = 0.028). M-mode septal thickness was significantly greater in MP than in MN group (Septum Z-score p = 0.0005; septum-to-cavity ratio (sepcavr) p = 0.00005; septum-to-posterior LV wall ratio (sepLVR) p = 0.0025) with some overlap between the groups in all measures. Screening performance was best in the last two measures. Sepcavr >95th centile for age (defined in Östman-Smith, Devlin, Eur J Echocardiography 2001; 2:22) gave a sensitivity of 50%, specificity of 93%, positive predictive value of 88% and negative predictive value of 66%, with corresponding figures for sepLVR >1.5 below three years of age, and >1.25 above three years of age being 43%, 83%,72% and 60%.

Conclusions: Long-axis M-mode is the best simple technique to predict mutation carriers. A sepcavr >0.27 at birth, > 0.24 by one year of age, >0.22 after two years of age, and >0.23 after eleven years of age has a high specificity of 93% for identification of mutation carriers, but sensitivity is only 50% since many children do not develop a pathological phenotype during childhood. The smaller LV cavities make sepcavr more sensitive than septum Z-scores.

O4-6

3D imaging of heart specimens: a new teaching tool for understanding the anatomy of double outlet right ventricle

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Introduction: One of the key success factors of the surgical treatment of complex congenital heart defects remains a detailed and in-depth understanding of the intracardiac anatomy. Currently, heart specimens are the best available teaching tools for studying the spatial relationships of the various components of the heart in complex congenital heart defects, like double outlet right ventricle (DORV). However, the access to anatomic collections is possible in only a few centres worldwide, and heart specimens may become damaged with time.

Objective: to study the intracardiac anatomy in DORV by using 3D imaging of heart specimens. The secondary objective is to build a database for teaching purposes.

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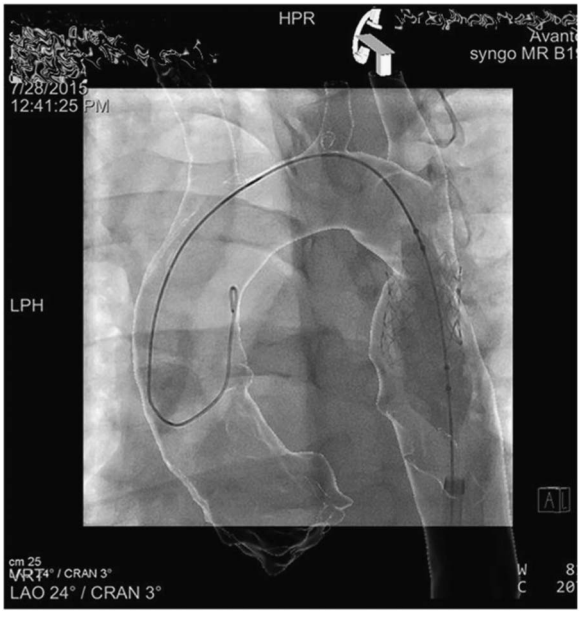
Methods and results: We performed CT scans with high resolution in 15 heart specimens with various anatomic types of DORV (with subaortic, subpulmonary and non-committed ventricular septal defect (VSD)). Heart specimens were fixed in 10% formalin. Openings were carefully stitched together before putting the heart in the CT scan. All 3D images were produced with a 3D reconstruction platform from Paris Descartes University. The 3D view from the right and left ventricles showed the anatomic details very neatly for all hearts. We described the VSD, its localization, borders and surface. We described also the relationship of the VSD with the aorta and the pulmonary trunk and the length and orientation of the outlet septum (figure). The ability to navigate through the heart cavities and vessels was very useful to understand the specific anatomy of the malformation.
Conclusion: This study underlines the role of 3D scan reconstruction as an imaging modality to increase our understanding of the anatomy of complex congenital hearts defects like DORV. This could constitute an innovative pedagogic approach, and a way to preserve the anatomic collections in the future.

O5-1
Radiation dose reduction in congenital heart disease patients during cardiac catheterization by a novel protocol
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Objectives: Cardiac catheterization remains as a major source of radiation exposure for patients with congenital heart disease, particularly for those patients with complex cardiac pathologies. As children are more prone to both deterministic and stochastic effect of radiation, every effort should be tried to reduce radiation exposure. One of the ways to reduce the radiation dose is to use lower pulse fluoroscopy rates. This study reports the magnitude of radiation exposure with 3.75 frame per second (fps) pulse fluoroscopy rate and compares the reduction with our previous 15fps protocol during cardiac catheterization for paediatric and adult congenital heart disease.
Methods: All diagnostic and interventional cardiac catheterization procedures from a single tertiary centre were analyzed from 1 January 2014 to 31 December 2015 which is 1 year before and after implementation of lower starting pulse fluoroscopy rates. Radiation dose was quantified as air kerma dose (mGy) and dose-area product (DAP; μGy/m2). Radiation exposure were analyzed for diagnostic and interventional procedures; diagnostic group was subdivided to cyanotic and acyanotic patients whereas interventional group was subdivided according to most common indications.
Results: A total of 786 procedure details were analysed. The mean fluoroscopy times and contrast amount were almost identical for both periods (621 vs 601 sec). In the first year of study (n = 371) the median air kerma for all procedures was 357.2 mGy, It was decreased to 104.60 mGy which means 70% reduction after lower pulse fluoroscopy rates during second year (n = 415). When we analysed procedures as interventional and diagnostic angiograms doses were 164.4mGy and 493.6mGy respectively in 2014 and 158,1mGy and 67.8mGy respectively in 2015. Among all patient groups the most striking decrease was observed in diagnostic procedures which we use prominently fluoroscopy rather than cine angiography.
Conclusion: We demonstrated a significant radiation dose reduction by implementing 3.75 fps pulse fluoroscopy rate which is the lowest possible rate and never reported before. We claim that novel radiation dose reduction protocols could be easily applied without any increase in fluoroscopy time and should be applied both for patient and health care provider safety.

O5-2
Next generation 3D-guidance for cardiac catheterization in congenital heart disease (CHD)
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Introduction: 3D-guidance in the cath lab for CHD is significant. The currently used 3D-models in VRT-format are not standardized and the commercially available registration needs an intra-procedural acquired 3D-dataset (Rotational angiography) with significant radiation.
Objectives: To evaluate feasibility and impact of tessellated 3D-models (triangulated surface-reconstruction) with new 2D-3D registration for now biplane 3D-guidance in catheterization of CHD.
Methods: Tessellated and segmented 3D-models (mesh in STL-format) are created from pre-interventional acquired MRI or CT by dedicated medical software (Mimics innovation suite). Accuracy of the tessellated models is evaluated in comparison to VRT-models. 2D-3D-Registration (between 3D-model and biplane fluoroscopy) was performed using a prototypic Siemens software solution (Monaco Workstation). Feasibility, accuracy of registration, visibility of the 3D-mesh and utility for intervention were evaluated in more than 50 cases.
Results: Measurements in STL-Meshes are significantly more accurate than in VRT-models. Overlaid on fluoroscopy visualization of STL-meshes are superior to VRT-Models. Nearly radiation-free 2D-3D-registration is practicable, using the trachea as fiducial marker.
Conclusions: 3D-meshes are the base for accurate, standardized and reproducible 3D-models; they can be used for 3D-roadmapping in the cath lab by 2D-3D-Registration. This technique has the potential to replace the rotational angiography in the pediatric cath lab.



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O5-3
Growth of the aorta in foetuses with evolving hypoplastic left heart syndrome after prenatal aortic valvuloplasty

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Objectives: Foetal aortic valvuloplasty(FBAV) may alter natural history of critical aortic stenosis(AS) and prevent progression to hypoplastic left heart syndrome(HLHS). Despite prenatal intervention substantial subset of patients ends up with univentricular circulation. The aim of this study was to analyse whether the FBAV facilitated aortic growth in foetuses who ended as HLHS, and therefore improved surgical conditions for 1st-stage repair.
Methods: Between 2012–2015 we evaluated foetal and newborns' echocardiographic examinations of 26 patients after FBAV and 37 without intervention(evolving HLHS with patent mitral valve and AS or atresia with visible aortic valve leaflets)who ended with univentricular circulation. Growth of the aortic annulus(AOV), ascending aorta(AAO), and isthmus(AOI) was analysed.
Results: All dimensions were larger in FBAV group pre-intervention and in last prenatal examination (appropriately 24.and 34.weeks in both groups). Postnatal data from 22 newborns after FBAV and 23 HLHS showed significant difference in the diameter of AOV and AAO, but not the AOI. Growth rate of measured structures, expressed as a difference in absolute values over time(mm/week) was significantly bigger in the intervention group for AOV (0.02 vs. 0.01, $p = 0.008$) and AAO (0.036 vs. 0.013, $p = 0.000007$), with no difference for AOI (0.014 vs. 0.016, $p = 0.53$). AOI diameter correlated better with size of the ductus arteriosus than with AOV in HLHS group.
Conclusions: Baseline aortic dimensions in foetuses suitable for the FBAV were larger than in those with HLHS. In spite of that, opening of the aortic valve seemed to be an important factor for the aortic growth, so update of indications for FBAV should be considered. In neonates AAO in FBAV group was almost within normal limits, contrary to HLHS. Lack of impact on AOI size may be due to its dependence on ductal rather than aortic flow.

Exam	Parameter	FBAVgroup	HLHSgroup	P
1st prenatal	AOV Z-score	-1.03 ± 0.98	-4.19 ± 2.13	<0.000001
	AAO Z-score	-0.60 ± 1.54	-4.56 ± 2.18	<0.000001
	AOI Z-score	-0.91 ± 1.18	-1.86 ± 1.28	0.0045
Last prenatal	AOV Z-score	-0.80 ± 1.30	-4.65 ± 2.53	<0.000001
	AAO Z-score	-0.57 ± 1.15	-5.19 ± 1.94	<0.000001
	AOI Z-score	-0.69 ± 1.31	-1.99 ± 1.32	0.0003
Newborn	AOV [mm]	5.4 ± 0.8	3.7 ± 1.0	<0.000001
	AAO [mm]	7.8 ± 1.7	4.1 ± 1.9	<0.000001
	AOI [mm]	4.1 ± 0.8	4.3 ± 1.0	0.52
	AOV Z-score	-2.65 ± 1.68	-6.72 ± 2.71	0.000001
	AAO Z-score	-1.75 ± 1.99	-8.44 ± 3.90	<0.000001
	AOI Z-score	-1.99 ± 1.09	-1.75 ± 1.32	0.55

O5-4
Novel Treatment for Post-Fontan Plastic Bronchitis by Direct Lymphatic Embolization

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Introduction: Plastic bronchitis is a rare, potentially life-threatening complication of Fontan procedure for single ventricle palliation. For these patients, the 5 year mortality rate is as high as 50%, increasing dramatically if the diagnosis is made less than one year post surgery. Current conservative treatment includes frequent bronchial lavage, anti-heart failure medications, anti-pulmonary hypertension medications, anticoagulation, and tissue plasminogen activator, DNase, N-acetyl cysteine, and hypertonic saline inhalations. Recent publications suggest selective lymphangiography and embolization as a promising new treatment option with good short term outcome.
Methods: We present a case of a 3.5-year-old boy with double inlet left ventricle, malposed great arteries, who developed severe plastic bronchitis 4 months post his Fontan completion. After recurrent life threatening episodes and failure of 8 month of conventional therapy, the patient was referred for lymphatic catheterization with direct embolization. Intranodal lymphangiogram was performed with injection of Lipiodol® (ethiodized oil-based contrast agent) into the inguinal lymph nodes. Under fluoroscopic guidance, the cisterna chyli was fluoroscopically accessed via anterior transabdominal approach. A guidewire and microcatheter were advanced into the thoracic duct and manipulated towards distal branches. Methylene blue injection into the lymphatic system resulted in observed staining of the right bronchial tree on simultaneous bronchoscopy. Embolization of distal lymphatic collaterals was performed by Lipiodol injection in the distal lymphatic vessels, followed by coil embolization of the thoracic duct and injection of glue.



Figure. Selective lymphangiography demonstrating abnormal dilated thoracic lymph vessels. These vessels were occluded using Lipiodol® injection with simultaneous balloon inflation in the left superior vena cava to avoid contrast embolization into the pulmonary tree.

Results: Post procedure the patient required mechanical ventilation and inotropic support for several days. He developed bilateral pleural effusions and moderate ascites that resolved with conventional treatment with no need for drainage. He was discharged

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home 18 days after the procedure. Currently, 2 months post intervention he is free of plastic bronchitis casts, only on anti-cardiac failure and anti-pulmonary hypertension medications.
Conclusions: Direct lymphatic embolization may be an effective novel treatment for plastic bronchitis with excellent short term prognosis.

O5-5
Atrial decompression in patients with HLHS and Hybrid approach by self expanding stents

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Background: A restrictive interatrial communication can be lifethreatening for patients with HLHS and hybrid approach. In some patients, recurrence of restricted interatrial communication can be observed despite initially successful ballon atrioseptostomy. We report our institutional experience performing atrial decompression by stenting the atrial septum.
Methods: From 2012-2015, atrial decompression was performed by stent implantation in 14 infants with HLHS and Hybrid procedure (Giessen procedure Stage I consists of bilateral pulmonary banding and PDA stenting). The median weight was 3.9 kg (2.7-5,38). Interatrial stenting was performed at an median age of 48days (5-116days). 7 Patients received ballon atrioseptostomy in prior interventions. In all patients self expanding stents (Sinus Superflex DS, Optimed) were implanted (5 x (8 × 12 mm), 3x (7 × 15mm) 6x (8 × 15mm)). In 3 patients additional stent dilatation was performed with 8 × 20 or 7 × 20 sterling ballon.
Results: The mean trans-septal gradient was reduced from 15 mmHg to 3 mmHg. In one patient dislocation of the stent occurred, which has been recovered without complications. This patient received a successful interatrial stent implantation a few days later.
Conclusion: Atrial decompression in infants with HLHS by self expanding stents (SS-DS) is a safe and effective method.

O5-6
Catheter intervention in complex congenital heart disease planned with the help of patient-specific computer models

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Introduction: Patient-specific models, based on each individual clinical images and data, can potentially be used to plan intervention for complex congenital heart disease. Overall to date, the translation of such computational tools has been limited to only single case reports. In this work, we report the results of our effort to develop a modelling framework that allows the use of realistic simulations of cardiovascular devices to prospectively predict clinical outcomes.
Methods: A small cohort of patients (n = 14) who were referred to our Centre for percutaneous pulmonary valve implantation (PPVI) and aortic coarctation stenting in complex anatomies was included in this study. Image data routinely acquired for clinical assessment (MRI, CT, echocardiography, x-ray) were post-processed to set up the patient-specific models of the implantation site together with functional characteristics. Various models of devices were virtually implanted and analysed in each patient-specific model by means of finite element and computational fluid dynamics simulations in order to predict structural and haemodynamic changes. The analysis results were presented during our clinical unit's multidisciplinary meeting where the optimal treatment strategy was decided. Measurable clinical outcomes from the real procedures were compared with the computer model predictions.
Results: A visual evaluation of many potential post-operative scenarios, as generated by computer simulations, supported the assessment of intervention feasibility. The computational outcomes provided additional quantitative information such as contact areas between device and implantation site, distributions of stresses on the vessel wall and flow data such as velocity and pressure fields. These measurements contributed to assess the different device options in each case and to indicate the optimal approach. Decision on the procedures including feasibility, choice of the device and size were in accordance with the computational predictions in all cases except one PPVI.
Conclusions: Computer simulations are a mature tool to predict outcomes of cardiovascular interventions. The early results of our Centre in translating patient-specific models to support treatment planning in complex cases of congenital heart disease are promising. These tools can provide information about the performance of existing devices and indications for the development of new ones.

O6-1
Growth in children with pulmonary arterial hypertension: a longitudinal multi-registry study

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Introduction: For an adequate interpretation of growth measurements in the clinical management of children with pulmonary arterial hypertension (PAH), there is a need for a description of growth in a large contemporary cohort. This study aimed to describe growth and its associated determinants in children with PAH.
Methods: A global longitudinal study of height and body mass index (BMI) in reference to WHO growth standards was conducted by pooling four contemporary prospective paediatric PAH registries representing 53 centres in 19 countries. Main outcome measures were median height-/BMI-for-age percentiles, percentage of patients below the 5th growth percentile and longitudinal deviation of height-/BMI-for-age Z-scores (HFAZ/BMIFAZ) from WHO standards. Determinants of growth were identified using linear mixed effects models.
Results: 601 children were followed for a median (IQR) of 2.9 (1.5-4.4) years and the total number of height and weight measurements was 4726 and 4932. Baseline median (IQR)

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height-for-age and BMI-for-age percentiles were 26 (4-54) and 41 (12-79). The number (%) of patients below the 5th percentile was 164 (27%) for height and 103 (17%) for BMI. Mean (95% confidence interval) HFAZ and BMIFAZ were significantly lower than the reference:-0.81 (-0.93 to -0.69); $p < 0.0001$ and -0.12 (-0.25 to -0.01); $p = 0.0466$. Figure 1 depicts means of HFAZ according to subgroups of age and aetiology. Although in individual patients increases and decreases occurred over time, in the total cohort there was no significant increase or decrease in HFAZ ($p = 0.5711$) or BMIFAZ ($p = 0.4776$) before taking account of covariates. Multivariable linear mixed effects modelling revealed that age, aetiology, ex-prematurity, WHO functional class, Trisomy-21, and time since diagnosis were associated with HFAZ, whereas age, ethnicity, and Trisomy-21 were associated with BMIFAZ. A favourable WHO functional class course was independently associated with increases in HFAZ ($p = 0.0070$).

Conclusions: PAH is associated with impaired growth. The degree of impairment is independently associated with aetiology and comorbidities, but also with disease severity and duration. As a favourable clinical course is associated with catch-up growth, height-for-age can serve as an additional and globally available clinical parameter to monitor the child's clinical condition in the management of paediatric PAH.

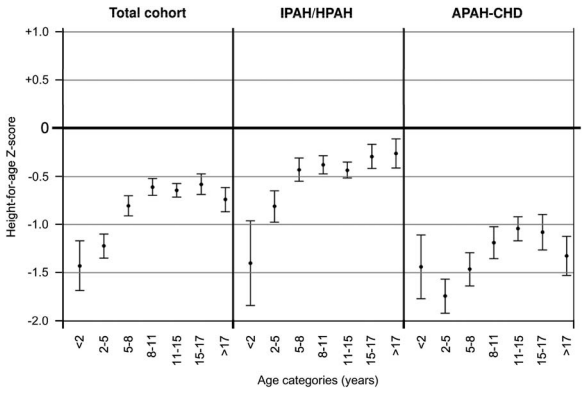


Figure 1. Mean height-for-age Z-scores within incremental age categories. Error bars represent 95% confidence intervals. IPAH = idiopathic pulmonary arterial hypertension, HPAH = hereditary pulmonary arterial hypertension, APAH-CHD = congenital heart disease associated pulmonary arterial hypertension.

O6-2

Composite clinical worsening endpoint in Paediatric PAH - clinically meaningful and feasible for clinical research

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Introduction: Clinical worsening (CW) composites are used in adult Pulmonary Arterial Hypertension (PAH) clinical research and is in discussion for future use in paediatric PAH research across all age ranges.

Methods: This study, using data from the Tracking-Outcomes–in-Paediatric-Pulmonary-Hypertension (TOPP) registry, describes the occurrence of individual outcomes and its CW composite: death, lung transplantation (LT), PAH-related hospitalisation (i.e. increased right heart failure, hemoptysis), atrial septostomy, deterioration in WHO functional class(FC) (change in 1 FC), initiation of parenteral prostanoids, syncope, PAH worsening (i.e. occurrence/progression of at least 2 symptoms: dyspnea, cyanosis, cough, fatigue, chest pain, dizziness). Predictive Cox proportional hazards models of time to death/LT were conducted for the aforementioned individual outcomes and CW composite (excl. death/transplantation).

Results: 255 incident (diagnosed ≤3 months) patients (i.e. idiopathicPAH/familialPAH, PAH associated with congenital heart disease) were included in the analysis. 155 (60%) were female and the mean (±SD) age at diagnosis was 7.5 (±5.2) years; 159 (62%) had iPAH/fPAH; 109 (43%) were in WHO FC II and 87 (34%) in class III. The highest incident rates per 100 person-years (95% CI) were observed for deterioration in WHO FC 24.8 (20.8, 29.5) and PAH related hospitalisation 18.3 (15.0, 22.4). In univariate models, first event of deterioration in WHO FC, (hazard ratio (HR) = 6.7; 95% CI 3.1,14.4), and first occurrence of PAH worsening (HR = 4.8; 95% CI 2.5, 9.0) were highly predictive of time to death/lung transplantation. PAH related hospitalisation and initiation of parental prostanoids had similar HRs.The predictive value of the occurrence of syncope did not reach statistical significance. The HR of the CW composite was 2.7 (95% CI 1.4, 5.3). The multivariate predictive models which included all individual variables (all univariate p-values < 0.15) revealed deterioration of WHO FC (HR = 3.5; 95% CI 1.5, 8.3), PAH related hospitalisation (HR = 2.6; 95% CI 1.3, 5.2) and occurrence of PAH worsening (HR = 2.1, 95% CI 1.0, 4.4) to be significant independent predictors of death/LT.

Conclusions: The chosen CW composite as well as the individual components (except syncope) occurred to be predictive for death/LT, thus supports the usefulness for clinical research but also for risk assessment during follow-up in paediatric PAH.

O6-3

Bilateral Lung Transplantation and PostOp VA-ECMO: A Novel Approach for Children with Endstage Pulmonary Arterial Hypertension (PAH)

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Introduction: Despite improvement in pharmacotherapy, bilateral lung transplantation (BLuTx) or combined heart-lung-Tx (HLuTx) remain the only established treatment options for children with endstage PAH. Although PAH is the second most common indication for BLuTx in children, data on the best perioperative management, pre- and postoperative cardiac function and mid/long-term outcome are lacking. We hypothesized

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that BLuTx, followed by early extubation and awake VA-ECMO, is associated with excellent short- and mid-term outcomes.
Methods: Retrospective study on children with PAH who underwent BLuTx at Hannover Medical School from January 2010 to September 2015. Pre-, peri- and postoperative data (demographics, echocardiography, cardiac MRI, CPR, ECMO, survival) were collected and analyzed.
Results: 7 consecutive patients with endstage PAH underwent minimal invasive BLuTx (mean age 13yrs; range 7-16yrs; diagnosis: 5 idiopathic, 1 after d-TGA arterial switch repair, 1 pulmonary capillary hemangiomatosis, i.e. group 1' PH). Average time on HU waiting list was 52days (range 1-130d); 3 patients were resuscitated prior to BLuTx, 2 of which were bridged to BLuTx on VA-ECMO. Intraoperative VA-ECMO or cardiopulmonary bypass was applied in 5 and 2 patients, respectively; 5 patients received scheduled post-BLuTx VA-ECMO support (mean duration 8.4days; range 6-12d). The goal "early extubation and awake VA-ECMO" could be achieved in 4/5 patients. Preoperative echocardiography and cardiac MRI showed severely compromised enlarged right ventricles (RV), i.e. suprasystemic or systemic RV pressure, endsystolic septal shift with LV compression, and pericardial effusion. Hemodynamic compromise rapidly improved after BLuTx, i.e. normalization of RV volumes and systolic function, and fade of LV compression and pericardial effusion. As of September 2015, all 7 patients are alive post-BLuTx, currently with a median survival time of 2.7years (range 0-5yrs).
Conclusions: BLuTx in children with endstage PAH is associated with encouraging results in our center. Post-op "awake VA-ECMO" might help facilitating RV and LV recovery, reducing perioperative and midterm mortality, and thus allowing better outcomes.

O6-4
Pulmonary arterial hypertension in children with transposition of the great arteries after successful neonatal arterial switch operation

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Introduction: Pulmonary arterial hypertension (PAH) has been sporadically described in children after neonatal arterial switch operation (ASO) for transposition of the great arteries (TGA). To describe epidemiology and clinical course of this particular concurrence, we present a series of children with PAH after neonatal ASO for TGA.
Methods: In nine dedicated pediatric pulmonary hypertension centers in Europe and North-America data were collected of children diagnosed with PAH after neonatal ASO (≤6 weeks after birth) for TGA between 1989 and 2014. Children with significant residual shunt-defects were not included.
Results: Twenty-five children were identified (median age of ASO 8 days). Six children (24%) had a concomitant ventricular septal defect. In 14 children (56%), PAH was diagnosed within six months after ASO. The remaining children were diagnosed after a median of 30 months (interquartile range: 14–93 months). Twenty-three children (92%) received PAH-targeted therapies during their disease course. During a median follow-up after ASO of 5.1 years, 2 children received a Potts shunt, 4 children underwent lung transplantation and 8 children died. One-, 5-, 10-and 15-year Potts shunt- and lung transplantation-free survival after ASO was 100%, 73%, 65% and 30%, respectively. Of the survivors without lung transplantation or Potts shunt (median follow-up after ASO 4.7 years), all but one child were in World Health Organization Functional Class I-II at last follow-up.
Conclusions: PAH after successful neonatal ASO for TGA represents a specific disease entity with a putative incidence of 0.5-0.9% of children that undergo ASO for TGA. Onset of PAH varies from directly after ASO to first detection only in adolescence. Its clinical course varies from rapid deterioration and death to a prolonged course in good clinical condition. Since prolonged exposure to unfavourable hemodynamics is not present in these children, specific prenatal pulmonary hemodynamics in TGA or genetic make-up may play a role in the typical development of PAH in these children.

O6-5
Pharmacological management of paediatric heart failure: results of a European survey

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Background: Paediatric heart failure (HF) has an important economic and social impact in public health. Drugs acting on the renin-angiotensin system are regarded as mainstay to lower the burden of HF for patients and families. A safe and efficient use especially in young children has been debated since several years and remains a challenge for physicians.
Aim: To characterise the different therapeutic strategies for the management of paediatric HF that are currently practiced across Europe with special focus on the use of Angiotensin Converting Enzyme Inhibitors (ACE-I).
Methods: A Europe-wide web-based survey was developed in the context of EU's Seventh Framework Programme under grant agreement n°602295 using standard recommendations for survey design. The questionnaire consisted of 23 questions addressing different aspects of drug therapy for HF in children. Use patterns of ACE-I i.e. dosage by age group, effectiveness and toxicity assessment according to HF aetiology were investigated. Clinicians from 204 different hospitals of 39 European countries were invited via e-mail to participate.
Results: Survey was conducted between January and May 2015 achieving a response rate of 50%. All participants reported using ACE-I for the management of at least one type of HF. Captopril was preferred for newborns (73%) and infants and toddlers (66%), whereas enalapril was the first choice in older age groups (57% for children, 59% for adolescents). Lack of consensus among survey participants was observed regarding benefits of drug treatment

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depending on aetiology and stage of HF or concerning the optimal ACE-I maintenance dose. Regarding safety parameters, up to 74% of the participants claimed to follow serum creatinine increase for decision-making when deterioration of renal function is detected. Selection of cut-off points for serum creatinine differed.
Conclusions: This survey provides an overview of the clinical treatment routine of paediatric HF across Europe. ACE-I seem to be a crucial part of the treatment strategies. Nevertheless, marked variability exists regarding the effective and/or safe use of this drug class. The results may help to start a consensus discussion about a standardized use of ACE-I treatment to provide guidance for an efficient and safe use of ACE-I in children with HF.

O6-6
Effect of bosentan in adults with pulmonary arterial hypertension due to congenital heart disease with and without Down's syndrome
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Background: Oral bosentan is effective in pulmonary arterial hypertension (PAH) related to congenital heart disease (CHD). In literature, the effect of bosentan in patients with Down's syndrome is largely unknown. Aim of the study was to evaluate the long-term effects of bosentan in adult patients with CHD-related PAH with and without Down's syndrome.
Methods: WHO functional class, resting oxygen saturation, 6-minute walk test (6MWT) and hemodynamics were assessed at baseline and after 24 months of bosentan therapy in patients with CHD-related PAH with and without Down's syndrome.
Results: Ninety-five consecutive patients were enrolled: 25 with and 70 without Down's syndrome. After 24 months of bosentan therapy, both with and without Down's syndrome patients showed an improvement in WHO functional class (Down: 2.4 ± 0.5 vs 2.8 ± 0.6, $p = 0.005$; controls: 2.5 ± 0.5 vs 2.9 ± 0.5, $p = 0.000002$), 6-minute walk distance (Down: 281 ± 66 vs 232 ± 69 m, $p = 0.0007$; controls: 383 ± 75 vs 336 ± 81 m, $p = 0.00003$), and hemodynamics (pulmonary flow, Down: 3.7 ± 1.5 vs 3.2 ± 1.3 l/m/m^2, $p = 0.006$; controls: 3.2 ± 1.3 vs 2.5 ± 0.9 l/m/m^2, $p = 0.0005$; pulmonary to systemic flow ratio, Down: 1.3 ± 0.6 vs 09 ± 0.3, $p = 0.003$; controls: 1.0 ± 0.6 vs 0.8 ± 0.2, $p = 0.012$; pulmonary vascular resistance index, Down: 15 ± 9 vs 20 ± 13 WU.m^2, $p = 0.007$; controls: 20 ± 10 vs 26 ± 15 WU.m^2, $p = 0.002$). We did not find any difference in the efficacy of therapy between the two groups.
Conclusions: Bosentan was safe and well tolerated in adult patients with CHD-related PAH with and without Down's syndrome during 24 months of treatment. Clinical status, exercise tolerance, (evaluated by 6MWT), and pulmonary hemodynamics (evaluated by right heart catheterization), improved, regardless of the presence of Down's syndrome

O7-1
Long-term Follow up after the Norwood procedure
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Objectives: Changes in operative techniques have brought significant improvement of early postoperative results after the Norwood procedure. This study is meant to examine long-term follow up concerning mortality and morbidity after the Norwood procedure regarding several treatment methods.
Methods: A single center retrospective study was done using data from the center's database. 317 consecutive patients who underwent a Norwood procedure between 1997 and 2014 were included. Patients were followed until september 2015 and the following parameters were analysed for the 3 stages of palliation: mortality, interstage interventions and longterm morbidity during the Fontan circulation.
Results: The mortality rate until the BDG was 18.9% (30 days mortality 13.2% and late interstage mortality 5.7%). The patient group with right sided RV-PA conduit (181 patients) showed the best results with 30 days mortality of 7.8% and late mortality of 4.4% compared to patients with modified BT-Shunt or left sided RV-PA conduit. The BDG operation was done in 254 patients at a median age of 121 days. 30 days mortality was 2%. Mortality in the interstage II was 10.2% with median age at death of 328 days and chronic cardiac failure in 65% of cases;1 HTX. Extracardiac Fontan operation was done in 176 patients (86% fenestrated) at a median age of 3.44 a with a 30 days mortality of 0.6%. During the follow up between 10 m – 17.5 a 5 patients died late after Fontan, 2 HTX. 6.3% of the Fontan patients showed PLE, 2.8% plastic bronchitis, 6,8% thrombi during follow up. Additional reoperations were necessary in 11.04% during interstage I, 3.5% during interstage II and 7.4% in Stage III. Interventions took place in 7.89% of pat. during interstage I, in 15.35% in interstage II and in 10.23% of Fontan patients.
Conclusions: Survival rates improved significantly during the observation period favouring the group with right sided RV-PA conduit showing a 5 years survival expectancy of 78.1% (95%-CI: 71.8-85.0%). Mortality rate is highest in stage I, still remarkable in stage II, but low in Stage III. The summarized reoperation and reintervention rate apart from the 3 stage palliation remains high.

O7-2
Does arterial switch for d-transposition of the great arteries alter myocardial deformation of the ventricles?
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Introduction: The arterial switch operation (ASO) is currently the surgical technique of choice for repair of d-transposition of the great arteries. The main pulmonary artery is moved forward (Lecompte maneuver) and its branches are stretched on either side of the ascending aorta. The coronary arteries are removed from and reinserted into the aorta. We sought to assess myocardial deformation changes in the right (RV) and left ventricles (LV) as signs of subclinical myocardial functional impairment after ASO and re-positioning of the coronary arteries.
Methods: Patients after ASO and normal controls underwent cardiac magnetic resonance (CMR) imaging including 2D SSFP for ventricular function. 2D SSFP cine images were post-processed with a feature tracking software (TomTec 2D CPA). Global circumferential strain was measured on short axis mid-ventricular slices and global longitudinal strain on horizontal long-axis images, separately for each ventricle. Patients with pulmonary arteries stenoses or history, symptoms, or CMR findings suspicious for coronary compromise were excluded.
Results: Eighteen patients after ASO (age 16.8 ± 6.7y) were compared to 18 normal controls (age 22.2 ± 11.4y; $p = 0.098$). RVs of ASO patients showed lower longitudinal strains (-14.1 ± 6.4% vs. -18.3 ± 3.8%; $p < 0.05$) but higher circumferential strains

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(-16.6 ± 3.2% vs.-13.1 ± 4.3%; $p < 0.01$) compared to normal RVs. LV longitudinal strain (-15.4 ± 5.1% vs.-17.5 ± 4.6%; n.s.) and LV circumferential strain (26 ± 5.6% vs.-23 ± 13.1%; n.s.) were not significantly different in patients vs. controls. There were no differences between ASO patients and controls regarding ejection fractions of the RV (54 ± 6% vs. 52% ± 5%; n.s.) and LV (58 ± 8% vs. 60% ± 5%; n.s.) or regarding end-diastolic volumes of the RV (91 ± 21 ml/m^2 vs. 94 ± 12 ml/m^2; n.s.) and LV (87 ± 26 ml/m^2 vs. 80 ± 11 ml/m^2; n.s.) indexed to body surface area, respectively.
Conclusions: LV deformation is preserved after the ASO operation, despite coronary artery surgery. In contrast, even in the absence of significant pulmonary artery stenosis, RV deformation is altered with decreased global longitudinal strain and increased circumferential strain, while preserving RV volume and ejection fraction. This may be the result of abnormal ventriculo-arterial coupling after the Lecompte maneuver and its changes in the outflow tract geometry.

O7-3
New Insights into Surgical Anatomy of Double Outlet Right Ventricle – An Echocardiography and 3D Model correlation study

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Introduction: Within the umbrella diagnosis of double outlet right ventricle (DORV), there is considerable heterogeneity of intracardiac anatomy. Precise 3-dimensional (3D) understanding of anatomy is crucial as complicated VSD baffling to an arterial trunk is often required. 3D-print cardiac models can provide a visual roadmap to assist with surgical planning.
Objective: To correlate transthoracic echocardiography (TTE) with 3D-print models and intra-operative findings and analyze discrepancies in the pre-surgical diagnosis between modalities. We sought to determine if 3D-print models affected anatomic diagnoses and decision-making.
Methods: Since 2009, 40 3D-print models with DORV were provided for clinical decision-making. We retrospectively reviewed TTE studies, models, intra-operative findings and final operative decisions.
Results: Anatomic characteristics included situs abnormalities (10%), dextrocardia (18%) and outflow tract obstruction (78%). Ventricular septal defects (VSD) were remote in 23%. Great artery relationships were malposed in 63%. There was a degree of discrepancy in 25% of cases between TTE and findings shown with 3D-print models and surgical reports, including inaccurate VSD description (13%), its commitment to arterial valve/s (8%) and mechanism of outflow tract obstruction (5%). The most significant contribution of 3D-print models was considered the apparent demonstration of the route for intraventricular baffling when echocardiographic findings were not completely conclusive. 3D-print models clearly showed the extent of the tricuspid valve annulus bordering the VSD margin, alignment of the VSD to arterial valve/s and extent of the muscular infundibulum. However, information regarding valves, chordae and papillary muscles was insufficient in 3D models.
Conclusion: 3D-print models provided indisputable information regarding cardiac anatomy, especially when the VSD involved the ventricular inlet or if the infundibulum was excessively long. Correlation of echocardiography with 3D models enhanced understanding of surgical anatomy and, therefore, improved surgical decisions with a higher level of confidence.

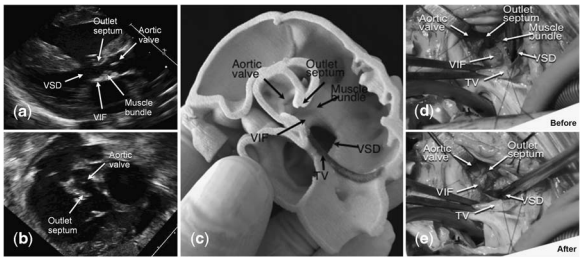


Figure: (a,b) Transthoracic echocardiographic images and (c) 3D-printed model of DORV showing subaortic obstruction from muscle bundles and posterior deviation of the outlet septum, with persistence of the ventricular infundibular fold (VIF). The ventricular septal defect (VSD) is aligned with the aortic valve. Intra-operative images before (e) and after (f) biventricular repair with subaortic resection and VSD closure. TV: tricuspid valve.

O7-4
Influence of aortic arch anomalies on long-term outcome of patients with TGA-VSD after the arterial switch operation

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Objectives: Early results after the arterial switch operation (ASO) for patients with simple transposition of the great arteries (TGA) are excellent, but studies on patients with an additional ventricular septal defect (VSD) are scarce. We aimed at analysing the long-term outcome of this subgroup of patients, focussing on reoperations and the influence of associated anomalies on outcome.
Methods: All patients with TGA-VSD, operated between 1983 and 2014 were included in a retrospective study. Study endpoints were survival and reoperation after the ASO. A reoperation was defined as a major cardiac procedure.
Results: A total of 207 patients with TGA-VSD underwent an ASO during the study period. Associated anomalies were multiple VSDs in 22 (10%), hypoplastic aortic arch (HAA) in 17 (8%), coarctation in 15 (7%), and interrupted aortic arch (IAA) in 6 (3%) patients, respectively. Early mortality was 6%, late mortality was 4%. Freedom from death was 93 ± 1.8% at 1 year and 87.9 ± 2.6% at 20 years. During a mean follow-up time of 12.1 ± 9.2 years (maximum 31 years), a total of 36 reoperations were required in 24 patients (11%). Freedom from reoperation was 92.3 ± 2% at 1 year and 73.9 ± 4.3% at 20 years. Reoperations were performed for right ventricular outflow tract obstruction in 20 (9%), residual VSD in 11 (5%), aortic insufficiency in 7 (3%), and for aortic obstruction in 7 patients (3%), respectively. Long-term survival was not significantly different for patients requiring a reoperation ($p = 0.6$) and for patients with associated anomalies ($p = 0.1$). Patients with associated anomalies required more often a reoperation than patients without associated anomalies ($p < 0.001$). Independent risk factors for death were age greater than 6 months at ASO and concomitant aortic arch repair ($p = 0.01$). Independent risk factors for a reoperation were multiple VSDs ($p = 0.03$), HAA ($p = 0.001$), IAA ($p = 0.02$) and the use of a pericardial patch for reconstruction of the coronary buttons ($p = 0.02$).
Conclusions: Long-term outcome after the ASO for TGA-VSD are excellent, but major reoperations are necessary in about 11% of the patients. Patients with associated anomalies, especially aortic arch anomalies, have a higher risk for death and for reoperation in the long-term.

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O7-5

Staphylococcus aureus adhesion to tissues used for RVOT reconstruction under static and shear stress conditions

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Introduction: Patients after repair of congenital heart disease (e.g. Tetralogy of Fallot) frequently develop dysfunction of the right ventricular outflow tract (RVOT). Implantation of cryopreserved pulmonary homografts, bovine jugular valved conduits or stent-mounted valves are often used for reconstruction. This improves patients' quality of life but inherits an increased risk of infective endocarditis (IE) often caused by *Staphylococcus aureus*. In this work, we focus on the susceptibility of different graft tissues to *S. aureus* adherence.
Methods: Standardized tissue pieces prepared as for clinical use (cryopreserved pulmonary homograft, bovine jugular venous conduit and pericardium patch) were mounted in a 6-well plate and incubated for 1 h at 37°C with 10^7 CFU/mL of *S. aureus* Cowan (labelled with carboxy-fluorescein) for static adhesion and adherence under laminar shear stress of 10 dyne/cm^2 in a flow chamber. Bacterial adhesion was confirmed using the fluorescence microscope IN Cell Analyzer 2000 (GE Helathcare) and quantified by CFU count on blood agar plates (expressed as Mean Log CFU/mL ± SD). Tissue pieces were sonicated in 1 mL of 0.9% NaCl for bacterial detachment and serial dilution were spotted on blood agar plates. Immunohistochemistry was also performed in fixed tissue pieces.
Results: Using the IN Cell Analyzer 2000 we visualize equal bacterial attachment to all tested tissue surfaces. The graft tissues showed similar susceptibility for bacterial adhesion in static condition with an average of 3.52 ± 0.31 Log CFU/mL ($P > 0.05$, one-way ANOVA). Moreover, the shear stress increased significantly bacterial adhesion for all graft tissues (4.81 ± 0.01 Log CFU/mL; $P < 0.05$, one-way ANOVA) although there was no difference among them ($P > 0.05$, one-way ANOVA). The immunohistochemistry revealed a 'single-cell like' bacterial attachment in static conditions and a 'cluster like' pattern under shear stress.
Conclusions: Our results indicate that all graft tissues tested have similar susceptibility to be colonized by *S. aureus*. The important influence of shear stress in bacterial adhesion has been verified. It seems that the tissue surface itself has no significant influence on bacterial adhesion, however it might modulate endothelial cell adhesion and inflammatory responses that are crucial for IE development. Our following studies will focus on these aspects.

O7-6

Fate of pulmonary artery branches after hybrid palliation for hypoplastic left heart syndrome: univentricular palliation versus biventricular repair

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Objectives: To revise midterm outcome of PA in patients who underwent neonatal hybrid stage I palliation and either subsequent comprehensive Norwood stage I-II (UVP) or biventricular repair (BVR). We focused on the overall freedom from re-intervention and on the potentially different impact of hybrid on PA branches development between UVP and BVR.
Methods: Since October 2011, 44 consecutive patients underwent hybrid stage I in our Institution. Twenty-two patients (50%) had surgical stage II and have been regularly followed-up for at least 8 months. Eleven patients had UVP while the remaining 11 had BVR. Patients' and surgical characteristics were homogeneous. Of note, in the biventricular group 5 patients had borderline left ventricle and would have not been eligible for biventricular repair at birth.
Results: Mean follow-up was 33.6 months (18-43) and 22.1 months (12-31) for UVP and BVR group, respectively. Freedom from re-intervention was 9.6 months (2 days – 28) and 7.5 months (3–13) in UVP and BVR group, respectively.
All 11 patients who survived UVP required re-intervention on PA branches. PA surgical enlargement only was adopted in 2 patients while interventional catheterization was performed in 9 patients. Eighteen interventional procedures were performed, consisting in balloon dilatation (12 cases) and PA stenting (6 cases). Left pulmonary artery was more hypoplastic and 6 out of 7 stents were implanted on left PA.
In the BVR group 4/11 patients (36%) needed re-intervention. Balloon dilatation was performed in all patients, one requiring also left PA stenting. In one case surgical enlargement was performed for concomitant occurrence of left ventricular outflow tract obstruction.
Conclusions: Hybrid palliation clearly demonstrated to be a reliable bridge to both UVP or BVR in neonates. However, this approach is associated with a high incidence of PA branches morbidity. According to the literature, need of re-intervention is particularly significant in patients with UVP, apparently due to the bulk of the aorto-pulmonary anastomosis. Conversely, in the biventricular group PA branches morbidity has a lower impact on outcome and, particularly in case of borderline left ventricle, PA disease is clearly less significant compared to the benefit of a delayed biventricular repair.

O8-1

A valuable tool in predicting poor outcome due to sepsis in pediatric intensive care unit: Tpeak-end/QT ratio

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Objective: To assess the feasibility of 12-lead electrocardiographic measures such as P wave dispersion (PWd), QT interval, QT dispersion (QTd), Tpeak-end interval (Tp-e), Tp-e/QT and Tp-e/QTc ratio in predicting poor outcome in patients diagnosed with sepsis in pediatric intensive care unit (PICU).
Methods: Measurements and Main Results: The study included 102 consecutive patients were enrolled into the study based upon diagnoses of systemic inflamatuar response syndrome (SIRS), sepsis, severe sepsis, and septic shock. The control group consisted of 103 age- and sex-matched healthy children. Patients' demographic features, capillary refill time, C-reaktif protein, procalcitonin, lactate values at the time of diagnosis were obtained from the hospital records. PWd, QT interval, QTd, QTcd, Tp-e interval, Tp-e/QT, Tp-e/QTc ratios were calculated in 12-lead-standart electrocardiogram at a speed of 25 mm/sec and an amplitude of 1 mV/cm.

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Results: We found that PWd and QTd were higher in the study group ($p < 0.001$). Tp-e interval, Tp-e/QT, Tp-e/QTc ratios were also significantly higher in septic patients compared to the controls ($p < 0.001$, $p < 0.001$, and $p = 0.003$ respectively). We found that of the 102 patients, 9 (8.8%) were diagnosed with SIRS, 35 with sepsis (34.3%), 39 with severe sepsis (38.2%), 19 with septic shock (18.6%). These subgroups were compared in terms of ECG carecteristics and we found no statistical differences between them. During the follow-up period 60 (58.8%) patients were cured but 42 (41.2%) had died. Deceased children had statistically significant higher Tp-e/QT, Tp-e/QTc ratios than the survivors. In multivariate logistic regression analysis only Tp-e/QT ratio were found to be independent predictors of mortality ($p = 0.016$, OR: 2.12; 95% CI:1.31-3.98).
Conclusion: This study demonstrated that the ECG measurements can predict the poor outcome in patients with sepsis. The Tp-e/QT ratio may be valuable tool in predicting mortality for patients with sepsis in the PICU.

O8-2
The Two Center Study Of Flecainide Efficacy And Safety In Childhood Arrhythmias With And Without Structural Heart Disease

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Introduction: Historical CAST study findings in adults have led to concern about the safety of flecainide in the presence of structural heart disease (CHD), ischemia and impaired cardiac function. Extrapolation of an adult study to paediatric patients has been questioned in the recent era with its safe use in fetuses and children. We aimed to review the effectiveness and safety of flecainide use in two centres in the treatment of paediatric arrhythmias associated with normal hearts (NH), structural heart disease (CHD) and cardiomyopathy (CMP).
Methods: We reviewed all patients receiving flecainide from 01/2005-07/2015 at two paediatric cardiology sites. Age at arrhythmia presentation, duration of arrhythmia treatment, ECG findings, patient outcomes and cardiac function were recorded. Flecainide efficacy, toxicity, and proarrhythmia rates were analysed.
Results: There were 175 patients, 22 had CHD/CMP. A significant QRS widening (55% vs. 22%, $p < 0.0001$) or QTc prolongation (22% vs. 8%, $p = 0.05$) occurred more often in CHD/CMP patients. Moderate to severe cardiac dysfunction was observed more commonly in the CHD/CMP group compared to NH (50% vs. 17%, $p = 0.002$) at baseline. Three patients (1.7%) showed a decline in cardiac function secondary to CHD ($n = 2$) and incessant SVT ($n = 1$). Flecainide was discontinued only in one (5.3%) patient in CHD/CMP from centre one due to worsened LV function. Flecainide levels were abnormal in 9 cases and of which 2 developed wide complex tachycardia. There was no difference in proarrhythmia between the CHD/CMP and NH groups (5.3% vs. 4.0%, p = NS). One infant died from other causes that was not linked to flecainide. Arrhythmia control did not differ between groups: CHD/CMP success = 19 (86%) vs. NH success = 116 (76%). CHD/CMP unsuccessful = 3 (13.6%) vs. NH unsuccessful = 35 (23%). Centre one success rate was 39 (52%) vs. centre two 92 (92%).
Conclusion: Flecainide was well tolerated in this cohort with less than 3% discontinuation of medication due to adverse outcomes. There was no mortality even in CHD/CMP group. Flecainide success rate in controlling arrhythmia was higher in cases from the centre two. A larger study is warranted to further validate the safety and effectiveness of flecainide.

O8-3
Predictors of Successful Catheter ablation of Ventricular Arrhythmias in Children: 12 Years Experience in 532 Pediatric Patients

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Objective: to determine the efficacy of catheter ablation (CA) approach based on evaluation of the endocardial mapping (EM) results and prediction of the treatment success in children with ventricular arrhythmias (VA).
Methods: In 2003-2015 532 pts (213-females) aged 5 to 18 yo received CA for VA in one hospital. Pts were divided into two groups depending on the selected mapping approach of VA. Group 1: 119 pts, 8-17 yo (54 – females): choice of a site for CA was based on the conventional mapping. Group 2: 413 pts, 5-18 years (159 – females). In these pts, a site for CA was chosen according to the calculation of the probability for successful ablation: if presystolic activation time (T) < 28 ms, then the probability of successful CA was considered low. In case of T was 29-72 ms - moderate. If T > 72 ms - high. Efficacy of CA, fluoroscopy time, effective dose, and the CA time were evaluated.
Results: The change in the approach to EM significantly affected the efficacy of CA of VA (ORi = 3.22; CI = 1.94-5.35; $p = 0.00001$). The efficacies of CA of VA were 73.11% and 89.83% in group 1 and group 2, respectively ($\chi^2 = 21.57$; df = 1; $p < 0.00001$; C = 0.197). The fluoroscopy times in group 1 ranged from 2 to 101 min (Me = 12 (IQR:7-20) exceeding those in group 2: 1 to 37 min (Me = 4 (IQR:2-7) ($p < 0.00001$). Effective doses in group 1 was 0.176-17.6 mSv (Me = 4 (IQR:2-7) which exceeded that in group 2: 0.06-4.95 mSv (Me = 0.325 (IQR:0.176-0.616) ($p < 0.00001$). The CA time in group 1 was also significantly longer compared with that in group 2: 1-35 min in group 1 (Me = 6 (IQR:3-16) versus 1-27 min in group 2 (Me = 4 (IQR: 2-7) ($p < 0.00001$).
Conclusions: An optimal protocol of the endocardial mapping for VA in children should include calculation of the probability of successful CA based on the comprehensive evaluation of EM results. Implementation of the approach to CA of the VA based on a comprehensive evaluation of the EM results and prediction of the probability of successful intervention may increase the efficacy of treatment for VA in children.

O8-4
Efficacy and Adverse Events Related to Initiation of "Second-Line Medications" for Supraventricular Arrhythmias in Children

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Introduction: Sotalol and Flecainide are used as second line agents for the treatment of supraventricular arrhythmias in children and are typically initiated in an inpatient setting. Efficacy and adverse events in this cohort have not been well described.

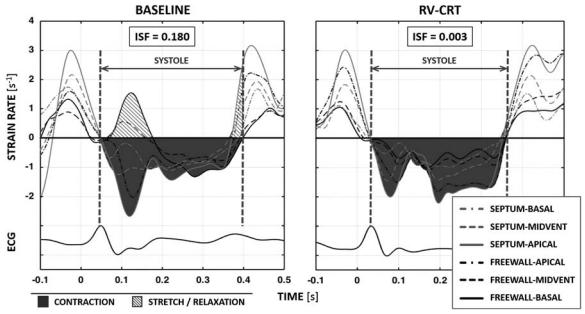
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Methods: Utilizing an institutional pharmacy database (2002-2014), all inpatients < 18 years of age initiated on sotalol or flecainide as second line treatment of supraventricular tachyarrhythmias were identified. Medical records, pharmacy registers, ECGs and Holters were retrospectively reviewed.
Results: In total, 335 pts were identified (Sotalol: n = 254, Flecainide: n = 81), with most (64%) being infants (<1 yr). Congenital heart disease (CHD) was present in 46% and 7% of pts initiated on sotalol and flecainide, respectively. The median starting dose of sotalol was 138 mg/m^2 (IQR:117-150). In total, arrhythmias were controlled with sotalol initiation in 210 (83%) of children, with an additional 6 (3%) controlled after addition of a betablocker. Sotalol dosing had to be decreased in 5 (2%) and stopped in 3 (1%) due to QT prolongation. Two pts had torsades de pointes and died on sotalol, both of whom had complex CHD. The median starting dose of flecainide was 100 mg/m^2 (84-119). Arrhythmias were controlled with flecainide initiation in 62 (77%), with an additional 5 (6%) controlled after addition of a betablocker. Flecainide dosing had to be decreased in 4 (5%) and stopped in 1 (1%) due to QRS prolongation. One pt with known cardiomyopathy had sustained VT and subsequently died on flecainide. In 11 pts, sotalol and flecainide were used in combination and successfully controlled the arrhythmia in all cases without proarrhythmia.There was no difference in efficacy between those with and without CHD (p = NS) or between infants and noninfants (p = NS) for either medication.
Conclusion: In children who fail therapy with first line agents for treatment of supraventricular arrhythmias, sotolal and flecainide are efficacious in the majority of cases. Although predominantly safe in otherwise healthy patients, ECG changes can occur, and in patients with CHD, proarrhythmic side effects can be seen. Inpatient monitoring should thus be considered during initiation, particularly in those with preexisting cardiac disease.

O8-5
Echocardiographic effects of pulmonary right ventricular resynchronization in congenital heart disease

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Introduction: Right ventricular (RV) electromechanical dyssynchrony may contribute to long-term pulmonary RV dysfunction in patients after surgery for congenital heart disease. We sought to evaluate immediate changes in RV mechanics after RV resynchronization (RVCRT).
Methods: 22 consecutive patients aged median 11.7 years with tetralogy of Fallot (N = 14), pulmonary atresia (N = 6), double outlet RV (N = 1) and arterial trunc (N = 1), resp., were echocardiographically studied following surgical RV revalvulation. RV-CRT was applied in the presence of complete right bundle branch block (RBBB) by atrial-triggered RV free wall pacing in complete fusion with spontaneous activation using temporary postoperative pacing wires.
Results: RV-CRT carried significant decrease in QRS duration (P < 0.001) along with elimination of the RBBB pattern, increase in RV filling time (P = 0.009), pulmonary artery velocity time integral (P = 0.015) and RV max.+ dP/dt as estimated from the tricuspid regurgitation jet (P = 0.011). RV mechanical synchrony improved: Septal to lateral RV mechanical delay decreased from median 65 to 28 ms (P < 0.001). RV internal stretch fraction (ISF) reflecting the ratio of myocardial stretching and contraction during systole diminished from mean (SD) 0.18 (0.09) to 0.09 (0.04) (P = 0.001, Figure). The q-RV interval at the pacing site (mean 77.1% of baseline QRS duration) confirmed pacing from a late-activated RV area.
Conclusions: RV-CRT carried multiple positive effects on RV mechanics including improvement in filling, systolic function and mechanical synchrony and efficiency.
(Supported by Ministry of Health of the Czech Republic, grant nr. 15-28029A)

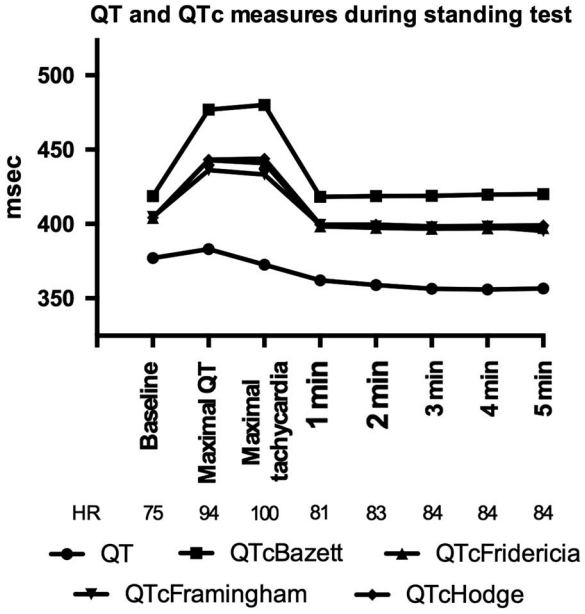


O8-6
QT interval dynamic changes from supine to standing in children

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Introduction: QT interval shortening in response to heart rate (HR) acceleration usually occurs within seconds, and may vary between healthy and disease conditions. QT hysteresis has been reported in children and adults performing exercise testing, and more recently in adults undergoing the standing test. A paradoxical QT interval increase has been used to confirm diagnosis of long QT syndrome in patients with borderline corrected QT (QTc). Studying QT hysteresis during a quick standing challenge may compensate for exercise testing; yet no pediatric data is available.
Methods: The standing test was performed in 51 healthy children (mean age 9.2 ± 2.8 yrs, 34 male) having a normal echocardiogram and negative family history for sudden death and long-QT syndrome. Following 10 min supine, fast standing with continuous ECG recording was performed. QT intervals were measured at baseline, maximal tachycardia, maximal QT and each minute of recovery (5 min), using a computerised software (leads II/V5), and corrected for HR using Bazzett (QTcB), Fridericia, Framingham, and Hodge formulas.
Results: In response to standing, HR increased from 75.2 ± 12.9 to 100.4 ± 12.7 beats/min. QT intervals at baseline and maximal tachycardia were similar (377 ± 33 vs 373 ± 37 msec, p = 1), however QTcB increased (419 ± 20 vs 480 ± 39 msec, p < 0.001). The 95th percentile for QTcB was 451 msec at baseline and 548 msec at maximal tachycardia. QT interval decreased throughout recovery (362 ± 28 msec at 1 min, 356 ± 24 msec at 5 min, p = 0.003). QTcB reached baseline values after 1 min of recovery and remained stable (418 ± 22 msec at 1 min, 420 ± 26 msec at 5 min, p = 0.97). Similar trends were recorded with the various correction formulas (Figure). Corrected QT using the Bazett formula was significantly higher at all point in times compared to the Fridericia, Framingham and Hodge formulas (p < 0.001).
Conclusions: This study provides pediatric reference values for normal QT and QTc interval responses to the brief tachycardia provoked by standing, which differ from published adult values. Potential applications of this easy bedside diagnostic test include, amongst other, screening for long QT syndrome.

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O9-1

Treatment of valvular aortic stenosis in children; a 20-year experience in a single institution

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Objective: The mode of treatment of valvular aortic stenosis, especially in the neonatal period, is still controversial. Valvotomy is the treatment of choice in our institution. We reviewed our experience regarding mortality, long-term outcome and need for re-intervention.
Methods: All patients <18 years who underwent relief of congenital valvular aortic stenosis with first intervention from January 1994 to December 2013 were identified in local catheter and surgical registries. Our referring area covers approximately 50% of the Swedish population concerning paediatric cardiac surgery. Medical charts of all patients were reviewed and followed until 18 years of age. Survival was cross-checked against the Swedish Population Registry as of Oct 1st 2015. No drop out occurred, except for three patients moving abroad. Patients with aortic regurgitation, subvalvular or supravalvular stenosis as main indication for surgery or other significant heart defects were excluded.
Results: 114 patients were identified (25 girls, 189 boys). Mean age at diagnosis was 2 days (0-13.3 years). Mean age at initial intervention was 2.9 months (0-17.9 years) with 43 < 1 month, 33 1-12 months and 38 patients >1 year. 93 had surgical valvotomy, 11 balloon dilatation, 4 Ross procedure, 5 prosthetic valve replacement, 2 Hegar dilatation as first procedure. In total 207 interventions were performed (surgical valvotomy 101, balloon dilatation 32, Ross procedure 17, mechanical prosthesis 18, biological prosthesis 7, Hegar dilatation 2, homograft 5, conversion to univentricular heart 3, heart transplant 2 and 20 interventions not related to the aortic valve). There was no 30-day mortality. One late death occurred at 10 months of age due heart-failure and concomitant pulmonary infection. Mean follow-up regarding mortality was 11.6 years (2-21.8 years). Re-intervention was required in 50 patients at mean 3.5 years (0-16.7, median 1.2 years). Freedom from re-intervention was 80, 69, 60, 57% at 1, 5, 10 and 15 years respectively.
Conclusion: Long-term survival was excellent. Many patients needed several interventions with valve replacement, including Ross surgery, in 33%. Our data do not allow comparison of catheter and surgical treatment, but based on these results we find no reason to change our current policy of surgery as first intervention.

O9-2

The Fontan circulation over time: attrition or natural decline of aging? A cross sectional study of functional status in Fontan patients

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Introduction: The nature of the impaired functional status of Fontan patients is far from unraveled, but is believed to be related to impaired cardiac function due to abnormal ventricular loading conditions, and to restricted pulmonary function. Furthermore, most studies reported the absolute value of peak oxygen uptake (VO2index) during exercise, whereas the peak VO2 as percentage of predicted could provide important information on how functional performance of the Fontan patients over time compares to healthy individuals. This study aims to investigate the functional status in a cohort of Fontan patients, and to identify its determinants, including cardiac characteristics, pulmonary characteristics and time since Fontan completion.
Methods and results: Eighty-five consecutive Fontan patients ≥10 years who performed adequate cardiopulmonary exercise testing (respiratory exchange ratio > 1.01) in 2012 or 2013, were included. Mean time since Fontan completion was 15 ± 9 years (range 2-37 years). New York Heart Association Functional Class (NYHA-FC) was I in 36 patients (42%), II in 41 patients (48%) and III in 8 patients (9%). Peak oxygen uptake during exercise (VO2index) was 25.7 ± 7.9 ml/min/m^2 (58 ± 14% of predicted). NYHA-FC and peak VO2index both correlated with time since the Fontan operation, but peak VO2 as percentage of predicted did not. In multivariate analyses, peak VO2 as percentage of predicted was independently associated with maximum heart rate, oxygen pulse at peak exercise and forced expiratory volume in 1 second (Rsquare = 0.579), but not with cardiac output in rest.
Conclusions: In Fontan patients, functional status is restricted compared to normal values, already early after completion of the Fontan circulation. The decrease in peak exercise capacity with longer time since the Fontan operation appears to be comparable to natural decline of aging. Additionally, functional status in Fontan patients appears to be related with pulmonary function and cardiac functional parameters during exercise, but not with conventional cardiac measurements at rest.

O9-3

Maternal and Fetal Outcomes in Cyanotic Congenital Heart Disease: a Multicentric Study of 55 Pregnancies

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Background: Many cyanotic congenital heart diseases (CHD) are deemed unsuitable for radical repair but are compatible

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with survival. Previous studies suggest that cyanotic CHD are of great maternal and fetal risk to pregnancy, but information on management of these pregnant women is lacking. The purpose of this study was to determine maternal and fetal outcomes in patients with cyanotic CHD.
Methods: This multicentric retrospective study included pregnant women with cyanotic CHD followed in 10 French specialized centers from 1992-2015. Patients with pulmonary hypertension were excluded. All pregnancies were reviewed. We observed maternal, obstetrical and neonatal outcomes.
Results: Twenty four patients (27 ± 6 years old) had 55 pregnancies. There were 10 miscarriages (19%), 40 complete pregnancies (≥20 week gestation (WG)), 4 abortions and 1 ectopic pregnancy. All pregnancies were singleton. Severe cardiac events occurred in 6 (11%) pregnancies. There was no maternal death. Cardiac complications were arrhythmia (n = 3), heart failure (n = 3) and stroke attack (n = 1). Six patients experienced deep hypoxia during the peri-partum period. No infectious endocarditis occurred. Half of cardiovascular events occurred in patients with single ventricle, and were more frequent in older patients (33 vs 27 y.o., p = 0.03). Obstetric complications occurred in 20%. Small for gestational age was diagnosed in 33%.The mean birth weight was 1868 ± 641 g at a mean gestational age of 34 ± 3WG, and 76% of newborns were premature. Neonatal death occurred in 4/40 live births. No CHD was diagnosed in the offspring.
Conclusion: Women with cyanotic CHD can go through pregnancy with a low risk to themselves during pregnancy and postpartum period. However, cyanotic CHD is associated with a high incidence of fetal and neonatal complications, with high rates of premature births, small gestational for age neonates and neonatal death. Close fetal monitoring and management in referral centers are required in this complex cardiac condition.

O9-4

Pregnancy outcomes in Eisenmenger syndrome in the modern management era

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Background: There is emerging evidence, that maternal mortality in pregnant women with PAH is lower in modern management era. This concerns patients with Eisenmenger syndrome (ES). In order to obtain further data, we retrospectively collected data of pregnancies in women with ES from 1997 to 2015 period.
Methods: This multicentric study included women with ES, followed in 7 French tertiary centers. All pregnancies were counted, including miscarriages and abortions. We observed maternal, obstetrical and neonatal outcomes.
Results: Twenty nine pregnancies in 18 women (25 ± 6 years old) with ES were managed during this period. There were 21 complete pregnancies (≥20 week gestation (WG)), 7 abortions, and 1 miscarriage. Six (32%) patients experienced severe cardiac events. The concerned females had lower saturation (79% vs. 89%) and were older. The most common cardiac complications during the complete pregnancies were heart failure (n = 4) and deep desaturation (n = 3), occurring mainly during the early postpartum period (n = 5/6) and after spinal anesthesia (3/6). Heart failure was particularly severe, needing inotropic treatment (n = 3) or ventricular assist device (n = 1), and leading to the only one death (mortality = 5%). Obstetric complications occurred in 38% of pregnancies. Small gestational for age was diagnosed in 33% (7/21) and was related to basal saturation level of the patient (p = 0.03), and to the maternal body mass index (p = 0.04). 12/21 (57%) pregnancies were delivered by cesarean section, with 7/12 for cardiac indications. The mean birth weight was 1824 ± 594 g at a mean gestational age of 34 ± 3WG. There was a high incidence of prematurity (57%), and no fetal or neonatal death.
Conclusions: Outcomes of pregnancy in women with ES are improved in the modern management era with a lower rate of maternal mortality. However, the severity of heart failure and the high rate of prematurity and SGA must still discourage ES patients to be pregnant.

O9-5

Clinical, neurohormonal and psychological characteristics predict on a long term basis, adverse cardiac events in patients with congenital heart defects

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Introduction: The growing cohort of adult patients with congenital heart disease (CHD) has an increased risk of mortality and morbidity, due to it's medical condition per se, but also due to psychological issues. Aim of this study is to identify the clinical, psychological and neurohormonal predictors of survival shared by this group of patients.
Methods: 60 consecutive clinical stable patients with various forms of CHD were recruited from a tertiary center. Patients' neurohormonal and psychological status, exercise capacity and cardiac function were assessed through plasma B-type brain natriuretic peptide (BNP) and interleukin 6 (IL-6) measurements, Beck depression inventory and Zung depression scale questionnaires, cardiopulmonary exercise test (CPET) and transthoracic echocardiography respectively. Patients were followed for major adverse cardiovascular events (MACE), including death or hospitalization for 5.1 ± 1.1 years.
Results: Most patients were symptomatic (48.3% with NYHA II and 36.7% with NYHA III). Mean plasma concentrations of BNP and IL-6 were 106.6 ± 98.6 pg/ml and 2.4 ± 2.6 pg/ml respectively. 17 patients (28.3%) were characterized as depressed. Patients with depression had higher plasma BNP levels (p = 0.030), limited exercise capacity, as expressed with peak VO_2 (p = 0.019) and higher probability of experiencing a MACE compared to non-depressed patients (95% CI: 1.630 to 3.616, p < 0.05). 22 patients (36.6%) experienced a MACE, among them 8 patients (13.3%) died. BNP, IL-6, peak VO_2, VE/VCO_2 were proved to be strong predictors of survival; BNP value > 241 pg/ml predicted MACE with a sensitivity of 65.38% and a specificity of 73.53% (AUC = 0.693, p < 0.0001), IL-6 value > 1.54 pg/ml predicted MACE with a sensitivity of 61.53% and a specificity of 73.53% (AUC = 0.627, p < 0.0001), VE/VCO_2 value > 38 predicted MACE with a sensitivity of 73.08% and a specificity of 76.47% (AUC = 0.808, p < 0.0001) and peak VO_2 value ≤ 21.4 ml/Kg/min predicted MACE with a sensitivity of 76.92% and a specificity of 70.59% (AUC = 0.794, p < 0.0001) respectively.
Conclusions: BNP, IL-6 levels, CPET parameters as well as depressive symptoms, predicted MACE and could be used as simple clinical markers for routine risk stratification and therapeutic manipulation of this population.

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O9-6

Influence of non-invasive hemodynamic CMR parameters on maximal exercise capacity in surgically untreated patients with Ebstein's anomaly

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Introduction: Ebstein's anomaly is often associated with RV dysfunction. Data on RV function in surgically untreated patients are however rare. Since a good quality of life correlates with a good exercise capacity in daily life, we investigated non-invasive quantitative data derived from cardiovascular magnetic resonance (CMR) and its impact on maximal exercise capacity in patients with surgically untreated Ebstein's anomaly.
Methods: We investigated 54 unoperated patients with Ebstein's anomaly, age 5 to 69 years (median 30 years) and examined these patients with CMR and cardiopulmonary exercise testing (CPET). We compared seventeen CMR parameters with CPET parameters. We performed univariate and multivariate analysis with the focus on the maximal exercise capacity in these patients. For the maximal exercise capacity peak oxygen uptake as the percentage of normal (peakVO_2%) was selected. The following CMR volume and flow parameters were correlated to peakVO_2%. Both right and left ventricular ejection fraction (RVEF and LVEF), the indexed end-diastolic and endsystolic volumes (RVEDVi, RVESVi, LVEDVi and LVESVi) as well as the indexed stroke volumes (RVSVi and LVSVi), the total normalized right and left heart volumes (volume of the atrium and the ventricle together) as well as the total right to left heart volume ratio (R/L-ratio). Indexed flow data as the antegrade (PA ante, Aorta ante) and the indexed net flow (PA netto, Aorta netto) in the pulmonary artery and the aorta as well as its normalized values on heart rate (CI-PA, CI-Aorta) were used.
Results: RVEF (r^2 0.2788), PA netto (r^2 0.2330), and PA ante (r^2 0.1912) showed the best correlation with peakVO_2% (all $p < 0.001$). Further significant linear correlation could also be demonstrated with CI-PA, LVEF, LVSVi, Aorta netto, RVESVi and Aorta ante. All other parameters did not show a significant correlation with peakVO_2%. Multivariate analysis for RVEF and PA netto revealed a r^2 of 0.4350.
Conclusions: CMR parameters reflecting cardiac function as RVEF and LVEF and flow data of cardiac forward flow best correlate to peakVO_2%. The evaluation of the indexed net flow in the pulmonary artery and the overall function of the right ventricle (RVEF) best predicts the maximal exercise capacity in patients with Ebstein's anomaly.

O10-1

Catecholaminergic polymorphic ventricular tachycardia, type 3: An autosomal recessive inherited cardiac arrhythmia caused by novel mutation in the TECRL gene

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Introduction: Primary cardiac arrhythmias could be one of the important causes of sudden cardiac deaths (SCD) in children and adolescents. Mutations in cardiac channel and their ancillary protein encoding genes have frequently been described in primary cardiac arrhythmias. Of them, KCNQ1, KCNH2, SCN5A, RYR2, CASQ2, CALM1-2 are the important ones. The aim is to present a new type of Catecholaminergic Polymorphic Ventricular Tachycardia (CPVT) and elucidate its genetic etiology.
Methods: Two families described in this study originated from Sudan. The parents in both families are first-degree cousins. Seven of their 13 children presented with exertion-induced arrhythmias or SCD. Five children died following the arrhythmic event. In the surviving two children, an Implantable Converter Defibrillator (ICD) was implanted in one child while the other suffered severe brain damage. Whole exome sequencing was performed to explore the genetic defect. Patient-specific stem cell induced-cardiomyocytes (hiPSC-CM) were made to evaluate the functional phenotype.
Results: ECG showed polymorphic ventricular fibrillation and torsade de pointes. ECG at rest showed borderline prolonged QTc interval of 450 ms. ICD interrogation of one surviving child revealed an episode of ventricular tachycardia. All affected children were homozygous for a splice donor site mutation, c.331+1 G > A in the TECRL (also annotated SRD5A2L2) gene on chromosome 4. iPSC derived cardiomyocytes demonstrated elevated diastolic Ca2+ concentration, action potential prolongation and adrenergic induced triggered activity. Antiarrhythmic medication flecainide significantly reduced the triggered activity.
Conclusion: CPVT type 3 is a novel malignant form of cardiac arrhythmia, caused by homozygous mutation in the TECRL gene. These findings have implications for diagnosis and treatment of inherited cardiac arrhythmias.

O10-2

Oral Triidothyronine Supplementation Increases Lactate-pyruvate Ratio As a Marker of Effective Availability of Energy Substrates

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Background: The incidence of euthyroid sick syndrome (ESS) was thought to be closely related to low cardiac output syndrome (LCOS) after congenital heart disease (CHD) surgery. Thyroid hormones exert their action on cardiovascular function by improving lactate and pyruvate utilization as effective energy substrates for mitochondrial respiratory function.
Objectives: To provide evidence that oral T3 can prevent and ameliorate ESS, thus subsequently reduce the incidence of LCOS, decrease serum lactate levels and lactate-pyruvate ratio as a marker of effective utilization of energy substrates.
Methods: The study was a single center, randomized, double blind, and controlled clinical trial. It was conducted in children, ≤3 years of age undergoing corrective open heart surgery. The treatment

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group received oral T3 supplementation 1 μg/kgBW while the placebo group received saccharum lactis every 6 hours from the induction of anaesthesia until 60 hours after the first dose.
Results: A total of 171 participants were enrolled to the study. ESS was already found in 22.2% before surgery and in 89% at 24 hours post cross clamp removal (FT3 less than 2.5 pg/mL). LCOS was higher in the placebo than the treatment group with a significant difference at 6 hours post cross clamp removal (37% vs. 20% respectively, odds ratio of 2.28 (1.15-4.52), $p = 0.02$. At 1 hour after cross clamp removal the serum lactate levels were significantly lower in the treatment group compared to placebo (1.50 (1.27–1.83) and 1.70 (1.55–2.23) mmol/L, respectively, $p = 0.04$). There was an increase of lactate-pyruvate ratio at 1 hour post cross clamp removal in the treatment group without LCOS compared to those with LCOS [20.99 (15.82–31.56) vs. 12.61(8.89–18.67), respectively, $p = 0.04$] and in the placebo group at 24 hours post cross clamp removal [35.22 (22.08–65.75) vs.17.05 (11.84–26.07)], respectively, $p = 0.01$).
Conclusion: Oral T3 prophylaxis could prevent and ameliorate ESS, which subsequently reduced LCOS after cardiac surgery. An increased lactate-pyruvate ratio in the treatment group may be a sign of an early increased pyruvate utilization for energy substrate.

O10-3
Influence of intrauterine growth restriction on postnatal osteoprotegerin concentrations and aortic intima-media thickness

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Introduction: Intrauterine growth restriction (IUGR) is considered a risk factor for atherosclerosis and coronary artery disease in adulthood. Understanding the pathophysiology of cardiovascular disease involves the identification of novel risk factors and biomarkers. There is emerging evidence that osteoprotegerin (OPG), a member of the tumor necrosis factor-receptor superfamily, participates in the pathogenesis of atherosclerosis by amplifying the effects of inflammation and traditional risk factors. In this case-control study we investigated whether IUGR affects postnatal OPG concentrations and the possible association between OPG levels and aortic intima-media thickness (aIMT), an index of preclinical atherosclerosis.
Methods: We studied 30 IUGR and 30 appropriate for gestational age (AGA) neonates matched for gestational age and gender. Quantitative determination of plasma OPG was performed with enzyme immunoassay on the second (DOL2) and fifth day of life (DOL5). Aortic intima-media thickness (aIMT) was measured in the distal abdominal aorta using a linear array probe, and adjusted for aortic lumen diameter.
Results: Neonates with IUGR had significantly higher OPG levels on both DOL2 and DOL5 as compared to controls (DOL2: 5.4 ± 1.0 mmol/L vs. 4.6 ± 1.0 mmol/L, $p = 0.002$ and DOL5: 5.1 ± 0.8 mmol/L vs. 3.9 ± 0.7 mmol/L, $p < 0.001$). Between DOL2 and DOL5, OPG concentrations did not change significantly in neonates with IUGR ($p = 0.087$), but decreased slightly in controls ($p = 0.003$). IUGR was also associated with increased aIMT (0.11 ± 0.03 vs. 0.06 ± 0.02, $p < 0.001$). There was a positive correlation between OPG and aIMT on DOL2 ($r = 0.494$, $p < 0.001$), which became stronger on DOL5 ($r = 0.791$, $p < 0.001$; Figure).
Conclusion: We report significantly increased concentrations of OPG in IUGR neonates and a positive correlation with aIMT. The dynamics of OPG concentrations during the transitional period support the fetal origin of the cytokine. Follow-up studies with repeat OPG and aIMT measurements may be indicated to evaluate whether these findings represent a permanent effect of IUGR on the offspring.

O10-4
Genotype-Phenotype correlations in pediatric SCN5A mutation carriers: new insights for a better risk stratification

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Introduction: Genotype-phenotype correlations of SCN5A mutations remain unclear. Given the relative rarity of cardiac sodium channelopathies in the pediatric population, risk stratification in the young diagnosed with a given SCN5A mutation need to be clarified.
Methods: A multicenter, international, 1990-2015 retrospective cohort study was conducted in 25 tertiary hospitals in 13 different countries. All patients 16 years of age or younger diagnosed with a genetically confirmed SCN5A mutation, whatever the clinical diagnosis were included in the analysis.
Results: 423 children fulfilled the study inclusion criteria, with a median age of 7.6 (0.0-16.7) years at diagnosis; 34.7% individuals were probands. Phenotypic spectrum was divided in 76 (18.0%) isolated LQT3, 33 (7.8%) isolated BrS type 1, 86 (20.3%) isolated PCCD, 3 (0.7%) isolated SSS and 102 (24.1%) overlap phenotypes; 123 (29.1%) kept a negative phenotype throughout follow-up. The risk of arrhythmic events in children was high, especially when a spontaneous BrS, LQTS, PCCD or overlap phenotype was displayed but also in those with a negative phenotype. Phenotype varied according to mutation type, missense pathogenic mutations being more frequent than radical mutations or variants of unknown significance in isolated LQT3, isolated PCCD and negative phenotype patients. Cardiac arrest or syncope as first symptom as well as appropriate ICD shocks in implanted patients were more frequently observed in case of mutation located to the transmembrane region. Compound genotype, double SCN5A mutation, sinus node dysfunction, age $\leq$1 year at diagnosis and absence of family history of BrS, LQTS, PCCD or PM implantation and cardiac arrest or ICD implantation were independent predictors of cardiac event giving new insights to identify high-risk subgroups in SCN5A mutation-positive infants and children.
Conclusion: In the largest series of SCN5A mutation carriers children, we found a high rate of cardiac events; ECG phenotype varied according to mutation type, whereas clinical severity was related to mutation location; several factors emerged as predictors of cardiac arrest or arrhythmic event.

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O10-5

NOS1AP variants affecting QTc in the long QT syndrome- mainly a male affair?

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Introduction: Single nucleotide polymorphisms (SNPs) in the NOS1AP gene have repeatedly been reported to influence QTc, albeit with moderate effect sizes. In the long QT syndrome (LQTS) this may contribute to the substantial variance seen in QTc among carriers of identical pathogenic sequence variants. Here we assess three previously reported NOS1AP SNPs for association with QTc in two large Swedish LQT1 founder populations.
Methods: This study included 312 individuals (180 females) from two LQT1 founder populations, whereof 227 genotype positive (133 females) segregating either Y111C (n = 148, 84 females) or R518* (n = 79, 49 females) pathogenic sequence variants in the KCNQ1 gene, and 85 genotype negative (47 females). All were genotyped for NOS1AP SNPs rs12143842, rs16847548 and rs4657139, and tested for association with QTc length (effect size presented as mean difference between derived and wildtype, in ms). Mean QTc was obtained by repeated manual measurement (preferably in lead II) by one observer using coded 50 mm/s standard 12-lead ECGs.
Results: A substantial variance in mean QTc was seen; all genotype positive 475 ± 33 ms (Y111C 482 ± 30 ms; R518* 462 ± 34 ms) and genotype negative 433 ± 24 ms. Female sex was significantly associated with QTc prolongation in all groups ($p < 0.01$) with effect sizes ranging from 14 ms (genotype negatives) to 16 ms (Y111C), 20 ms (genotype positives) and 30 ms (R518*). Two derived NOS1AP SNPs (rs12143842 and rs16847548) were significantly associated with QTc prolongation in genotype positives (10 ms, $p = 0.02$), in genotype negatives similar results were seen (9 ms, $p = 0.07$). Notably, among genotype positives, when stratified by sex neither of these two SNPs were significantly associated with QTc in females (all 6 ms, $p = 0.2$; Y111C 9 ms, $p = 0.1$; R518* 1 ms, $p = 0.9$) while in males, a prolongation of 16 ms, $p = 0.03$ was seen (Y111C 15 ms, $p = 0.07$; R518* 27 ms, $p = 0.007$). In genotype negatives, while non-significant, the same SNPs were associated with a 5 ms (females) and 10 ms (males) QTc prolongation.
Conclusions: Our findings suggest specific genotype and most importantly sex affects the effect size of NOS1AP SNPs on QTc. This may be of clinical significance when applying NOS1AP genotype to clinical risk stratification.

O10-6

Single nucleotide polymorphisms in KCNQ1-3'UTR associated with allele-specific repression of transcription segregate with the phenotypically benign Swedish Long QT Syndrome type 1 founder mutations

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Introduction: Strong allele-specific effects, worsening and/or possibly ameliorating Long QT Syndrome type 1 (LQT1) phenotypes depending on location, have been proposed for single nucleotide polymorphisms (SNPs) in KCNQ1-3'UTR. Here we assess three previously reported KCNQ1-3'UTR SNPs for allele-specific location and association with QTc in two large Swedish LQTS founder populations with a previously documented low incidence of sudden cardiac death.
Methods: This study included 312 individuals from two LQT1 founder populations, whereof 85 genotype negative family members and 227 genotype positive individuals segregating either the dominant-negative variant Y111C (n = 148) or the haploinsufficiency-causing variant R518* (n = 79) in the KCNQ1 gene. All were genotyped for KCNQ1-3'UTR SNPs rs2519184, rs8234 and rs10798. Allelic phase (cis/trans) was determined by trio analysis. Association between mean QTc, obtained by repeated manual measurement on coded 50 mm/s standard 12-lead ECGs, and allele-specific location of KCNQ1-3'UTR SNPs was tested within each founder population among carriers of identical mutations, testing between populations was not performed.
Results: Among all genotype positives, 92% had at least two KCNQ1-3'UTR SNPs in cis (rs8234 and rs10798 were in complete linkage disequilibrium) and 97% of R518* carriers had all three reported repressive SNPs in cis. Mean QTc was 482 ± 30 ms and 462 ± 34 ms among carriers of Y111C and R518*, respectively. No significant associations between QTc and allele-specific SNP location were found within the founder populations. Among the 16/148 Y111C carriers that lacked SNPs in cis, mean QTc was 490 ± 36 ms as compared to 481 ± 29 ms in carriers with SNPs in cis, and mean QTc was 4-10 ms longer in R518* carriers that in addition to 3 SNPs in cis also had 1-3 SNPs in trans (none of these findings were statistically significant).
Conclusions: Three KCNQ1-3'UTR SNPs, previously reported to repress transcription of the allele where they reside, were found to segregate with the Swedish LQT1 founder mutations (2/3 in cis with Y111C and 3/3 with R518X). This finding could relate to the surprisingly mild phenotypes previously described for both these populations, albeit no significant association between QTc and allele-specific location of SNPs could be seen in these relatively homogenous samples.

O11-1

Termination of Pregnancy Epidemiology following Fetal Congenital Heart Disease Diagnosis

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Objectives: To document the probability of Termination of Pregnancy (TOP) following fetal Congenital Heart Disease (fCHD) diagnosis in a Mediterranean population.
Methods: Retrospective study of medical reports and fetal echocardiogram findings performed over 6 years (1997-2013) in the only referral center for fetal cardiology available on an island of 600.000 inhabitants. Critical fCHD were considered cases with probable ductal dependent postnatal circulation. Caryotype findings and pregnancy outcome regarding TOP were obtained from families or treating physicians. Odds Ratios (O.R) and 95%C.I. for TOP following diagnosis of any fCHD, critical fCHD, presence of caryotype abnormalities have been estimated, using Pearson Chi Square analysis.
Results: 1808 out of 1847 fetuses (1804 pregnancies), with complete medical information, evaluated <24th GW, were included. The overall incidence of TOP was 1.8% (n = 32), of fCHD 27% (n = 499), of critical fCHD 1.9% (n = 34). Caryotyping was available in 10% of cases (n = 185), with 13% of them (n = 25) being abnormal.
The probability of TOP was 5.7% following diagnosis of any fCHD vs 0.2% in the absence of fCHD, corresponding to O.R: 26.2 (7.9-86.8), $p < 0.001$ for abortion following fCHD diagnosis. The diagnosis of critical fCHD was associated with an O.R: 185

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(76-449), $p < 0.001$ for TOP, as 19/13 cases (56%) with critical fCHD (with available outcome data) vs 12/1771 (0.7%) of non-critical or absence of fCHD opted for TOP.
CHD specific TOP rates were documented: VSD (2/239), AVSD (3/8), HLHS (3/5), P.Atresia (5/5), Tr.Atresia (1/2), DORV (2/4), TOF (3/9), Truncus Art (1/2), TAPVC (1/1), CoA (1/12), DILV (1/2),
The presence of caryotype abnormalities increased in a lesser extend the TOP probability (O.R: 3, 0.9-9.6) overall. However the incidence of known caryotype abnormalities in fCHD and critical fCHD groups did not differ significantly between subgroups deciding for TOP compared to those with pregnancy continuation.
Conclusions: Detection of fCHD and critical fCHD was associated 6% and 56% chance for TOP, respectively. fCHD type and severity influenced TOP probability, independently of fetal caryotype findings. As the epidemiology of liveborn CHD is affected by antenatal CHD diagnosis, further study is needed to document factors that influence family decisions while support services should be established.

O11-2
Quality of Life in Patients After Staged Operation For Hypoplastic Left Heart Syndrome (HLHS) and Their Families - One Center Experience

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Background: Improvement in results of staged palliation for HLHS necessitates assessment of quality of life in this group of patients and their families.
AIM: To assess the quality of life of children with HLHS and their families.
Methods: Mothers of 98 children with HLHS (73 boys – 75%) completed the questionnaire to assess quality of their children's life and impact of child's illness onto the family. All children (age 3-17 years) were operated at our institution (43 were after Glenn anastomosis and 55 after Fontan completion at the time of study). In 26% child with HLHS was the only child in family.
Results: Development estimated as normal was reported in 94% of patients with HLHS. 84% of patients attend to normal schools or kindergartens. Physical activity comparable with peers was reported in only 19%. Limited physical activity and exercise intolerance were reported in 81%, emotional problems in 25%, educational in 9%. Good tolerance of frequent hospitalizations was reported in 75% of cases.
Child illness was connected with strong parental stress (93%), but 91% of responders estimated that they successfully cope with the problem. Family support, support groups of parents and religious faith were considered as most helpful. Only 12% of mothers looked for professional psychological care. 94% responders assessed familial atmosphere as good, in 85% child's illness strengthened parental marriage, but in18% of families mother was the only parent. Impact of child's illness on family material situation was assessed as significantly negative in 79%. In 50% of families the father is the only working parent. 98% of responders would not change anything in their choice of the method and place of treatment.
Conclusions:

1. Patients with HLHS are active members of the society, they attend to normal schools and kindergartens although their physical activity is limited.
2. The family functioning is good but child's illness is a reason of strong parental stress and indicates material problems.
3. Increasing number of HLHS survivors indicates the need for continuation studies concerning neurodevelopmental outcome, quality of life, career planning and social functioning.

O11-3
Asymmetric septal hypertrophy in children on long-term parenteral nutrition – the glucose role?

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Introduction: Long-term parenteral nutrition(PN) is an effective method of treatment in children with intestinal failure, however it is related to several complications. Hypothetically there are several factors related to PN which might influence cardiac status, such as volume overload and metabolic influence of administered nutrition mixtures.
Objectives: The aim of this cross-sectional study was to evaluate the cardiac status in children with intestinal failure on long-term PN and to identify factors that might influence it.
Methods: 71 children with intestinal failure aged from 0.92 to 19.84 years (average – 7.6 year), being on parenteral nutrition for average of 4.9 years were examined by echocardiography with assessment of basic functional and structure parameters, chest X-ray with assessment of CTR and NTproBNP serum concentration. Results were compared with normal values established on the base of epidemiological studies and analyzed by T-student test. Correlation of the cardiac results with potential risk factors related to PN such as: nutrients and fluid content, fluid flow, frequency of PN administration per week, the period from initiating PN were analyzed with Spearman correlation test.
Results: The main stated aberrations were: the augmentation of the echocardiographic IVSd diameter and IVSd/LVPWd ratio above normal limits (>2 SD and >1,3) in19% patients (average of the abnormal IVSd diameter:140% of the mean normal value (130%-170%); average percentile in the studied group – 70th; $p < 0.001$), and elevation of CTR in 44% and of NTproBNP serum level in 32% .
Those aberrations correlated with glucose content in the PN mixture (CTR: $p = 0.008$; $r = 0.329$; NTproBNP: $p < 0.001$; $r = 0.491$; IVSd: $p = 0.024$; $r = 0.273$), with PN's frequency (CTR: $p = 0.032$; $r = 0.276$; NTproBNP: $p = 0.013$; $r = 0.295$) and with PN mixture's volume (CTR: $p < 0.001$; $r = 0.395$; NTproBNP: $p < 0.001$; $r = 0.327$; IVSd: $p = 0.014$; $r = 0.306$).
Conclusions: The main aberration of the cardiac status in children on long-term PN was the enlargment of IVSd diameter, which correlated significantly with glucose content in PN (resembling the situation of ASH in infants of diabetic mothers) and elevation of IVSd/LVPWd ratio. There was also frequently stated elevation of CTR on chest X-ray and of NTproBNP serum concentration, which both correlated significantly with PN volume.

O11-4
Family history of sudden death is not an independent risk factor for sudden death in childhood hypertrophic cardiomyopathy - risk is determined by ECG- and echocardiography predictors

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Background: Reports from tertiary centres in adult cardiology specializing in hypertrophic cardiomyopathy (HCM) have indicated family history of sudden death (SD) as a significant risk factor, and it is classed as one of three strong risk factors meriting implantation of an internal cardiac defibrillator (ICD) in the latest American Heart Association guidelines. However, several studies from paediatric cardiology centres with few events have failed to confirm this association in paediatric HCM patients. We have studied this question in a national cohort.
Patients and Methods: Patients with a diagnosis of HCM before age 19 years attending all five regional centres of paediatric cardiology in Sweden have been studied. There were 27 patients with sudden death or re-suscitated cardiac arrest, and 103 patients with at least two years follow-up and without heart-failure death (mean follow-up 9.2 years). These were studied with Cox proportional hazard regression in respect to previously suggested possible risk factors for sudden death.
Results: On univariate Cox-hazard analysis first and latest ECG risk score (both $p < 0.001$) (see Eur Heart J 2010;31:439-449), both first and latest septal thickness in percent of 95th centile (SEPPER) (both $p < 0.001$), VT on Holter ($P = 0.003$), left ventricular outflow tract obstruction at rest ($p = 0.001$), and latest left atrium: aortic ratio ($p = 0.002$) were significant risk factors whereas family history of SD ($p = 0.89$), and maximal wall thickness in mm ($p = 0.80$) were not. Beta-blocker therapy ($p = 0.002$) was the only therapy to significantly reduce risk, also evident as dose-related protection (propranolol-equivalent/kg $p = 0.012$). On multivariate Cox-hazard last ECG risk score ($p < 0.001$), last SEPPER ($p = 0.001$), gender ($p = 0.054$) and beta-blocker therapy ($p < 0.001$, protective) were significant. Out of 34 patients with family history of SD 7 died suddenly, and 27 have survived (median 8 years). Those who died were identifiable as high-risk phenotypes with initial SEPPER median 289% [IQR 194-435], and initial and last ECG risk scores 7 [4-9] and 8 [7-11], versus survivors: first SEPPER 134% [105-192] $p = 0.0022$, first risk score 2 [1-4] $p = 0.017$, and last ECG risk score 1 [0-5] $p = 0.0026$.
Conclusion: Family history of SD is not an independent risk factor in childhood HCM; the phenotype determines risk of sudden death.

O11-5
BMI in patients with CHD compared with the general population

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Objectives: Children with congenital heart disease (CHD) often have unique metabolic challenges such as exercise intolerance or failure to thrive, as well as potential recommendations of activity restriction. The purpose of this study was to examine the prevalence of obesity and underweight in the CHD population as stratified by disease severity with comparisons to the general population.
Methods: Since 1991 the Children's Database has recorded height, weight and body mass index (BMI) as measured by medical doctors and nurses at annual school-based preventive health checks offered to all Danish children. Using the Danish National Patient Registry we identified all individuals born and diagnosed with CHD in Denmark during 1991-2012. A unique personal identifier enabled identification of CHD subjects with at least one BMI measurement recorded in the Children's Database. BMI measurements were categorized in one-year increments (6-12). For each year increment, we identified a general population sample, matched (1:10) on age and gender. Obesity was defined as BMI above the 95th percentile and underweight as BMI less than the 5th percentile for age and gender. We determined the prevalence of obesity and underweight and used conditional logistic regression to compute the corresponding odds ratios (OR). CHD was stratified by severity (mild, moderate, and complex).
Results: We identified 2,394 CHD patients with at least one BMI measurement recorded. The prevalence of obesity in the CHD population was 5.3%, and the prevalence for underweight was 9.3%. The corresponding ORs were 1.1 (95% CI:0.9–1.3) and 1.9 (95% CI:1.7–2.2), respectively. The ORs for obesity were 1.2 (95% CI:0.9–1.5) for mild, 0.7 (95% CI:0.4–1.4) for moderate, and 1.1 (95% CI:0.8–1.6) for comp1ex CHD. For underweight the ORs were 1.6 (95% CI:1.3–2.1) for mild, 2.6 (95% CI:1.8–3.8) for moderate, and 2.6 (95% CI:2.0–3.4) for comp1ex CHD.
Conclusion: We found no evidence to suggest an increased prevalence of obesity in children with CHD compared with the general population. The CHD population had an increased prevalence of underweight, in particular individuals with moderate and complex CHD.

O11-6
Exercise, Lifestyle and Psychological Status of a Large Cohort of Children with Congenital Heart Disease

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Introduction: Improved survival among children with congenital heart disease (CHD) has shifted focus to the long-term physical and psychological outcomes for these patients. There is evidence that children with CHD have lower levels of daily physical activity and a higher prevalence of obesity compared to their normal peers. The benefits of an active lifestyle within the general population have been well described. They include better cardiovascular health, improved psychological, cognitive and social functioning and obesity prevention. This study aims to assess both physical and psychological functioning in children with CHD.
Methods: 430 patients aged between 5-10 years old with CHD were identified on Heartsuite Database and invited to participate. Each patient recruited underwent assessment as detailed below:
Biophysical assessments:

- Weight, height, waist measurements
- Baseline heart rate, blood pressure, oxygen saturation
- Exercise stress test (EST) – Graded cycle ergometer protocol
- Actigraph accelerometer worn at home for 1 week

Psychosocial assessments:

- Kidscreen27: To assess health related quality of life (HRQoL)
- Strengths and Difficulties Questionnaire: To assess for behavioral problems
- Butler Self-image Profile was completed by children over 7 years old

Results:

- 163 patients were recruited and underwent assessment, 100 were male (61.3%).

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- The mean age was 8.4 years (range 5.3–11.5).
- 136 of the patients had major CHD* (as defined by requiring catheter or surgical intervention).
- Patient subgroups: 18.4% acyanotic no intervention, 37.4% acyanotic repaired, 27.6% cyanotic corrected, 16.6% cyanotic palliated.
- EST: EST duration mean 5.89 mins (SD 2.02), METs mean 9.79 (SD 1.79), mean Maximal predicted HR 81% (SD 7.8)
  - The 'cyanotic palliated' subgroup had significantly lower EST duration, maximal HR and oxygen saturations at peak exercise compared with the other 3 subgroups.
- Actigraph: Average time spent in MVPA (Moderate-Vigorous Physical Activity) 45 mins (SD -27.2), percentage time spent in MVPA 6.3% (SD 3.7).
- No significant difference was found in Kidscreen27 HRQoL measures between minor and major CHD groups.

(*3 patients had not undergone intervention by the time of the baseline assessment but were scheduled for surgical repair were coded as 'major CHD').

Conclusions: Overall physical and psychological wellbeing is well preserved in the majority of children aged 5-11 years with CHD.

O12-1
Prenatal diagnosis of single ventricle physiology impacts morbidity and mortality

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Introduction (or Basis or Objectives): *Objectives:* Single ventricle (SV) lesions are most often detected in fetal echocardiography. We sought to evaluate the impact of prenatal diagnosis on morbidity and mortality.

Methods: All consecutive patients born between 2001 and 2013 with pre- or postnatal diagnosed SV and birth weight >1800 g from 1/2001 until 6/2013 were reviewed. Primary endpoint was 30days survival rate after Hemifontan palliation. Secondary endpoints were condition at admission, neonatal mortality (30d) and hospital morbidity after the first operation.

Results: 259 cases with SV physiology 160(62%) were prenatally diagnosed. After termination of pregnancy, intrauterine demise and comfort care a total of 181 alive newborns were admitted to our center for treatment. One patient died for non-cardiac cause. Thus 87 fetal cases and 93 postnatal cases were analysed. Prenatal and postnatal anatomical diagnoses showed similar distribution, including hypoplastic left heart syndrome 40/35%, atrioventricular septal defect 10/11%, tricuspid atresia 15/10%, double inlet left ventricle 14/13%, double outlet right ventricle 9/13%, others 12/18% and right ventricular predominance 56/55%. High-risk patients were equally present in both groups, and consisted of restrictive foramen or obstructive pulmonary veins 8/8%, right atrial isomerism3/2%, left atrial isomerism4/2%). Patients with a prenatal diagnosis were born earlier (38.2 ± 1.4 versus 39.3 ± 1.5, $p < 0.0001$), but birth weight was not significantly different (3033 ± 453 g versus 3167 ± 613,g $p = 0.09$). Lactate at admission was more often >10 mmol/l in postnatal cases (9/93 versus 1/87; $p = 0.02$). PH at admission was more often <7.20 in postnatal cases than in prenatal diagnosed cases (10/93 versus 1/87; $p = 0.001$). Postnatal diagnosed children presented ad admission with higher dose of prostaglandin ≥0.05 mcg/kg/min (14/93 versus 2/87, $p = 0.003$) and required more often mechanical ventilation (25/93 versus 2/87; $p < 0.0001$). Neonatal mortality was significantly higher in postnatal diagnosed children (14% versus 4.6%, $p = 0.03$). Overall mortality until 30d after Hemifontan palliation, including transplantation in one prenatal case, was also higher in postnatal diagnosed patients (24.7% versus 12.6%; $p = 0.04$).

Conclusions: Prenatal diagnosis helps reducing neonatal morbidity and mortality in children with single ventricle physiology. Overall mortality remains significantly lower until 30 days after Hemifontan palliation in prenatal diagnosed compared to postnatal diagnosed cases.

O12-2
Ventricular Doppler Inflow Duration Corrected by Cycle Length in Fetuses with Normal and Abnormal Diastolic Function

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Introduction: Myocardial performance index (MPI) and cardiovascular profile score (CVPS) quantify fetal cardiac dysfunction and heart failure. CVPS combines 5 echocardiographic variables (effusions; heart size; cardiac function; systemic venous flow; umbilical arterial flow) with deduction of 1-2 points from maximal 10 points per abnormal component. MPI, calculated as isovolumetric contraction plus relaxation times/ejection time, requires antegrade pulmonary/aortic flow. In fetuses with normal and abnormal cardiac function, we sought to determine correlation of another measure of function, ventricular inflow duration corrected by cardiac cycle length (ID) with MPI and CVPS.

Methods: Fetuses diagnosed with cardiomyopathy (CM; n = 30) and twin-twin transfusion syndrome (TTTS; n = 30) were compared with age-matched healthy fetuses (n = 120). All measurements were made offline. Observer agreement was evaluated in 30 cases.

Results: The table summarizes the main findings. For CM/TTTS, significant linear correlations were found among all variables, with best fit between IDs and MPIs (R2 0.5-0.56; $p < 0.001$). Using the 95th confidence limit as normal cutoff, ID < 35% and MPI > 0.43 were considered abnormal. In 86% of ID and MPI measurements, both results were either normal or abnormal. Intra- and inter-observer agreements were comparable for IDs (0.96; 0.89) and MPIs (0.95; 0.88).

Conclusions: ID is a valuable and reproducible index to detect and quantify abnormal fetal ventricular function and should be included in baseline echocardiographic assessment.

	TTTS	CM	Normal	p-values
Cases	30	30	120	
Age	21.8 ± 4.2%	24.9 ± 5.7%	23.8 ± 5.3%	NS
Left VID	31.9 ± 7%	31.8 ± 7.7%	41.1 ± 2.7%	<0.001
Right VID	26.7 ± 8.3%	32 ± 6.4%	40.7 ± 2.3%	<0.001
Left MPI	0.73 ± 0.23	0.83 ± 0.43	0.42 ± 0.05	<0.001
Right MPI	0.98 ± 0.94	0.85 ± 0.49	0.40 ± 0.06	<0.001
CVPS	6.9 ± 2.5	5.4 ± 2.2	10 ± 0.0	<0.001

O12-3
Serial Echocardiography to Prevent Major Immune-Mediated Heart Disease in the Fetus: Results of a Risk-Based Prospective Surveillance Strategy

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Background: Exposure of the fetus to maternal autoantibodies (AB) is the main etiology of congenital complete heart block (CHB) and endocardial fibroelastosis (EFE). Because of the perception that these immune-mediated conditions may be preventable if detected and treated at an early disease stage, weekly fetal echocardiographic surveillance during the period of highest fetal risk of complications has been advocated to all mothers with positive anti-Ro/La AB tests. In an earlier study, we found that all mothers of 40 children with immune-mediated CHB had high anti-Ro titers and concluded that serial assessments could be safely limited to women with these high levels. This prospective study examines the utility of this approach.
Methods: Included were referrals ≤22 gestational weeks for a positive maternal AB test from 2009-2014. At the baseline fetal echocardiogram, maternal anti-Ro sera titers were measured by ELISA and results graded as equivocal (group 1: anti-Ro < 8 U/ml), low-positive (group 2: 8-49 U/ml), or high-positive (group 3: ≥ P U/ml). Weekly echocardiograms to 24 weeks (no previous child with CHB or EFE) or 28 weeks (previously affected offspring) were recommended to group 3 mothers while a neonatal exam including an electrocardiogram was offered to all referrals. Chi-square, Fisher exact tests and the Student *t* test were used for intergroup comparisons.
Results: Of 232 mothers with 241 fetuses, 43 tested anti-Ro equivocal, 62 low-positive, and 127 high-positive. Numbers of fetal exams per patient were significantly less in group 1 and 2 (2; range: 1-7) when compared with group 3 (4; 1-21; p < 0.001). Immune-mediated heart disease, including CHB (n = 4), incomplete heart block (n = 4) and isolated EFE (n = 1), was diagnosed in 9 (8%) group 3 fetuses with Ro-titers >100 U/ml and none of the fetuses exposed to lower Ro-titers (odds ratio: 26.4; p < 0.001). Unlike CHB, incomplete block and EFE regressed with perinatal steroid and immunoglobulin therapy.
Conclusions: Restricting fetal echocardiography to women with high anti-Ro AB titers is safe and eliminated the need for ongoing exams in almost half of the referrals. Serial echocardiography allowed the detection of reversible immune-mediated cardiac injury in some but not all of our patients.

O12-4
Predicting Emergency Atrial Septostomy In Prenatally Diagnosed Transposition Of The Great Arteries

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Background: Prenatal detection of transposition of the great arteries (TGA) improves outcome, but mortality still occurs in the preoperative period due to inadequate mixing of the pulmonary and systemic circulations. Prenatal prediction of cases requiring emergency balloon atrial septostomy (BAS) remains difficult. It has been postulated that presence of oxygenated blood in the pulmonary artery causes relaxation of the pulmonary vasculature with subsequent increased pulmonary venous return predisposing to early postnatal closure of the interatrial communication. The objective is to identify prenatal markers which may predict the need for emergency BAS.
Methods: Prenatally diagnosed cases of isolated TGA in singleton pregnancies between January 2013 and December 2015 were reviewed. The appearance of the atrial septum (AS), the foramen ovale (FO) length and foramen ovale: total septal length (FO:TSL) were assessed at the third trimester fetal echocardiogram. The arterial valves, branch pulmonary arteries and arterial duct were measured from inner edge to inner edge in systole at maximal diameter. Assessments were made without knowledge of the postnatal outcome.
Results: 24 fetal echocardiograms were performed at a median gestational age of 34+ 2weeks (range: 29+ 0 to 36+ 2). An emergency BAS was required in 9/24 cases (table 1). All 3 cases with limited movement of the AS required emergency BAS. A hypermobile AS was not associated with emergency BAS (p = 0.60). An aneurysmal AS was identified in 21/24 cases and was seen more frequently in those cases which did not require an emergency BAS.
The FO length and the FO:TSL were significantly smaller in those who required an emergency BAS. An emergency BAS was required in 6/7 cases with FO < 6mm and 6/8 cases with FO:TSL < 0.4. There was no significant difference in arterial duct, semilunar valve, branch pulmonary artery size in those cases requiring emergency BAS and those that did not.
Conclusions:

1) Third trimester FO length and FO:TSL are smaller in those with inadequate mixing of pulmonary and systemic circulations after birth.
2) Third trimester FO < 6mm or FO:TSL < 0.4 should highlight the likelihood for emergency BAS.
3) Hypermobile and aneurysmal AS do not indicate inadequate postnatal mixing.

Table 1. Prenatal echocardiogram findings in TGA.

	Emergency BAS	No emergency BAS	P value
No. of cases	9	15	
Foramen ovale length (mm) (mean ± sd)	4.9 ± 1.0*	9.0 ± 2.8	0.005
Foramen ovale: total septal length	0.27 ± 0.07*	0.44 ± 0.11	0.015
No. of cases with aneurysmal AS	6/21*	15/21	P = 0.041
No. of cases with non-aneurysmal AS	3/3	0/3	
No. of cases with hypermobile AS	2/5	3/5	P = 0.60
No. of cases with mobile AS in left atrium	4/16	12/16	
No. of cases with fixed AS	3/3	0/3	
Arterial duct diameter (mm)	3.9 ± 1.3	3.6 ± 0.9	0.54
Pulmonary valve diameter (mm)	7.0 ± 1.2	6.2 ± 1.0	0.08
LPA z score	1.6 ± 1.1	1.3 ± 0.7	0.65
RPA z score	0.8 ± 0.4	0.4 ± 0.8	0.52

O12-5
Changes in right ventricular function after fetal pulmonary valvuloplasty in fetuses with pulmonary atresia with intact septum

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In-utero pulmonary valvuloplasty in fetuses with pulmonary atresia with intact ventricular septum (PAIVS) is technically feasible and should lead to beneficial changes of RV function with the goal to improve the chances for a postnatal biventricular circulation.
The aim of the study was to assess immediate changes in RV filling and function using 2D, Doppler and Tissue-Doppler measurements.
Patients: Since 2000 we performed 20 fetal pulmonary valvuloplasties in 14 fetuses with PAIVS (n = 8) or critical pulmonary stenosis (n = 6). Median gestational age at intervention was 27+ 6 weeks (23+ 6-32+ 1). The procedure was successful or partially

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successful in 12/14 (86%) and not successful in 2 patients. There were no fetal deaths. 8/12 children became definitive biventricular, 2 with one and a half ventricle, and 2 infants with BT Shunts still have an undetermined circulation. To assess immediate changes we measured RV morphologic and functional parameters 1-2 days before and 1-2 days after the procedure. Parameters were compared by paired student's t-test.
Fetal pulmonary valvuloplasty lowered RV pressures as measured by TR velocity and improved RV filling as assessed by longer RV inflow time and larger RV structures. RV Tei index decreased mainly due to shorter PEP.
Conclusion: Successful fetal pulmonary valvulopasty led to several significant immediate changes of RV dimensions and function. These changes should allow for beneficial in-utero RV remodeling and improved RV function, thus improving the chances for a postnatal sustained sufficient biventricular circulation.

	N	before - median (range)	after - median (range)	p-value
RV length z-score	10	-2,8 (-3,81–1,78)	-2,07 (-3,14–1,07)	0,026
RV/LV ratio	10	0,58 (0,51-0,67)	0,66 (0,59-0,87)	0,017
TV z-score	10	-1,43 (-2,52-0,39)	-1,23 (-2,22-0,74)	0,010
TV/MV ratio	10	0,85 (0,64-0,97)	0,78 (0,70-0,98)	0,307
RV inflow duration (%)	10	0,3 (0,19-0,4)	0,42 (0,30-0,46)	0,021
TR velocity (m/sec)	10	4,83 (3,28-5,5)	3,95 (2,10-5,38)	0,013
PV velocity (m/sec)	6	2,72 (2-3,3)	3,23 (2,06-4,4)	0,374
RV Tei index	10	0,89 (0,35-2,04)	0,57 (0,30-1,16)	0,030

O12-6
Prenatal Detection of Transposition of the Great Arteries does not reduce Mortality and Morbidity

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Background: Although neonates with transposition of the great arteries (TGA) often have an uncomplicated preoperative course followed by arterial switch operation with a low mortality risk, others become severely hypoxic and even die shortly after birth due to insufficient systemic arterial oxygenation. This retrospective study sought to review the impact of fetal diagnosis on neonatal morbidity and survival.
Methods: Included were 159 live-births with a fetal (n = 80) vs. neonatal (n = 79) diagnosis of TGA in Ontario from 2009-2014. Excluded were 4 fetal cases with pregnancy termination. Patients were identified from prospective databases. Collected data includes patient age at diagnosis, ICU admission and interventions, pre-operative health status, morbidity, and survival to 1-year of life. Student's t-test, chi-square test and Kaplan Meier estimates were used to compare the study cohorts.
Results: The table summarizes the main results.

	Fetal (n = 80)	Neonatal (n = 79)	P-values
Gestational age at birth (weeks)	39 (33-41)	39 (35-41)	0.02
Preadmission mortality	1/80 (1.2%)	-	
Age at hospital admission (hours)	1.4 (0.5-4)	10.5 (1-696)	<0.0001
Preoperative prostaglandin	79/79 (100%)	75/79 (95%)	0.12
Balloon atrioseptostomy	59/79 (75%)	68/79 (86%)	0.11
Age at PGE initiation (hours)	0.1 (0-1.7)	4.9 (0.8-643)	<0.0001
Age at atrioseptostomy (hours)	5.2 (1.3-655)	15 (3.5-645)	<0.0001
Age at surgery (days)	6 (3-68)	9 (3-62)	0.02
Length of hospital stay (days)	20 (10-175)	16 (8-99)	0.007
Mortality from admission to 1-year	1/79 (1.3%)	5/79 (6.3%)	0.21
Preoperative morbidity score	1 (0-4)	1 (0-3)	0.26
Postoperative morbidity score	0 (0-5)	0 (0-5)	0.89

Time intervals from birth to hospital admission, start of prostaglandin therapy, atrioseptostomy and surgery were significantly shorter for prenatally diagnosed livebirths. While hospital stays were significantly shorter for postnatally diagnosed cases, the cohorts were otherwise comparable, including in preoperative preductal oxygen saturation, pH, lactate, perioperative morbidity and one-year mortality rate.
Conclusions: The rate of prenatally diagnosed livebirths is about 50% in Ontario, suggesting the need of strategies to further improve detection rates. Prenatal diagnosis significantly shortened the time to neonatal tertiary care and surgery but did not improve the health status or survival of patients with TGA.

MP1-1
Maternal oral snuff during gestation alters heart rate variability in infants

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Introduction: Maternal smoking causes significant morbidity and mortality in the human infant. Following prenatal exposure, the newborn displays alterations in autonomic control and has increased risk for sudden infant death syndrome, SIDS.
Objective: This study aimed to establish whether snuff(non smoking tobacco) use by the mother during pregnancy is a less harmful alternative for the baby than smoking cigarettes.
Methods: A prospective observational study was conducted on infants (n = 56) of women who used snuff or cigarettes during pregnancy. Maternal nicotine use was determined both with questionnaires and with cotinine levels in urine of the infant. We studied heart rate variability in infants at 1-2 months of age with a 24 hour-ECG followed by spectral analysis of a 2 hour ECG segment during sleep. The outcome LF/HF ratio reflects the balance between the sympathetic and parasympathetic system.
Results: The LF/HF ratio was higher in snuff and smoke as compared to controls and was due to decreased parasympathetic activation. Prenatal nicotine exposure, both snuff and smoking, without any postpartal exposure showed an increased LF/HF ratio compared to controls. We could see no difference of non-smoke tobacco compared to cigarettes which suggests that the detrimental effects are mediated via nicotine. The alteration in autonomic regulation seems to be long lasting and due to neonatal reprogramming.
Conclusion: The results of this study indicates that nicotine is harmful for the developing infant in all forms. It thus emphasizes the importance of abstaining from all forms of nicotine during pregnancy and that there are no safe forms of tobacco or safe periods for its use during gestation.

MP1-2
Early outcomes following heart transplantation are not affected by stage of univentricular palliation

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Background: Following univentricular palliation, unfavorable factors might disqualify patients from progressing towards Fontan

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completion, necessitating heart transplantation (HT). HT for single-ventricle patients presents a difficult challenge and outcomes remain unclear according to the previous univentricular staging. We reviewed our experience in recent years.
Patients/Methods: From 2013-2015, 16 univentricular patients underwent HT (12 children, 4 adults). Clinical features/palliation stage: table. Primary diagnosis: HLHS (n = 9), PA+ IVS (n = 1), unbalanced AVSD (n = 1), tricuspid atresia (n = 3); DILV+ subpulmonary stenosis (n = 1) and TGA+ criss-cross+ VSD (n = 1). Post-operative complications/short-term outcomes were included. Comparative analysis between Fontan (n = 8) and single-ventricle non-Fontan (n = 8) patients was conducted (Fisher/Mann-Whitney tests).
Results: 4 patients (25%) were inotrope-dependent at listing. Berlin-Heart-EXCOR (bridge) was used in 1 patient. Median interval to HT was 90 ± -21 days (range 1-208). The following reconstructive techniques (bicaval technique) were performed: hemiarch repair (25%,n = 4), pulmonary artery (PA) plasty (31.2%, n = 5), hilum-to-hilum PA reconstruction (56.2%, n = 9), superior venae cavae (SVC) reconstruction (12.5%, n = 2) and stent removal from PA (56.2%, n = 9), inferior venae cavae (6.2%, n = 1) and lateral-tunnel-Fontan (6.2%, n = 1). Cardiopulmonary-bypass time was 244.6 ± 75.3 minutes (range 117-434); total-ischemia-time 217.3 ± 45.2 (range 139-283). Post-operative complications/early mortality: table. Post-operative ECMO was instituted due to ventricular dysfunction (n = 3). 1 patient developed subacute-humoral-rejection treated with plasmapheresis. No differences between post-op complications were detected. Survival was not affected by the previous univentricular stage (30-day mortality: intraoperative massive bleeding (n = 1) and sepsis following ECMO support (n = 1)). In-hospital stay was 46±16 days (range 23-161). At follow-up (14.4 ± -7.2 months), no mortality cases were detected; incidence for percutaneous interventions was higher for single-ventricle non-Fontan group (25% vs 0%; p < 0.04). All the survivors (n = 14) remain with optimal functional class.
Conclusions: HT is an effective option for patients following intermediate univentricular circulation with outcomes comparable to those with Fontan circulation. It can be performed with encouraging short-term results, reflecting current advances in surgical/perioperative management and immunosuppression strategies.

	SINGLE-VENTRICLE NON-FONTAN PATIENTS (n = 8)	FONTAN PATIENTS (n = 8)	p
Median age (years)	**Pediatric group** (n = 7): 6.1 ± 1.8 (range 1.5 months-9 years) **GUCH group** (n = 1): 25 years	**Pediatric group** (n = 5): 9.9 ± 1.2 (range 8-13.5) **GUCH group** (n = 3): 21.6 ± 0.6 (range 21-23)	–
Median weight (kg)	**Pediatric group** (n = 7): 18.5 ± 8.7 kg (range 3.7-36) **GUCH group** (n = 1): 64 kg	**Pediatric group** (n = 5): 28.4 ± (range 23-31) **GUCH group** (n = 3): 51.3 ± 8.6 (range 43-61)	–
Palliation type	Fontan take-down (n = 2); Glenn shunt (n = 5); BT shunt (n = 1)	Classical Fontan (37.5%, n = 3); extracardiac Fontan (62.5%, n = 5)	–
Post-op ECMO	12.5% (n = 1)	25% (n = 2)	p < 0.96
Delayed sternal closure	25% (n = 2)	12.5% (n = 1)	p < 0.94
Post-op percutan. procedures	25% (n = 2): RPA stent; ascending aorta stent	12.5% (n = 1): SVC stent	p < 0.34
30-day mortality	12.5% (n = 1)	12.5% (n = 1)	p < 0.71
Follow-up percutan. proc.*	25% (n = 2): SVC stent; RPA ballooning	0% (n = 0)	p < 0.04*

MP1-3
Arterial Stiffness and Motor Competence in Children and Adolescents with Univentricular Heart After Total Cavopulmonary Connection
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Objective: Studies suggest that patients with congenital heart disease have increased arterial stiffness and impaired motor competence. The aim of this study is to investigate whether current patients after total cavo-pulmonary connection (TCPC) still have augmented arterial stiffness and deficits in motor competence.
Patients and Methods: From July 2014 to December 2015, 58 patients (15 girls, 11.8 ± 3.3 years) with TCPC underwent an arterial stiffness measurement and performed a motoric test with five tasks to assess muscle strength (push-ups and curl-ups), lower back strength, shoulder and harmstring flexibility. Arterial stiffness was measured using the oscillometric Mobil-o-Graph.
Data was compared to a contemporary references pool of 1964 healthy subjects (956 girls, 12.7 ± 2.4 years) recently tested in different Bavarian schools. Patients' data was compared to healthy controls using a multivariate regression model with correction for age, sex and BMI.
Results: Central systolic blood pressure, a surrogate for arterial stiffness, was 4.0 mmHg higher in TCPC patients compared to healthy peers (B = 4.0, p < .002), whereas there was no difference in peripheral systolic blood pressure (p = .329). Motor competence of TCPC patients was impaired in all of the five performed motor tasks. Patients performed 2.4 push-ups (p = .041) and 10.0 curl-ups (p < .001) less. Moreover, they had impaired lower back strength (B = -9.0 cm, p < .001), and shoulder (B = -7.9 cm, p < .001) and harmstring flexibility (B = -5.3 cm, p < .001).
Conclusions: In patients with TCPC central systolic blood pressure, a surrogate of arterial stiffness, is increased and motor competence in terms of strength and flexibility is impaired. Therefore measuring both during routine follow-up is recommended and treated if indicated.

MP1-4
Intima Media Thickness Seems to Be Increased in Patients with Congenital Heart Disease in Comparison to Healthy Subject
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Introduction: Carotid intima media thickness (IMT) is a strong predictor of cardiovascular events and can be used as a risk marker of atherosclerosis. Studies suggest that patients with congenital heart disease (CHD) have an increased cardiovascular risk and impairments in arterial compliance. Therefore, this study compares the IMT of patients with CHD with a healthy control group and also focus on the potential correlation to exercise capacity.
Patients and Methods: From August 2015 to December 2015, 123 patients (51 female, 24.7 ± 11.9 years)with various CHD and 56 healthy volunteers (35 female, 30.5 ± 12.4 years) received an assessment of the IMT at the A. carotis communis with ultrasound. Afterwards they performed a cardiopulmonary exercises test.
Results: Mean IMT of patients with CHD was 0.492 ± 0.080 mm. IMT was associated with age (r = .668, p < .001), systolic blood

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pressure (r = .327, p < .001) and BMI (r = .179, p = .017). After adjusting for age, sex, BMI and systolic blood pressure in a multivariate regression model, patients with CHD tended to have higher IMT (B = .019; Beta = .106, p = .056) in comparison to healthy counterparts. Especially patients with left heart obstruction (B = .043, p = .002) and patients with univentricular heart after total cavopulmonary connection (B = .036, p = .055) seem to be at a higher cardiovascular risk. Exercise capacity was not associated with IMT (p = .421).
Conclusion: After adjusting for several confounders patients with CHD present higher IMT than healthy subjects, especially those patients with left heart obstruction and with univentricular heart after total cavopulmonary connection. More research with higher patient number is needed for diagnostic subgroup evaluation and to elucidate the association to exercise capacity.

MP1-5
Arterial stiffness evaluated by cardio-ankle vascular index (CAVI) in healthy Slovak children
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Purpose: Recently, the cardio-ankle vascular index (CAVI) is considered as a novel noninvasive index of the arterial stiffness from the beginning of the aorta to the ankle. CAVI represents an important marker of early atherosclerotic changes that is significant for a consequent evaluation of the atherosclerosis severity. We aimed to determine the CAVI values in the group of healthy children and adolescents necessary for a comparison with CAVI values in pathological states. Moreover, the second aim was to assess the age and gender influence on the CAVI in healthy children and adolescents.
Methods: We examined 520 healthy Slovak children at the age from 7 to 19 years (260 boys) without clinically observed cardiovascular risk factors. CAVI values were evaluated using the system VaSera 1500 (Japan).
Results: The CAVI normal values are presented in graphical forms for total group, and separately for boys and girls. In healthy children, the CAVI increased linearly with age from 7 to 19 years. Gender analysis did not show significantly differences between boys and girls at this age-period. Additionally, CAVI values were independent on the blood pressure measurement at the same time.
Conclusion: Our study firstly presented the CAVI normal values for Slovak population of children and adolescents at the age from 7 to 19 years. Importantly, the CAVI was dependent on the age during this developmental period. Concluding, our CAVI values can be used for detection of the early atherosclerotic changes in children with essential hypertension.

MP1-6
Children and adolescents with congenital heart defect show equal or even better health related quality of life than healthy controls
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Introduction: Improved treatments for patients with congenital heart disease (CHD) decrease the mortality rate and lead to a growing importance of long-term outcomes like health related quality of life (HRQoL). Although there already exist numerous studies, an influence of CHD on HRQoL is not certain. Especially the results concerning the impact of particular diagnoses are not homogenous, mostly even contradicting.
Methods: The cross-sectional study included 390 patients (143 girls) aged 6 to 17 years (12.7 ± 3.3 years), recruited in the German Heart Centre Munich between July 2014 and December 2015. To evaluate the HRQoL an age-adapted questionnaire (KINDL®) for self-report was used which includes six subcategories: physical, psychical, family-, social-, and school-related well-being as well as self-esteem. The data of the patients were compared to 734 healthy children (346 girls, 13.5 ± 2.1 years), which were examined between 2011 and 2013. Three analyses were performed: (1) comparison of healthy and CHD, (2) comparison of severity class simple (n = 98), moderate (n = 86) or severe heart defect (n = 166), (3) diagnostic subgroup analysis of patients with Aortic Coarctation/Aortic Stenosis (CoA/AS n = 81), Tetralogy of Fallot/Pulmonic Stenosis (ToF/PS n = 55), Atrial Septal Defect/Ventricular Septal Defect/Atrioventricular Septal Defect (Shunts n = 63), Transposition of the Great Arteries (TGA n = 44), Univentricular Heart (UVH n = 50). In all analyses the influence of age and sex was controlled by a multivariate regression model.
Results: Current children and adolescents with CHD have a better HRQoL compared to healthy peers (CHD: 78.6 ± 9.7 vs. healthy: 75.6 ± 10.1; p < .001). Still after correction for sex and age children with CHD present a 2.1 higher HRQoL (B = 2.1, p = .001). In the subcategories family-related well-being (B = 2.4, p = .009), school-related well-being (B = 7.3, p < .001) and self-esteem (B = 3.1, p = .005) they show even higher values. Children with simple, moderate or severe heart defect do not differ in a single domain. There was no difference in-between the diagnostic subgroups, only children with shunts showed worse HRQoL results (B = -3.1, p = .007).
Conclusion: Children with CHD can cope well with the adverse impact of their disease so that no negative influence on HRQoL is evident. This holds true for all severity classes and almost all diagnostic subgroups.

MP1-7
Impact of Exercise Training on Arterial Wall Thickness and Distensibility in Young Competitive Athletes
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Objective: The thickening of the carotid artery wall has been described as a marker of early atherosclerosis. Exercise is associated with decreased CV risk and higher fitness confers cardioprotection. However, studies examining the impact of exercise training on arterial wall thickness in healthy young adults lead to contradictory results. Little is currently known regarding exercise and remodeling processes with regard to diameter (wall-to- lumen ratio (W-to-L-ratio)) and arterial stiffness of the carotid artery. The aim of this prospective study was to examine the carotid intima-media thickness (cIMT) in young competitive athletes compared to reference values. Further to analyze associations between cIMT, W-to-L ratio as well as intraventricular septal thickness (IVSd), leftventricular posterior wall dimensions (LVPWd) and exercise performance.
Methods: cIMT, W-to-L-ratio and arterial stiffness of the A. carotis communis were measured with ultrasonography in elite youth

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soccer players (n = 34 boys, age 14-18 years, training duration 15-20 h/week). The athlete's performance was quantified by cardiopulmonary exercise testing, measuring the percentage of oxygen consumption (VO2%) and the relative exercise performance (Watt/kg). Echocardiography was performed to evaluate myocardial wall thicknesses.
Results: cIMT (cIMT_right 0.531 ± 0.03mm; cIMT_left 0.519 ± 0.04mm) were above the 75th percentile compared to age-matched reference values. Multivariate regression analysis revealed a significant model on cIMT_right only (adjusted for age and blood pressure) and could explain 17.4% of the variance (V02%: ß = 0.000 p = 0.312; relative performance (Watt/kg) ß = 0.037, p = 0.024, $R^2 = 0.174$. Regarding cardiac structure (IVSd: 10.1 ± 1.8 mm; IVSd z-score: 1.13 ± 0.88; LVPWd: 8.6 ± 1.57, LVPWd z-score: 0.80 ± 0.85) and arterial stiffness no outstanding detection or significant correlations occurred.
Conclusion: An increased cIMT in young athletes might be considered as a vascular adaptation to exercise. Like adaptation mechanisms in athlete's heart, the thickening could be interpreted as hypertrophy of smooth muscle cells, in order to economize the arteries work under elevated shear stress. The mechanisms explaining changes in the arterial wall by revealing the arterial elasticity as a result of exercise training are not fully understood. Identifying these stimuli in a larger cohort will help in the design and recommendation of optimal exercise training protocols to attenuate atherosclerosis burden and risk.

MP1-8
Peripheral Vascular Endothelial Function is Impaired in Childhood Cancer Survivors
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Introduction: Childhood cancer survivors are a growing population and have an earlier than expected morbidity and mortality from cardiovascular disease. Early identification of subclinical cardiovascular illness could help to prevent these adverse outcomes. We assessed peripheral vascular endothelial function using reactive hyperemia peripheral arterial tonometry (RH-PAT). Augmented reactive hyperemia index (RHI) has been linked in several studies to increased risk for future cardiovascular events.
Methods: 47 childhood cancer survivors with age 20-30 years without previous cardiovascular events and at least 5 years of freedom from cancer disease and 24 healthy controls were evaluated for RHI using the ENDO-PAT 2000 (Itamar Medical). Data on body mass index, arterial blood pressure, and subjective fitness (using a questionnaire) were collected.
Results: Nearly one-third of cancer survivors (31.2%) compared to only 8% of controls (p = 0.02) had RHI below the cut-off value of 0.5 (log). There was no association between this index and the patients´ cancer duration and therapy (p > 0.2).
Conclusion: Asymptomatic survivors of childhood cancer have subclinical signs of microvascular disease. This method could prove useful in prevention of cardiovasvular disease in this cohort.

MP1-9
Adapting Pulse Oximetry Measurements for Early Detection of Critical Congenital Heart Disease in Early Neonatal Period
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Introduction: The prevalence of Congenital Heart Disease (CHD) is worldwide 1% of all living births. 25% of them will suffer from critical CHD (c-CHD). Common examples: transposition of great arteries, tetralogy of Fallot, tricuspid valve atresia, pulmonary valve atresia, truncus arteriosus, total anomalous pulmonary venous drainage, critical aortic valve stenosis, severe coarctation of the aorta, interrupted aortic arch and hypoplastic left heart syndrome. These patients need early detection and surgical treatment in the first month of life. As fetal echocardiography is limited to significant indications and pre discharge from maternity wards physical examination carries a significant false negative diagnosis any new screening test for detecting c-CHD is vital.
Aim: of this paper is to draw the awareness of the medical practitioners that deal with neonates on the benefits of pulse oximetry measurements both pre and post ductal after the first day of life to detect c-CHD. Additionally, we are in the process of creating guidelines for the use of this test as a screening test in the early neonatal period.
Method: Review of literature on the subject.
Results: This test has been used today in many countries around the world. USA and China are among those. In Europe only a few countries have adopted it as a screening test. Others are running national studies to detect its use.
Discussion: The sensitivity of the test has been reported in various large multicenter studies to range from 63-83% and the specificity from 97.9-99.83%. From the false positive detected cases, approximately: 41% had other cardiac congenital or acquired diseases, 14% suffered from primary sepsis, 10% from respiratory disease. Pulse oximetry screening seems to be a very valuable screening test to detect c-CHD and other serious morbidities in early life. It's cheap and handy use must be encouraged by national legislation globally and our near future guidelines can assist this task.

MP1-10
Cerebral MR Morphometry and Neurodevelopmental Outcome in Children before Fontan procedure at 2 years of age – White and grey matter volumes are larger after Hybrid procedure
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Objectives: Advances in cardiac surgery and intensive care have reduced mortality in patients with congenital heart disease (CHD). Improvement of neurodevelopmental outcome is one of the most important goals for children with complex CHD. Delayed brain development has been described preoperatively in newborns with CHD, affecting mainly white matter (WM) structures. Perioperative complications may additionally cause brain injury. We set out to study brain volumes in children with hypoplastic left heart syndrome (HLHS) or univentricular heart (UVH) at the age of 2 years prior to Fontan surgery.
Methods: Prospective two-center cross-sectional study. Cerebral MRI (21 Zurich patients: 3 Tesla GE MR 750 scanner; 23 Giessen patients: 3 Tesla Magnetom Verio B17 Siemens scanner; FreeSurfer image analysis) was performed before Fontan.

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Morphometric findings were correlated with results from the Bayley Scales of Infant and Toddler Development III (Bayley III).
Results: 44 patients (male 28; age mean 26.7 ± 3.9 months) with HLHS (25/44) and non-HLHS (19/44) were included, treated by Hybrid or Norwood procedure. The average brain volumes derived were: 909.1 ± 83 ml for total brain volume, 277.1 ± 30 ml for WM, 610.5 ± 59 ml for gray matter (GM), and 42.7 ± 5 ml for subcortical GM. Ventricular CSF volume was larger in patients with abnormal neurologic examination (i.e. tonus and/or abnormal reflexes) and correlated with poorer Bayley III scores (CSS: rSpearman's Rho = -.31, p = .043; LCS: rSpearman's Rho = -.40, p = .009, MCS: rSpearman's Rho = -.31, p = .04). Brain volumes did not differ between type of UVH (HLHS vs. non-HLHS; p values all >.07). Subcortical GM (p = 0.01) and WM (p = .05) volumes were larger in patients receiving Hybrid procedure as first surgery. However, patients after Hybrid did not show significantly higher Bayley III scores (p > .69). Brain volumes were not different in infants with cerebral lesions compared to those without (p-values all >.38)
Conclusions: Cerebral MR morphometry is a good tool to measure brain volumes in children with CHD. In children undergoing Fontan procedure, larger ventricular CSF volume correlates negatively with neurodevelopmental outcome. WM brain volume loss was associated with the surgical approach, but not with the subtypes of CHD. Perioperative management in combination with routine neurodevelopmental follow up programs are needed to further improve outcome.

MP1-11
Aortic arch reconstruction in the Norwood procedure using a curved polytetrafluorethylene patch

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Objective: Children with hypoplastic left heart syndrome and related malformations inherently have an hypoplastic aortic arch. The aortic arch reconstruction in the Norwood procedure is classically carried out using an enlarging curved homograft patch on the inner curvature of the neoaorta. The widespread lack of homografts makes an alternative patch material a desired product.
Methods: Between 4/2007 and 12/2014 we alternatively used curved polytetrafluorethylene (PTFE) patches known from peripheral vascular surgery in 76 out of 171 Norwood procedures. The suture was done using prolene 6/0. In the other operations pulmonary homograft patches were implanted. The decision for either patch material was made due to anatomic reasons, preferring PTFE patches in larger aortas.
A retrospective analysis was carried out concerning postoperative course and long term follow up regarding aortic arch interventions and reoperations.
Results: The arch augmentation with the prosthetic material could be carried out in all 76 preoperatively selected patients. There were no material associated operative or postoperative complications. There were no significant differences regarding aortic clamp time or bypass time between the groups. 30 day mortality was 6.5%. Presently 22 pts. are in stage II and 36 pts. after Fontan completion. 9 children died late during follow up (12.7%) and 4 are lost to follow up.
One aortic isthmus dilatation was carried out 12 months after the Norwood procedure, no arch reoperation was necessary. In the remaining patients the aortic arch didn´t show need for intervention during the complete follow up. Potential gradients were assessed echocardiographically and invasively at time of angiography for fenestration closure.
Conclusions: The curved PTFE patch showed good qualities in operative technical demands and excellent long term results. In selected cases of hypoplastic left heart syndrome it can be well used as alternative to the commonly used pulmonary homograft. Its advantages are its availability, durability and lower costs. Its stiffness makes the application in very tiny aortas difficult.

MP1-12
Bicuspid neoaortic valve in patients with transposition of the great arteries after arterial switch operation: 25 years' experience

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Introduction: The function of the native pulmonary valve in systemic circulation in patients with transposition of the great arteries (TGA) after arterial switch operation (ASO) still remains the matter of concern and the neoaortic insufficiency is one of the most frequent abnormalities found in these patients in postoperative observation. As the native bicuspid aortic valve is more frequently associated with fibrosis and calcification which lead to its stenosis, insufficiency and root dilatation, patients after ASO with bicuspid neoaortic valve (NeoAoV) should be identified and the function of systemic valve needs to be watched carefully.
The aim of this study was to establish the frequency of bicuspid native pulmonary valve in patients with TGA, correlate its presence with major postoperative complications and establish the natural course of its function in systemic circulation.
Methods: For this retrospective case review study we included all 716 patients TGA who underwent ASO in our institution between years 1991-2015. All of the surgical procedures were performed by one cardiac surgery team lead by JJM, using the same surgical technique with his own modifications. The presence of pulmonary bicuspid valve was identified on the basis of data from surgical protocols.
Results: Bicuspid native pulmonary valve was present in 39 patients (5.4%). Early mortality in this group was 5,1%(2 patients) and mean follow up was 8,1 years (range 0,1-21 years). During postoperative observation neoaortic insufficiency developed in 21 patients (57%; 6-trivial; 10-mild; 4-moderate, 1-severe). Significant PS was observed preoperatively in 3 cases, 2 of them in postoperative period remains insignificant (PG < 16 mmHg), in one case reoperation (LVOTO) and catheter intervention(aortic valve balloon plasty) was performed. No significant stenosis developed postoperatively. In multivariate analysis bicuspid NeoAoV was not a significant risk factor for early mortality (p = 0.57); reoperations (p = 0.63), catheter interventions(p = 0.77) and neoaortic insufficiency (p = 0.52). Bicuspid NeoAoV was significantly more frequent in patients with VSD associated with TGA (p = 0.037) and correlated significantly with the non-facing commissures (p < 0.001) and patch reconstruction of the neopulmonary artery (p = 0.031).
Conclusions: In the analysed period of time bicuspid neoaortic valve did not increase the risk of postoperative complications, however this group of patients needs to be followed as the function loss may show at later stage.

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MP1-13

Porcine pulmonary prosthesis to repair the dysfunctional right ventricle outflow tract. Does it the same in children than adults? A word of caution

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Objectives: Pulmonary valve replacement for repair the dysfunctional right ventricle outflow tract (RVOT) is increasing in childhood to prevent the dysfunction of the right ventricle (RV). The question is whether results are so good in children than in adults. Our goal is to compare the results of the stented pulmonary porcine prosthesis (PPP) in older and younger than 18 years.
Methods: All patients who received a PPP between 1999-2015 for repair the sequela after primary surgery on the RVOT. Prosthetic dysfunction criteria: surgical/percutaneous reintervention, prosthetic gradient >50 mmHg or severe prosthetic regurgitation. Statistical analysis with SPSS 20.0.
Results: 102 PPP/101 patients (81/81 > 18; 21/20 < 18). 60% male. Fallot, most common primary disease in both groups. From 24 preoperative variables studied, statistically significant differences occur in 4: last surgery mean age before PPP, $p < 0.001$; NYHA status, $p = 0.005$; QRS time, $p = 0.007$ all of them greater in adults. Surgical indication (pulmonary regurgitation/pulmonary stenosis/double pulmonary lesion), $p = 0.036$, with more pulmonary stenosis in group <18 years.
Overall hospital mortality: 2.9% (3.7% adults vs 0% children, $p < 0.001$). If PPP is a single procedure mortality is 0%. From 14 perioperative variables, 7 were statistically significant, highlighting cardiopulmonary by-pass (CPB) with peripheral access, $p < 0.001$; associated surgical procedure, $p = 0.02$; CPB time $p = 0.01$; aortic cross-clamp need, $p = 0.043$; intubation time, $p < 0.001$, all variables greater in adult group. Complications in 32% of cases, most often tachyarrhythmia. No significant differences
Mean follow-up time 4 ± 3.7 years (4,4 adults vs 2.4 children, $p = 0.017$). Only 1 late dead in over 18 years group. From 16 postoperative variables, 6 proved statistically significant differences, highlighting systolic/diastolic RV volumes, $p < 0.001$, greater in adults. The need for surgical/percutaneous reintervention, $p = 0.029$ and reoperation for prosthesis dysfunction ($p = 0.001$), in contrast, are higher in <18.
Conclusions: The PPP improves the dysfunctional RVOT with low in-hospital and follow-up mortality in both groups of age. PPP dysfunction is greater and earlier in children than in adults, so the procedure must be indicated with caution in young people.

MP1-14

The left ventricular outflow tract after arterial switch operation: the benefit of geometry preservation

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Basics: The arterial switch operation (ASO) is the standard operation for transposition of the great arteries (TGA). Pulmonary stenosis remains the most frequent late complication, beside potential early and late coronary artery problems. The evolution of the left ventricular outflow (LVOT) tract is less frequently analyzed. The aim of this report is to examine the long-term outcome of the LVOT after ASO.
Methods: We retrospectively studied 271 patients who underwent ASO between January 1985 and December 2008 in our institution. Anatomic data and surgical procedure details were collected. The aim of the surgery was to preserve the neo-aorta sino-tubular junction and to use direct coronary reimplantation, whenever possible. The geometry of the neo-aortic root was as a result, mostly preserved. Echocardiographic data, and when available, MRI or CT-scan follow-up data were collected and analyzed.
Results: Median age at time of ASO was 10 days (1-497). 75,6% (n = 205) were male. Follow up reached 12,12±- 6.62 years (maximum 29.82 years). Direct reimplantation without any trap door technique was done in 60% of the cases. Two deaths from cardiac causes (0,7%) were encountered. Mean Valsalva sinus diameter Z-score was +3,72 (42.4 mm for adults). An aortic root z-score > 3 or diameter > 40 mm (in adults) was found in 57 patients (22.7%). Length of follow-up and male gender were significantly associated with aortic dilatation. Moderate aortic insufficiency was seen in only 5 patients and none had severe regurgitation. No significant risk factor for aortic insufficiency could be identified. There were no aortic or coronary events and only one patient required an elective LVOT surgery (subvalvar stenosis).
Conclusion: Our results, when compared to other literature data, show a very low incidence of significant aortic regurgitation or dilatation of the neo-aortic root, with no re-intervention. Our hypothesis is that this results from a deliberate effort to conserve geometry during coronary reimplantation. This effort at the time of neonatal surgery is probably part of the way to lower the incidence of root dilation and aortic insufficiency and is possible without increasing coronary risk.

MP1-15

Diaphragm plication yields similar results in patients following univentricular palliation and biventricular repair. A 10 years experience

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Background: Adequate respiratory function is essential following univentricular palliation. Hence, diaphragm paralysis after staged Fontan surgery due to uni- or bilateral phrenic nerve palsy may require prolonged mechanical ventilation. In this setting, diaphragm plication has been proposed.
Objective: We sought to determine incidence and results of diaphragm plication focusing on potential differences between patients following biventricular repair and univentricular palliation in our center during a 10 years experience period.
Design/Methods: A retrospective analyse of all consecutive patients who underwent diaphragm plication between 2005 and 2015 was performed. Outcomes were compared between the groups following univentricular palliation and biventricular repair.
Results: Diaphragm plication was performed in 25 patients in 27 phrenic nerve palsy identified cases following 8120 procedures (0.3%). 11 of them presented with functional univentricular heart (7 following a Glenn anastomosis,1 TAPVR repair , 1 Blalock Taussig shunt, 1 total cavopulmonary connection, 1 unknown procedure). The remaining patients had undergone biventricular repair (3 AVPR, 5 VSD, 1 GVA, 1 Fallot,1 pulmonary valve agenesis,1 troncus and 1 aortic coarctation). Mean age at plication was 10 months in univentricular group and 2.1 in biventricular one ($p = 0.65$), with median delay between surgery and plication of 16 days in uni and 14 days in biventricular surgeries, with a trend to be shorter now. There was no hospital mortality in both groups.

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Mean time on mechanical ventilation, mean time on the intensive care unit, and mean time of hospitalization after plication was 24 hours, 3 and 10 days in univentricular group and 24.5 hours, 3 and 11 days in biventricular one, respectively (all p values > 0.05).
Conclusions: The incidence of diaphragmatic paralysis following congenital heart surgery is very low. Diaphragm plication is an effective and safe treatment of diaphragmatic paralysis. The procedure yields similar results in patients presenting with functional univentricular heart and in patients following biventricular repair. Early application is warranted to prevent prolonged ventilation and length of stay.

MP1-16

Cone reconstruction for Ebstein's anomaly: What defines the need for additional bidirectional cavopulmonary anastomosis?

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Objectives: We aimed to investigate whether the tricuspid valve (TV) diameter after "cone reconstruction" (CR) as primary biventricular, "anatomical" correction of Ebstein's anomaly decisively influences the need for additional bidirectional cavopulmonary anastomosis (BCPA), s.c. "one-and-a-half" repair.
Methods: Retrospective echocardiographic analysis of the postsurgical TVannulus in 4-chamber view (mean value of 4 measurements) in patients who underwent CR between 06/2013 and 12/2015, determination of the Z scores (Detroit) and correlation with the bi or "one-and-a-half" result.
Results: Thirty patients (median age 4 (6 months-36) years, median weight 18.6 (6.6-74) kg) received a CR, 14 (52%) of them with and 16 (48%) without BCPA. An additional pulmonary valve repair or replacement was necessary in 7 patients.
The was no mortality and the early postsurgical course was uneventful (median duration of the mechanical ventilation 19 hours and hospital stay 8 days) except for temporary ECMO support in one patient. Central venous pressure at the end of the operation was in median 12 mmHg and showed no correlation with BCPA necessity. According to the Carpentier classification of tricuspid valve dysplasia (class A: 2 (6%), B: 7 (23%), C: 16 (53%), D: 3 (10%)) we found no correlation with regard to BCPA. Postsurgical TV regurgitation was absent in 13 (43%), low in 13 (43%) and mild in 4 (14%) patients.
The mean diameter of the TV annulus and equivalent Z score were significantly lower ($p = 0.002$ and $p = 0.01$, respectively) in the patients who needed an additional BCPA.
Conclusion: The resultant diameter of the TV created using CR seems to be co-decisive: in the case of Z score > -2 the additional BCPA may be avoided even with severe dysplasia of the TV; however, it seems to be necessary with smaller TV diameter/Z score. Intraoperative TV-Z score determination after cone reconstruction may be helpful in the decision-making with regard to 'biventricular' or "one-and-a-half" correction of the Ebstein anomaly.

MP1-17

Double arterial cannulation for aortic arch reconstruction in newborns and infants

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Introduction: Aortic arch reconstruction (AAR) is a highly demanding procedure in congenital heart surgery. The two conventional options of cardiopulmonary bypass - deep hypothermic circulatory arrest or antegrade cerebral perfusion – both mean lack of perfusion of the entire or lower body. 1999 Imoto et al. presented a perfusion technique with a second arterial cannula into the descending aorta (double arterial cannulation, DAC). However, there is still limited experience with large series of patients.
Methods: All operations including AAR in newborns and infants performed in our institution between 2003-2013 were reviewed. Median age of our 348 patients at time of operation was 8.0d [6; 16], median weight 3.3 kg [2.9; 3.8]. The cohort was divided into 3 groups: simple biventricular (1), complex biventricular (Taussig Bing Heart etc) (2) and univentricular (Norwood procedure, others) (3).
The cannulation technique has not been changed substantially over the years.
We used descriptive analyses to evaluate i.a. complications or in hospital mortality and multiple linear regression analyses to investigate influence of variables on lactate and creatinine related endpoints.
Results: There was no lethal incident associated with DAC. We evaluated median in hospital mortality between 2.6% in group 2 and 10.1% in group 3. Postoperative lactate decreased to 2 mmol/l after 12 hours at the latest in group 3. The highest creatinine levels on postoperative day 3 were found in group 3 (0.66 mg/100 ml). 7 patients (2.2%) required peritoneal dialysis postoperatively. As complications of DAC we found 1 intraoperative local lesion of descending aorta (repaired immediately), 1 reoperation and 4 secondary pleural drainages due to prolonged chylothorax.
Conclusion: DAC is a simple and save method for perfusing lower parts of the body in AAR. Dissection of the descending aorta has to be done with great attention on local lymphatic vessels. Perioperative results as in hospital mortality or lactate levels and necessary reoperations or peritoneal dialysis are satisfying. Comparison to patient cohorts with other perfusion techniques in our centre wasn't done, since we never stepped back to other perfusion techniques due to the simplicity of DAC.

MP1-18

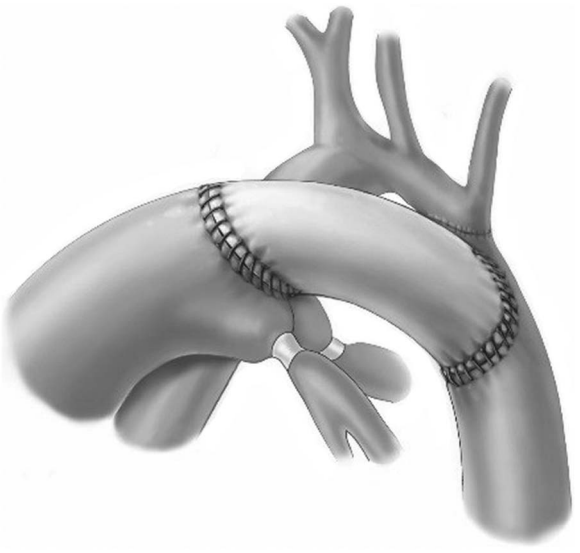
Reversed shunt and bilateral branch pulmonary artery banding as bailout for backward left heart failure after adequate neonatal coarctectomy in borderline left heart

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Introduction: An adequate neonatal coarctectomy with good gradient relief may still result in circulation failure due to borderline small left heart. We present bail-out procedure for backward left heart failure after coarctectomy.
Methods: in 2005 – 2015 89 neonates presented with 'isolated' critical coarctation and variable degree of small left heart. Nine patients underwent a hybrid procedure (ductal stent and bilateral PA banding) because of borderline left heart.
Results: Eighty had extended end-to-end coarctectomy; 2 developed, despite adequate coarctectomy, after initial stable hemodynamics, suprasystemic retrograde pulmonary hypertension with severe RV dilation and compression of the small LV with progressive circulatory failure non-responsive to medical treatment.
Pt1 (4,4 kg, coarctectomy D5); patient 2 (3,7 kg, coarctectomy D7, progressive lactate and renal failure). Through a sternotomy a "new duct" or "reversed shunt" procedure was created as an urgent bailout respectively 9 and 7 days after the coarctectomy:

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6 mm Goretex® graft between the main pulmonary artery and thoracic aorta, 3.5mm Goretex® bands around branch pulmonary arteries. Immediate circulatory improvement was observed with extubation 4 and 7 days later.
The left heart was allowed to grow; Pt1 evolved to Damus–Kaye and Fontan circulation, Pt2 had 4 months later biventricular repair by debanding, shunt take-down and arch optimalisation.
Conclusion: Backward left heart failure after neonatal coarctectomy can be managed by surgical construction of a reversed shunt in combination with bilateral PA banding. If high risk for backward failure, a primary hybrid procedure is a more elegant strategy.



MP1-19
Experience with Berlin-heart Excor® devices in children in south of France
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Background: Techniques for long-term circulatory support in pediatrics are limited and the number of implantations remains low when compared to the adult population. Description and analysis of follow-up data are essential in order to progress on indications, technical difficulties and management of complications.
Methods: A retrospective observational multicenter study was conducted in the south of France. All 3 university hospitals (Bordeaux, Lyon, Marseille) performing circulatory support in children participated. All children (<18 years of age) who needed a long-term ventricular assist device support were included. Only the Berlin Heart EXCOR® was used. We sought to analyze the morbidity and mortality data in this population and identify risk factors for complications and death.
Results: 45 patients were included. Median age at time of support was 16 months (range: 3-156), 49% were males. The predominant indication for circulatory support was cardiomyopathy (97.2%). In 53% the assist device was biventricular, the remaining had left assist device. 35.6% patients had extracorporeal membrane oxygenation-support before implantation of the Berlin-heart Excor. The total amount of days of circulatory support for the whole group reached 2272 days, with a mean duration of 34 days (range: 0-199 days). 62% of patients were transplanted, 11% were weaned and 27% died. Complications occurred in 82,2% of the patients and were mainly infections (48.9%), severe bleeding (35.8%), stroke (35.6%) and non-neurologic thromboembolic events (37.8%). Preimplantation disorders of consciousness were significantly associated with the occurrence of stroke ($p < 0.005$, log rank test). The number of complications tended to increase with the ventricular assist duration, initial value of BNP (or pro-BNP) and the presence of initial hepatic dysfunction. Male gender was also significantly associated with higher mortality or severe complications (composite criteria - $p < 0.005$-Fischer Test).
Conclusion: Despite the severity of the underlying disease, the length of the support and the high number of serious complications, the survival was encouraging (73%) in our population and comparable to other published series. Better control of infectious events and more homogenous anti-coagulation protocols are required. More detailed follow-up, in particular with regards to long-term neurological development is required.

MP1-20
Outcome of arterial switch operation for Taussig Bing anomaly versus transposition of the great arteries and ventricular septal defect
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Objective: Patients with Taussig Bing anomaly (TBA) usually considered to have increased mortality and morbidity when compared to those with transposition of the great arteries and ventricular septal defect (TGA+ VSD) after arterial switch operation (ASO). In this report we analize our results in those patients.
Methods: Between November 2010 and December 2015, 94 ASO was performed. Among them 36 patients (38,2%) had associated VSD and was diagnosed as TBA ($n = 14$) or TGA+ VSD ($n = 22$). Median ages were 17 days (range: 6-62 days) and 16 days (range: 2 days-7 months) respectively ($p = NS$). Six patients had aortic arch anomalies (coarctation $n = 4$; interruption $n = 2$) in TBA group (42,8%), while 3 patients had aortic coarctation in TGA+ VSD group (13,6%) ($p = 0.11$). Coronary anomaly was present in 3 (21,4%) and 6 (27,2%) patients respectively ($p = NS$). All VSDs were large in size and subpulmonary (except one with multiple VSD) in TBA group. Neveretheless VSDs were large in 14 (multiple VSD $n = 3$) and moderate in 8 patients in TGA+ VSD group. Only 1 patient had previous palliative surgery in TGA+ VSD group. Besides ASO, VSD closures were performed via transneoaortic approach in 10 patients with TBA. Others had transatrial approach. Pericardial patch was used in most of the patients except 7 patients who had moderate sized VSD, underwent primary VSD closure. One patient in each group had additional pulmonary banding because of multiple VSD or apical VSD.
Results: Early mortality was 1 patient in TBA group and 2 patients in TGA+ VSD group (mortality: 7.1% vs 9% respectively;$p = NS$). Delayed sternal closure was used in 85.7% and 72.2% respectively; $p = NS$). One patient needed ECMO support in each group. One of them (in TBA group) weaned and discharged uneventfully. Long duration of ventilatory support was necessary for 3 (21.4%)

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and 4 patients (18,1%) respectively (p = NS). One patient underwent permanent pacemaker implantation in TGA+ VSD group. One patient in TBA group had VAC therapy due to mediastinitis.
Conclusions: Although the incidence of aortic arch anomalies is higher in TBA group, early and intermediate term outcomes are similar with TGA+ VSD group.

MP2-1

Inhibition of histone acetylation by curcumin reduces alcohol-induced fetal cardiac apoptosis

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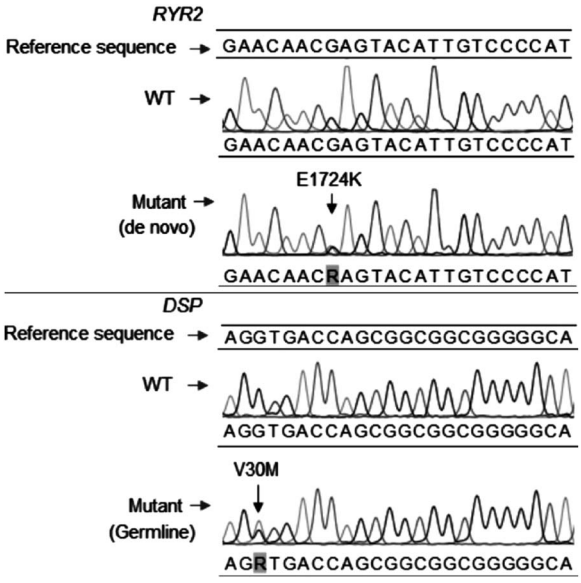
Background: Prenatal alcohol exposure may cause cardiac development defects. It is known that alcohol could induce cardiac apoptosis and myocardium dysplasia. However the underlying mechanisms are still not clear. Our previous studies suggest that histone modification play a vital role in alcohol induced fetal cardiac development abnormalities. So, the objective of this study was to investigate the effect of histone acetylation regulation mechanisms on alcohol induced cardiac apoptosis.
Methods and Results: C57 pregnant mice were exposed of alcohol by gavage (5 μl/g, 56% v/v in saline). TUNEL assay showed positively stained cells were significantly higher in alcohol group. Q-PCR result showed an increase of caspase-3 and caspase-8, and a decrease of bcl-2. Western blotting also showed that, alcohol could raise active-caspase-3 and active-caspase-8, reduce caspase-3 caspase-8 and bcl-2. Meanwhile, alcohol exposure also enhanced acetylation of histone H3K9 in embryonic hearts. ChIP assay showed that alcohol significantly increased the acetylation of histone H3K9 in the promoter of caspase-3 and caspase-8, and decrease acetylation of histone H3K9 in the promoter of bcl-2. The second part was in vitro experiment. We found alcohol (200 mM) treatment increased the expression of active-caspase-3, active-caspase-8 and the acetylation of histone H3K9, and also decreased the expression of caspase-3, caspase-8 and bcl-2 in cardiac cells. Then we intervened cardiac cells with curcumin, a HATs enzyme inhibitor. Surprisingly, the up-regulation of active-caspase-3, active-caspase-8 and acetylation of histone H3K9, and the down-regulation of caspase-3, caspase-8 and bcl-2 were reversed. Moreover, annexin-V/PI assay showed that the high apoptosis level that induced by alcohol of cardiac cells were down-regulated after curcumin treating.
Conclusions: This study indicate that histone modification might play an important role in mediating alcohol induced fetal cardiac apoptosis, which is possibly through the up-regulation of acetylation of H3K9 in the promoters of the apoptosis genes. Curcumin could reverse alcohol induced fetal cardiac apoptosis, suggesting curcumin is protective against alcohol abuse during pregnancy.

MP2-2

A novel de novo mutation in the Cardiac Ryanodine Receptor Gene (RyR2) in a patient with concealed Long QT Syndrome

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Introduction: The congenital long QT syndrome (LQTS) is a potentially life threating hereditary channelopathy and fifteen genetic forms have been defined. Here a concealed LQTS patient with a de novo mutation in the *cardiac ryanodine receptor (RyR2)* gene and a known germ line pathogenic mutation in desmoplakin *(DSP)* gene simultaneously is presented.
Case: A nine-year-old male patient, who had syncope episodes with spontaneous resolution in few minutes, admitted with cardiac arrest. After resuscitation electrocardiogram revealed a QT interval of 600 msec, corrected QT (QTc) interval of 620 msec and no T-wave abnormality or dysrhythmia. Echocardiography showed no structural heart disease. At 45th day of hospitalization the ECG showed a QTc of 430 msec. Epinephrine stress test confirmed the diagnosis of concealed LQTS and a de novo heterozygote missense mutation at nucleotide 5170 in exon 37 of *RyR2* gene (GenBank: NM_001035) which replaces glutamic acid with lysine at residue 1724 (c.5170 G > A-p.Glu1724Lys) and a germ line heterozygote missense mutation at nucleotide 88 in exon 1 of *DSP* gene (GenBank: NM_004415) (c.88 G > A (p.Val30Met), were identified on the genetic screening of the patient. (Figure)



Discussion: On genetic screening of our patient, a heterozygote missense mutation, (c.5170 G > A-p.Glu1724Lys), on 37th exon in *RyR2* gene was identified. This mutation was first reported in a Dutch female patient with diagnosis of CPVT (1) and it affects the cytoplasmic loop and protein interaction (1,2). To the best of our knowledge this is the first concealed LQTS patient to have this de novo missense mutation. CPVT patients may be misdiagnosed as concealed LQTS. In both diseases the resting ECG is normal, but induction of bidirectional ventricular tachycardia and/or polymorphic ventricular premature beats during exercise or stress test is typical for CPVT. In our case epinephrine stress test was performed and a positive paradoxical QT response was observed, however any dysrhythmias or ventricular premature beats were not observed.
Conclusions: This is the first concealed LQTS patient to have this de novo missense mutation. In patients with concealed LQTS, provocative tests and genetic screening should be performed and identification of new gene mutations will be helpful for the definitive diagnosis.

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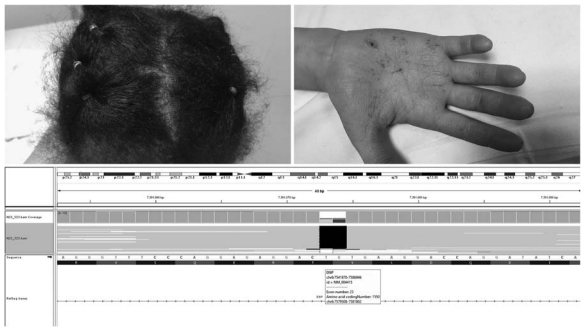
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MP2-3

A novel mutation in desmoplakin (DSP) gene in two siblings with Carvajal Syndrome

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Introduction: Dilated cardiomyopathy is the most common form of cardiomyopathies and it may be a component of cardiocutaneous syndromes. Carvajal syndrome is a cardiocutaneous syndrome characterized with dilated cardiomyopathy, woolly hair and keratoderma (1). Here a new homozygote frameshift mutation in desmoplakin (DSP) gene detected in two female siblings with diagnosis of Carvajal syndrome is presented.
Case: A five-year-old female patient, first child of parents with second-degree consanguinity, admitted with complaints of malaise and abdominal pain. She had findings of overt congestive heart failure, peculiar woolly hair, and palmar keratoderma and was diagnosed as DCM. After being treated with pharmacological agents for congestive heart failure for two years; left ventricular assist device (LVAD) was implanted and on the 501th day of LVAD implantation, the patient had heart transplantation. The sister of the patient also had peculiar woolly hair, palmoplantar keratoderma and DCM. She is being treated with pharmacological agents for congestive heart failure. Genetic screening of both siblings were performed with suspicion of Naxos/Carvajal disease and revealed a new homozygote frameshift mutation, *c.4650_4651delTG (p. V155Efs⋆75),* in *desmoplakin (DSP)* gene. (Figure)



Discussion: Cardiocutaneous syndromes are a group of syndromes associated with DCM and other types of cardiomyopathies. Dilated cardiomyopathy with woolly hair and keratoderma (DCWHK), also known as Carvajal disease, is an autosomal recessive cardiocutaneous syndrome caused by mutations in *DSP* gene which encodes desmoplakin, on chromosome 6p24 (1,2). Mutations in 10 different desmosomal proteins and more than 40 different mutations in desmoplakin leading to distinct cardiac and cutaneous features have been identified so far (3). In our case genetic screening of both siblings revealed a new homozygote frameshift mutation, *c.4650_4651delTG (p. V155Efs⋆75),* in *desmoplakin (DSP)* gene.
Conclusions: Genetic screening is an important tool for early diagnosis and for predicting the presence of disease in asymptomatic family members. As hereditary diseases are an important cause of DCM, early diagnosis of diseases by genetic screening may be life saving for patients. Detection of new mutations and prenatal genetic counseling may help parents in decision of future children.

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MP2-4

A restrictive ventilatory pattern is common in patients with univentricular heart after Fontan palliation and associated with a reduced exercise capacity and a reduced quality of life. A European multicenter retrospective cross-sectional study with 232 patients

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Objectives: The Fontan circulation is highly dependent on ventilation, which improves pulmonary blood flow and cardiac output. A reduced ventilatory function was reported in these patients. This analysis investigates the extent of this impairment and its relation to exercise capacity and quality of life.
Methods: This multicenter retrospective cross-sectional study included 232 patients (140 females, 92 males, 25.6 ± 10.8 years old) with univentricular heart after Fontan palliation (19.8% atrioventricular connection; 20.3% atriopulmonary connection; 59.9% total cavopulmonary connection). Resting spirometry, cardiopulmonary exercise tests, and health-related quality-of-life assessment (SF-36) in those 14 years or older were performed between November 2003 and May 2015.
Results: FEV1 was 74.7 ± 17.8%pred. and FVC 71.1 ± 16.9%pred. In those 58.3% of patients with a FEV1 < 80%pred., all had a normal FEV1/FVC >70% suggestive of a restrictive ventilatory pattern. The reduced FEV1 was associated with a reduced peakVO2 of 67.0 ± 17.6%pred. ($r = 0.43$, $p < 0.0001$), even if analyzed together with possible confounding factors (sex, BMI, age, years after Fontan palliation, total number of interventions, presence of scoliosis, diaphragmatic paralysis, and PLE).
Synergistically to exercise capacity, FEV1 was weakly associated to quality of life in the aspects of physical functioning ($r = 0.25$, $p = 0.008$), bodily pain ($r = 0.22$, $p = 0.02$), and general health ($r = 0.16$, $p = 0.024$).

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FEV1 itself was associated to diaphragmatic paralysis ($p = 0.001$), scoliosis ($p = 0.001$), high total number of interventions ($p = 0.002$), and reduced BMI ($p = 0.01$). No correlation was found with ventricular morphology, timing/type of surgeries, catheter and echo data, and others peri-operative or long-term complications.
Conclusions: The common restrictive ventilatory pattern in patients with a Fontan circulation has a substantial impact on exercise capacity and quality of life. Risk factors are diaphragmatic paralysis, scoliosis, a high total number of interventions, and cachexia.

MP2-5
Number of Thoracotomies Predicts Impairment in Lung Function and Exercise Capacity but not survival in Patients with Congenital Heart Disease
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Objective: Almost all of the patients with congenital heart disease (CHD) require open heart surgery to ensure survival into adulthood. History of previous thoracotomy is associated with respiratory muscle weakness, impairments in chest wall compliance and moderately to severely impaired lung function. This study aimed on the functional outcome of patients with CHD with regard to the number of thoracotomies.
Patients and Methods: In total 1372 adolescents and adults with various CHD (624 female, 32.4 ± 11.5 years) undergone lung function testing and accompanied cardiopulmonary exercise testing in our institution from January 2010 to August 2015.
Results: After adjusting for confounding variables, with every thoracotomy the risk for a restrictive lung pattern increased by 1.8-fold (CI: 1.606-2.050; $p < .001$) and the risk of impaired exercise capacity by 1.2-fold (CI: 1.054-1.346; $p = .005$). There is a strong correlation of forced vital capacity and peak oxygen uptake ($r = .464$, $p < .001$). In 1066 patients surgical treated, thoracotomy during the first year of life was associated with a 1.5-fold increased risk (CI: 1.118–1.984; $p = .007$) impaired lung pattern but not with exercise capacity. During follow-up 21 patients died. Survival increased by 6.6% (CI: 1.035–1.098; $p < .001$) for every percentage increase in peak oxygen uptake (%predicted). Thoracotomies ($p = .522$), cyanosis ($p = .612$) and forced vital capacity ($p = .389$), were not predictive for survival.
Conclusions: Independent for CHD complexity, multiple thoracotomies lead to limitations in lung capacity yielding to further limitations in exercise capacity which is still the strongest predictor for survival in multivariable analysis.

MP2-6
New QT and JT correction methods in right bundle branch block in children
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Introduction: QT interval prolongation on the surface ECG is a known marker of abnormal repolarization and the potential for arhythmogenesis. In patients with right bundle branch block (RBBB), the assessment of ventricular repolarization remains controversial. We set out to compute the best derived QT and JT formula correction factors in children with RBBB.
Methods: we enrolled a cohort of 96 children with RBBB. In a quiet state a digital 12 lead electrocardiogram was recorded and stored. In 9 patients, > 1 ECG at different time intervals were obtained (total of 129 ECG's studied) The QT, JT and RR intervals were measured digitally in lead 2. The QT/RR and JT/RR curves were fitted with 2 regression analysis. Firstly a linear regression for constant α, whereby $QTc = QT + \alpha \times (1\text{-}RR)$, and $JTc = JT + \alpha \times (1\text{-}RR)$ and secondly a natural log-linear regression analysis for constant β whereby $QTc = QT/RR\beta$ and $JTc = JT/RR\beta$. Additionally, linear regression analyses of QTc/RR and JTc/RR for each two formulae were performed as well as QTc/JTc vs QRS duration to obtain slope and R2. A slope and R2 close to zero judged to eliminate the effect of heart rate on QT interval.
Results: mean age 8.4 years, range 0.3-18 years, median 7.0 years, Mean QRS duration was 124 ms SD + 18 ms, median 120 ms, range 90– 174 ms. From linear regression analysis, correction factor for JT was $\alpha = 0.19$ and $\beta = 0.43$ and for QT α was 0.22 and β 0.39. Linear Regression plots for QTc and JTc against RR intervals: QTc linear: slope < 0.005, R2 < 0.01 QTc log: slope < 0.05 R2 < 0.01, JTc linear slope 0.039 R2 > 0.001, JTc log slope -0.03 R2 < 0.001. QRS duration plotted against JTc α R2 0.028 and JTc β R2 0.019; QTc α R2 0.3, QTc β 0.32.
Conclusion: Correction for heart rate was good for both JT and QT new formulae. For QRS duration correction, unsurprisingly, the JT formulae were superior. For pediatric subjects with RBBB, these new JTc and the QTc correction formulae perform well.

MP2-7
Usefulness of Andrastents for dilation of different vessels
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Aim: To present our experience with application of new cobalt - chromium stents (namely Andrastents XL/XXL).
Methods: There were 91 patients treated with 93 Andrastents - 53 (aged 8-65 years) with native CoA or ReCoA, 16 (aged 6-64 y) with left or right pulmonary stenosis (PS) closely to the bifurcation (native or postsurgical). In 19 pts (aged 11-40 y) the procedure was presenting before Melody valve implantation (in calcified pulmonary homograft or native RVOT). In 3 patients Andrastents were implanted in different places to dilate stenosis of: superior vena cava (in 7.5 y o child), Fontan tunnel (in 17 y old boy), and PFO (interatrial septum in complex heart defect in 19 old yo boy). Mean follow-up was 3.4 (0.2-5.4) years.
Results: All procedures were finished successfully in all but two patients without any complications with good clinical improvement. Two migration of stents occurred – one in RVOT and another in LPA (without clinical consequences). In all cases successful dilation of stenosed place with significant gradient reduction occurred. In 2 cases of native CoA (23 and 34 y old man) in early follow-up (6 and 8 months after the procedure) in angio CT small aneurysm formations was observed. Both patients were treated successfully with covered stents. In follow-up no fracture of the stent nor any others complications were observed.
Conclusions: Implantation of Andrastents XL and XXL is a good therapeutical option for the treatment of stenosed great vessels.

MP2-8
Comparison of efficacy and safety of 6 different types of nitynol wire mesh occluders in 1321 patients with atrial septal defect (ASD) closure
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Objectives: To compare efficacy and safety of 6 different nitynol wire mesh occluders in ASD closure: Amplatzer ASO (St Jude

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Med), Cardi-O-Fix (Starway Comp- without CE mark from 2012), Figulla (Occlutech GmbH), Cera, HeartR (both produced by LifTech Comp) and Hyperion (Comed Comp).
Methods: Between 1997 and 2015 1321 patients (pts) in our center had performed percutaneous closure of ASD using standard transvenous technique.
Results: In 1281 pts (97%) the procedure was successful (in 40 pts - mainly with ASO occluders were withdrawn). There were 1077 single ASD and 204 double/multiples ASD. Some of the clinical and procedural data are shown in the table 1. There were 8 early implant embolizations - 7 ASO, 1 Figulla device – mainly in the early years of its usage. ASD diameter and implant sizes, fluoroscopy time were similar in all groups. In the ASO group the age and weight of the pts were lower and the follow-up longer. No serious complications (such as wall erosion, fracture of the device, or thrombus formation) were observed in any pt. In late observations 1 pt developed endocarditis due to incomplete endothelialization and in 2 others complete AV block occurred, with the need of pacemaker application (all treated with ASO).
Conclusions: All types mentioned above nitynol wire mesh occluders for ASD closure have the same effectiveness what cann promote some of them as cheaper alternative. The advantage of the smallest sheath makes Amplatzers the optimal device in the closure of ASD in small children.

Table 1. Some clinical and procedural data.

Implant type	No of pts	Age (years) mean	Weight (kg) mean	ASD in TEE (mm) mean	Implant size (mm) mean	Fluorosc. (min) mean
Amplatzer	**1013**	**21** (0.5-77)	**44.4** (6-119)	**12.8** (3-34)	**18.1** (4-40)	**5.0** (1-39)
Figulla	**87**	**35** (3.4-79)	**68.5** (19-121)	**16.7** (6-33)	**22.9** (12-39)	**4.3** (1-13)
Cardi-O-Fix	**62**	**27.3** (2.5-72)	**55.5** (14-103)	**13.6** (7-22)	**19** (9-30)	**3.6** (0.8-11.4)
Cera/Heart R	**27/31**	**34.6** (5-74)	**59.2** (18-107)	**14** (5-26)	**18.3** (10-30)	**3.6** (1-9.3)
Hyperion	**61**	**28.5** (5.2-77)	**57.2** (17-117)	**13.2** (4-26)	**17.9** (9-34)	**3.2** (0.5-11.5)

MP2-9
Lung Function in Patients With Congenital Heart Disease. Prevalence, Severity and Correlation to Diagnosis

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Objectives: Since restrictive lung function is an independent predictor of mortality in congenital heart disease (CHD), investigation of lung function in patients with CHD was performed; including description of prevalence, type of lung disease and severity and its correlation with the underlying diagnosis.
Methods: 536 patients with CHD aged 6 to 69 years (mean 28.2 ± 13.4 years) underwent spirometry between 2012 and 2015. The ratio of forced expiratory volume in the first second to forced vital capacity (FEV1/VC Ratio) and vital capacity were measured. Cut-off: restrictive lung function: VC < 80% of predicted, obstructive lung function FEV1/VC < 0.8. Patients were divided in groups by diagnosis (see fig. 1).
Results: Patients with normal lung function: 32.5%, restrictive lung function: 29.3%, obstructive lung function: 23.3%, mixed-type impairment: 14.9%. Highest percentage of restrictive lung defects was found in patients with complex cyanotic heart defects, univentricular anatomy, and Fallot tetralogy. Least patients with obstructive lung defect were found in patients with TOF and TGA after atrial switch operation (13.8 and 14.6%), whereas all other groups contain more than 22% of patients with obstructive lung disease. A decreased vital capacity correlates significantly with the number of thoracotomies ($r = -0.41$; $P = 0.0001$). There is a significant difference in lung impairment releated to underlying diagnosis $P < 0.0001$.
Conclusion: A significant amount of our patient population demonstrated impaired lung function. Restrictive lung function, detected in more than 40% patients with complex cyanotic anatomy, univentricular hearts and Fallot tetralogy may significantly affect the long term outcome. Since this is a known and independent risk factor for mortality (Alonso-Gonzalez, et al. 2013), it is recommendable to routinely perform spirometry especially on these patients to determine individual risk by impairment or decline in lung function.

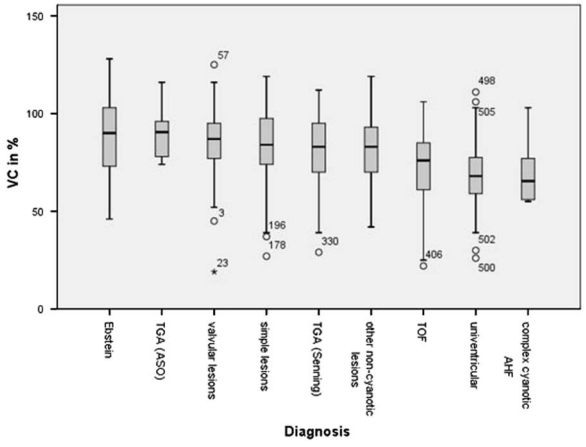


Figure 1. Median and quartiles of VC% in different CHD. TGA – transposition of great arteries, ASO arterial switch operation, TOF – Fallot tetralogy.

MP2-10
Transcatheter Closure of Perimembranous Ventricular Septal Defects with Amplatzer Ductal Occluders

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Inroduction: This study reports on our experience with transcatheter closure of perimembraneous ventricular septal defects (pm-VSDs) with Amplatzer duct occluder I (ADO I) and Amplatzer duct occluder II (ADO II)
Methods: Transcatheter device implantation with ADO I and ADO II was attempted in 17 patients with pm-VSD between August 2014 to December 2015. We usually decided the appropriate type and size of device according to the measurements by left ventricular angiograms. We choose 1-2 mm larger device than the defect size in our first cases. Recently we prefered 2-4 mm larger device due to prevent the residuel leakage or device embolism.
Results: Patient mean age was 10.59 ± 3.55 years and mean weight was 37.12 ± 16.36. The mean defect size was 5.17 ± 1.66 mm , mean Qp/Qs was 1.88 ± 0.5, mean PVR/SVR ratio was 0.042 ± 0.028 and mean fluoroscopy time was 25.11 ± 7.97 minutes. ADO I was used in 13 cases and ADO II used in remaining 4 patients. The Amplatzer duct occluders were succesfully implanted in 16 of 17 patients. In one patient, ADO I

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suddenly embolised into the left pulmonary artery soon after its release. This device was retrieved and the patient underwent surgical closure of the ventricular septal defect succesfully. One of 4 patients underwent pmVSD closure with ADO-II had residuel leakage and one of them had transient left bundle branch block after the procedure. In addition one of patient had residuel leakage after ADO I deployment. No atrioventricular block (AVB) was determined during follow-up.

Conclusions: We prefer the Amplatzer duct occluders in percutenous closing to pm-VSD among several devices. Amplatzer duct occluder I has an advantage due to does not have a proximal disc and thus does not squeeze the AV bundles. Although ADO II has a two disc, its low-profile and flexibility provides some advantages compared to other devices. Residuel leakage and one case of device embolism were seen in our first cases. Therefore we started to choose 2-4 mm larger device if adaquete defect rims were present and no residual leakage, embolism or AVB were detected . Duct occluders provide an effective and safe treatment option in selected patients with pm-VSD.

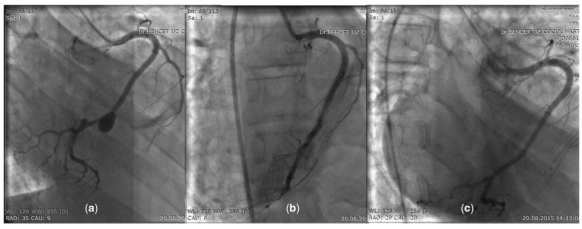
MP2-11

Pericardium-covered stent implantation in a complicated giant coronary artery aneurysm in an adolescent boy

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Introduction: We report an adolescent in whom pericardium-covered stent was successfully implanted in a giant coronary aneurysm which is ruptured and caused an intrathoracic hematoma.

Case: A 16-year-old boy Syrian refugee, admitted to our emergency room because of sudden onset of chest pain and respiratory distress 8 months ago. Echocardiography was demonstrated a mirror-image dextrocardia, massive pericardial effusion and compression of right heart chambers. After the successful pericardiocentesis, control echocardiography showed a cystic formation contiguous with the right atrioventricular groove that was not previously noted. The coronary CT was demonstrate a giant aneurysm originated from distal part of the right coronary artery (RCA). Diagnostic coronary angiography confirmed the presence of giant aneurysm 20 mm in diameter located at the distal part of RCA (fig. 1a). However, the family and the patient refused therapeutic options and they left the hospital without permission. The patient re-admitted to our emergency department with the complaint of chest pain. Electrocardiogram demonstrated ST-T wave changes in precordial leads. Transthoracic echocardiography showed a mild dilatation of left heart chambers with mild systolic dysfunction. Echocardiography also demonstrated giant coronary aneurysm previously detected and huge thoracic mass compressing the heart and left lung. A multislice CT showed a huge mediastinal hematoma arising from the pericardial space with bulging into the distal lobe of left lung. A giant aneurysm (24 × 32 mm) and a small one (6 × 4 mm) at the distal part of a right coronary artery were also detected in multi-slice CT. Blood leakage from coronary aneurysm was considered as the source of pericardial hematoma. Therefore, we decided to use covered stent for the exclusion of giant aneurysm. A 3.0 mm × 27 mm pericardium-covered stent was implanted to exclude both aneurysms (fig. 1b). However, persistence of stenosis at the proximal site was detected after implantation. Thereafter, we implanted an another pericardium-covered stent at the proximal site so that which resulted that stents overlapped each other. The final angiogram showed both improvement of blood flow and the disappearance of stenosis (Fig. 1c). We did not encounter any complications during the procedure. Elective surgery was planned, as there was no regression of hematoma previously occurred.



MP2-12

Percutaneous Pulmonary Valve Implantation in Patient with Native Right Ventricular Outflow Tract

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Background: Right Ventricular Outflow Tract (RVOT) dysfunction due to transannular patch or valvulotomy remains as a risk factor that increase the mortality and morbidity on late period. Surgical pulmonary valve implantation is the traditional treatment strategy. Recurrent surgeries, may leads complications of cardiopulmonary bypass, sternotomy etc. Percutaneous pulmonary valve implantation (PPVI) is an alternative, in order to prevent the patient from cumulative risk of multiple surgeries. However, anatomical complexity and differences and very large sizes for balloon expandable valves of RVOT, makes the PPVI technically more difficult. In this paper we present PPVI with Edwards Sapien XT valve in patients with native RVOT.

Patients and Methods: Fourteen (14) patients were taken to catheterization laboratory for PPVI procedure. In patients with transannular patch who has no stenosis, stent implantation was done at first session. Least two months after stent implantation, PPVI was performed. In one patient with stenosis and free regurgitation PPVI procedure was performed at the same session with stenting.

Results: Mean age was 13 4.5 (6-26) years and mean weight was 28 7.9 (17-75) kg.

In 92% of (13/14) patients PPVI was performed successfully. Ten (10) patients had free pulmonary regurgitation,3 patients had severe pulmonary stenosis and moderate pulmonary valve insufficiency. Andra XXL stent were used for prestenting in 11 cases. CP stent were implanted in two cases who had stenosis. Mean maximal diameter of the RVOT's was 23 6.8 (12-26.8) mm. Edwards Sapien XT valve were used in all patients. Size of the implanted valves were 20 mm in two cases, 23 mm in two cases, 26 mm in 3 cases and 29 mm valve was used in 7 cases. In one patient, stent embolization occurred during prestenting. Stent exteriorized surgically without any complication and surgical valve inserted. Mild paravalvular insufficiency remained in one patient.

Conclusion: PPVI is an alternative in the treatment of RVOT dysfunction in patients with native RVOT. Presenting is essential for valve implantation in order to create a landing zone. Using of 26 and 29 mm sizes of Edwards Sapien XT valve extend the possibility of percutaneous valve implantation in larger RVOT's.

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MP2-13

Extended application of the Amplatzer Vascular Plug II for elongated arterial duct occlusion

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Introduction: Although used very successfully in the majority of attempted transcatheter arterial duct occlusions, the St. Jude ADO 1 is less suitable for elongated or complex arterial ducts as the aortic retention disc must distort to allow safe capture of the narrowest diameter. We aim to describe the use of the St. Jude, Amplatz Vascular Plug (AVP) II for patients with significant arterial ducts in 2 UK centres.
Methods: From September 2013 to October 2015 a total of 35 patients (24 female) underwent attempted duct occlusion using the AVP II. The median age was 1.3 years (range 0.46-14.5) and median weight 9.25 kg (range 4-54). 17 patients were symptomatic with tachypnoea ± failure to thrive and in 21 patients the left ventricular end diastolic dimension z score was over 2. Using angiography, the median narrowest diameter was 3 mm (range 1.2-7.2) and median length was 12.5 mm (range 6.2-21). Krichenko duct shapes were A (n = 1), C (n = 3), D (n = 8) and E (n = 23). Fourteen implantations were via an antegrade approach and the remainder retrograde.
Results: Sizes of AVP II devices (diameter times unconstrained length) employed were 6 × 6 (n = 11), 8 × 7 (n = 21) and 10 × 8 (n = 3). There were 2 complications, in one patient the device migrated to the left pulmonary artery and was successfully retrieved before successful ADO 1 implantation. In a further patient, the AVP II appeared unsatisfactory before release and was retrieved before an ADO 1 was implanted which subsequently migrated requiring surgical retrieval. In all other patients implantation was successful with complete duct occlusion and no disturbances to flow in the left pulmonary artery and aorta.
Conclusions: The morphology of the arterial duct must be carefully considered before selecting a suitable device. In elongated and complex ducts, the AVP II performs well and should be considered as a first line device.

MP2-14

First Evidence in Humans of Neointimal Regeneration after Stenting of Native Right Outflow Tract: an intracardiac Ultrasound Study

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Background: Percutaneous pulmonary valve implantation (PPVI) is a challange in pts treated with outflow patch. After an adeguate selection by echocardiography, cardiac MRI and CT, pts with favorable anatomy are selected for this procedure but a PPVI is considered feasible if a stable percutaneous conduit is built. Aim of our study is to demonstrate neointimal proliferation growth by ICUS in implanted stents in the right native outfow tract. At date, only animal models or authoptic samples have been used to report neointimal stent proliferation in aortic position and IVUS studies have been performed only in adult patients with coronary stents.
Methods: We prospectively analyzed 7 children, previously surgically corrected for tetralogy of Fallot by outflow patch, with severe pulmonary regurgitation defining the indication to PPVI. All patients underwent a 'two step' procedure for PPVI. At first stage an hybrid stent was implanted in the outflow patch. At the same time, an ICUS study was performed using an Ultrasound Catheter. After 2-8 months a second ICUS study was performed to demonstrate presence and entity of neointimal stent proliferation, preceeding covered stenting and PPVI. Neointimal stent proliferation quantification was performed by two independent reviewers, blinded to clinical data. Spearman rho analysis was performed to analyze interobserver variability.
Results: All 7 patients were boys (age 12.6 ± 2.7years, weight 48 ± 15Kg). At baseline no neointimal thickness stent was visible in any of the patients. At second procedure (4.5 ± 2.3 months after stent implant) the ICUS study showed presence of endothelization in all children, with a mean neointimal proliferation thickness of 1.3 ± 0.2mm (range 1.0-1.6mm), with low interobserver variability (Spearman's rho = 0.81; p = 0.027). Interestingly, endothelization thickness was mildly positively correlated to time from implant (r = 0.53). At second procedure, all patients underwent successful PPVI.
Conclusions: Our preliminary study is the first evidence in literature of a stent neointimal regeneration in humans with native right outflow patch. Our study demonstrated that neointimal proliferation occurs in all patients in which a stent is implanted and that this phenomenon can be accurately assessed by ICUS. Evidence of neointimal proliferation supports the proposed approach of a 'two step' PPVI procedure.

MP2-15

ECMO after cardiac surgery in infants: does early cardiac catheterization improve outcome?

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Objectives: To report our experience with cardiac catheterization performed during extracorporeal membrane oxygenation (ECMO) in children who underwent cardiac surgery in our Institution.
Methods: We retrospectively reviewed our ECMO registry (February 2011-October 2015) and identified patients who underwent cardiac catheterization while on ECMO. All patients had complex congenital heart disease and had undergone cardiac reparative or palliative surgery in the past 48 hours. Indication for cardiac catheterization was hemodynamic and anatomical assessment when weaning from ECMO was not possible after 48 hours and surgical complication was suspected. Primary outcome was considered weaning from ECMO. We sought to observe if cardiac catheterization influenced primary outcome.
Results: Fifty-three consecutive patients (mean age 50 days) received artero-venous ECMO support for cardiac arrest or failure to wean from cardio-pulmonary bypass. Fifteen cardiac catheterizations were performed on 13 patients (24.5%).
Catheterization was diagnostic in 12 and interventional in 3 cases. In 8/13 (61%) catheterization modified therapeutic approach. A structural anomaly requiring repeat surgery was detected in 5 cases. In details, catheterization identified a left ventricular outflow obstruction after arterial switch, a pressure gradient through a Damus-Kaye anastomosis, too tight a pulmonary branch banding after hybrid stage I palliation and two cases of right ventricular outflow obstruction after Fallot repair. Interventional catheterization was performed in 2 patients with narrowing of the right pulmonary artery after comprehensive Norwood stage I-II, readily treated with successful stenting. Also, coil embolization of systemic-to-pulmonary collateral was performed during catheterization in one patient who was then scheduled for repeat surgery the day after, based on the evidence of elevated right ventricular

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pressure. In one case catheterization diagnosed pulmonary arterial hypertension and epoprostenol was started.
Weaning from ECMO support was achieved in 9/13 patients (70%), a mean of 3.2 days after cardiac catheterization/surgery. Conversely, 18/40 patients (45%) who did not undergo cardiac catheterization were weaned from ECMO. The Odds Ratio for the probability of weaning from ECMO was higher after cardiac catheterization (OR 2.750).
Conclusions: Cardiac catheterization during ECMO provides useful information that can actively modify treatment approach. Weaning from ECMO is positively influenced by early cardiac catheterization.

MP2-16
Infective endocarditis after PPVI with the Melody® valve Is residual RVOT gradient important?
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Introduction: Melody® valve used for percutaneous pulmonary valve implantation (PPVI) is susceptible to infective endocarditis (IE). Multiple risk factors have been postulated. We analyzed the presence of residual RVOT gradient, age and sex as risk factors for developing IE in Melody.
Patients and methods: Single center analysis of Melody® valve implanted between 2006 and 2014. Analysis of valve function by echocardiography. Screening of patients records for IE and other events.
Results: 133 Melody® valves were implanted in 132 patients with mean age 19,9 years (range 3,9 to 81,6y), and a total of 302,2 patient years of follow-up (mean follow-up 2.3y; maximum follow-up 7.6y). IE occurred in 10 patients (7.5%). after a mean period of 2.0 years (range 0.7 to 4.0y). Freedom from IE was 78.93% after 5 years. Male patients with a Melody® valve are clearly more prone do development of IE ($p = 0\,0451$) and young age (between 10-20 years) gives an increased tendency to develop IE, however not significant (hazard ratio < 1; $p = 0.2323$). Residual gradient over the RVOT ($p = 0.1085$) or rapid increase of RVOT velocity ($p = 0.0811$) appear not to be independent risk factors or predictors for IE.
Conclusions: With current data IE in Melody valve can not be predicted by residual gradient in the RVOT. Male adolescents are clearly more prone to develop IE in Melody. Prevention and instructions remain important.

MP2-17
Multiple different metal stents in the RVOT: is electrolysis a threat?
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Introduction: Different metals in close physical contact lead to electrolysis and corrosion; this may downsize the strength and durability of the stents. Lack of data precluded pre-stenting in several trials by FDA.
Methods: Stents used in the analysis: Cheatham-Platinum (Pl-Ir-Au), Andrastent (Co-Cr), Optimus (Co-Cr), Sapien (SS316L) SapienXt (Co-Cr). All tests performed in saline Plasmalyte AKE0324 at 37 °C. At the end of each test visual and stereomicroscopic investigation was performed to type the corrosion as severity. 1/galvanic corrosion test: open circuit corrosion potential (OCP) was determined against an Ag/AgCl reference electrode (exposed surface 1.5 cm^2). The samples are coupled to each other. The galvanic zero resistance amperometry current and the mixed potential are measured for 1 hour after contact, 1, 2 and 4 weeks. 2/Exposition corrosion test: couples Andrastent (Co-Cr)+ CP stent (Pt-Ir-Au) and Optimus stent (Co-Cr)+ CP stent (Pt-Ir-Au) were tested for 3 months. Round test samples with both stents folded in each other resulting in a direct galvanic connection. Visual and stereo microscopic investigation; weight decrease analysis (uniform corrosion rate). 3/Electrochemical cyclic polarization tests : measurement of the open circuit corrosion potential after an equilibration period of 16 hours; repeated measures for 3 months. A cyclic polarization gives a polarization scan : measured current in relation to the applied anodic and cathodic overpotential.
Results: 1/couples CP-Andrastent, Andra-Sapien and Andra-SapienXT have lowest galvanic corrosion rates (0.000001 mm/year), resulting in material loss of 10 µg/year. The couples CP-Sapien and CP-SapienXT have a higher corrosion rate (0.000003 mm/year) resulting in material losses of respectively 17 and 24 µg/year. 2/No signs of corrosion could be observed on both materials of the couple Andrastent + CP stent. 3/The calculated corrosion rate of expanded and non-expanded Andrastent based on Tafel slope analysis was estimated as ~0.00002 mm/year
Conclusion: The corrosion rate and material loss of all tested materials is extremely low. No mechanical integrity problems are expected: after 100 years only 0.3% of the initial diameter of the wires of a stent will be lost as a consequence of corrosion.

MP2-18
Supportive medical therapy for inter-stage of the Gießen Hybrid procedure
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Objectives: To report on our 5-year single-center experience with supportive medical therapy (SMT) for the Gießen hybrid procedure for complex congenital heart disease (CCHD).
Methods: BLS (Bisoprolol, Lisinopril, Spironolactone) is the cornerstone of the Gießen SMT, which was developed by a pathophysiological driven concept addressing the neuro-humoral and cardiovascular effects of the specific flow pattern of postnatal parallel circulation i.e. I) blocking the inadequately activated neuro-humoral axis and lowering heart rate II) balancing systemic and pulmonary flow III) enhancing anti-fibrotic activity and promote cardiac remodeling. The dosage of B and L was in most patients 0.05-0.1 mg/kg S 1-2 mg/kg; all drugs given once daily. Diuretics were avoided in order not to further activate the neuro-humoral axis. Data was retro- and prospectively analysed (study period 12/2010 to 12/2015).
Results: 52 newborns underwent the "Gießen hybrid" procedure with uneventful surgical bilateral pulmonary artery banding (bPAB) and percutaneous arterial duct stenting for primary treatment of CCHD (hypoplastic left heart syndrome/complex (HLHS/HLHC), $n = 30/16$; others, $n = 6$) at a median age of 6 days (range 1 – 50) with a median weight of 3.0 kg (range 1.9–4.4; $n = 9 \leq 2500$ g) and a median Aristotle score of 17.0 (range 14.5–21.5). Prior to bPAB, 16% of patients received bisoprolol, 4% lisinopril, 12% spironolactone, 24% furosemide, 12% hydrochlorothiazide. At discharge, 90% of patients received bisoprolol, 73% lisinopril, 78% spironolactone, 18% hydrochlorothiazide. The mean resting heart rate was reduced from 138/min at admission (range: 112–172/min;

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n = 52) to 123/min at discharge (range: 99–139/min; n = 51) to 110/min before stage II/biventricular repair/heart transplantation (range: 90–140/min; n = 37). No side effects were observed; the parental compliance for treatment was excellent.
Conclusion: SMT for the Gießen Hybrid procedure appears to effectively reduce full-body and myocardial oxygen consumption. In single cases reduction of the diastolic left-right flow across the stented duct could be monitored. Further prospective studies are warranted whether the pathophysiological concept sufficiently addresses the specific hemodynamics of the parallel circulation and in which agonistic and antagonistic neuro-endocrine parameters are affected.

MP2-19

Transcatheter pulmonary valve implantation in small right ventricle to pulmonary arteries conduits

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Objectives: Evaluate the feasibility and the outcome of Melody valve (Medtronic Inc, Minneapolis, USA) implantation in small right ventricle outflow conduits.
The current guidelines recommend a minimal conduit diameter of 16 mm and no dilatation to a diameter greater than 110% of the original implant size for deployment of a percutaneous pulmonary valve.
Methods: Seven consecutive patients (May 2008-October 2015) from 5 institutions underwent transcatheter pulmonary valve implantation in less than 16 mm conduit diameter (n = 7). Three additional patients with a conduit diameter of 16 mm (n = 2) or 17 mm (n = 1) underwent a 22mm Melody valve implantation. The median age and weight of the 10 patients was 10.85 (7.7 to 15) years and 41.5 (19 to 60) kg, respectively.
Results: The median diameter of the conduit at the time of the procedure was 12 (10 to 14) mm and the median right systolic right ventricular pressure was 62 (40 to 94) mmHg. The procedure was successful in all cases with a median diameter of the implanted pulmonary valve of 22 (18 to 22) mm. A confined conduit rupture occurred in 4 patients and was treated with covered stent implantation priori to melody valve implantation. The post-implantation median systolic right ventricular pressure was 30 (29 to 50) mmHg. One patient who had implantation of an 18 mm diameter Melody valve experienced moderate post implantation systolic right ventricular hypertension (50 mmHg). After a median follow-up of 13.7 (2.9 to 85.2) months, all the patients are asymptomatic and no further intervention was required.
Conclusions: Transcatheter pulmonary valve implantation is feasible in small conduits, with a high rate of ruptures that remain confined because of the surrounded fibrosis. In this setting, one should always try to implant the largest available pulmonary valve.

MP2-20

Three dimensional rotational angiography imaging in diagnosis and treatment of complex congenital heart diseases. A single-centre experience

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Introduction: Three dimensional rotational angiography (3DRA) is a relatively recent but rising imaging technique used in paediatric and adult catheterization laboratories. Fusion of 3-dimensional (3D) image data with fluoroscopy can potentially overcome limitations of 2-dimensional (2D) angiography for visualizing complex cardiovascular structures and facilitating accurate diagnoses as well as drive interventional procedures. 3D images in real time can be useful both for diagnostic purposes and as a roadmap to guide interventions, especially in complex congenital heart diseases (CHD). We want to evaluate the impact of 3DRA in CHD catheterization management.
Methods: A database of all patients catheterized between March and December 2015 at our Hospital, were examined. Among all treated patients, 59 cases of complex CHD were approached with 3DRA. About considered population mean age and main weight were respectively 18.7 ± 12 yrs (range 2 months - 61 years of age) and 47.2 ± 23.8 Kg (range 2.5-90 Kg), including 28/59 paediatric patients (47.4%). Procedure types and trend were analysed.
Results: 3DRA was used respectively in 11 diagnostic procedure and 48 interventional catheterization. Diagnostic procedures consisted in n = 3 pulmonary atresia with VSD and MAPCAs, n = 5 TOF after surgical repair, n = 1 discontinuous pulmonary artery, n = 1 single ventricle after cavo-pulmonary connection, n = 1 aortic coarctation after stenting. Interventional catheterizations involved n = 18 percutaneous pulmonary valve implantations, n = 18 percutaneous treatments of aortic coarctation (15 stent implantation, 3 stent re-dilatation), n = 12 percutaneous treatment of pulmonary branch stenosis (10 stent implantation, 2 balloon angioplasty). In total investigations median dose-area product (DAP) was 8687.53 μGym2 (range 146.32-48 397 μGym2), median fluoroscopy time was 30.6 min (range 3.29-186.22 min) and median contrast dose was 355.33 ml (range 50-999 ml). Separately in diagnostic investigations and in interventional procedures, DAP, fluoroscopy time and contrast dose were respectively: 2220.6 vs 9791.6 μGym2, 11.37 vs 35.2 min, 156 vs 403.9 ml.
Conclusions: 3DRA used in diagnostic procedures and interventional catheterizations, especially for complex CHD, provides significant additional information as compared to bi-planed angiography, without need to do multiple injections and with a real reduction of X-ray exposure.

MP3-1

Assessment of cardiac angle in fetuses to predict cases of tetralogy of Fallot and common arterial trunk

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Introduction: Only one third of cases of Tetralogy of Fallot (TOF) and Common Arterial Trunk (CAT) are diagnosed prenatally. Fetal cardiac axis has been shown to be rotated towards the left in these anomalies. The objective of our study was to evaluate the performance of fetal cardiac angle as a screening tool for the above cardiac anomalies. In addition, we aimed to compare its utility to that of maternal diabetes and smoking which have recently been reported in a review as risk factors for TOF and CAT.
Methods: The fetal cardiac angle was retrospectively measured using a previously published method on archived images in cases of the above-mentioned cardiac anomalies (N = 63). These were compared to a control group with a documented normal postnatal outcome (n = 151). The presence of maternal diabetes and smoking were ascertained from medical records. The karyotype

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and pregnancy outcome were ascertained in all cases of TOF and CAT. Logistic regression analysis was used to assess the performance of the fetal cardiac angle and maternal history of smoking and diabetes in the prediction of TOF and CAT. The Area under the Receiver Operator Characteristic Curve (AUROC) was calculated to assess the performance of the screening model.
Results: The 63 cardiac anomalies included 58 cases of Tetralogy of Fallot and its variants and five cases of common arterial trunk. The mean fetal cardiac angle was larger in fetuses with TOF and CAT compared to controls (63.5° vs 44.1°, $p < 0.0001$). The prevalence of diabetes (6.3% vs 11.9%, chi-square $p = 0.2$) and smoking (10% vs 6.9%, chi-square $p = 0.4$) was not different between TOF and CAT and controls. In a multivariate logistic regression model, the only predictor of these lesions was the fetal cardiac angle (logit (TOF and CAT) = -11.05+ 0.198 * cardiac angle, $R2 = 0.6$, $p < 0.0001$). The AUROC was 0.899 (95% CI, 0.846-0.951).
Conclusions: The fetal cardiac angle is an effective screening tool for identification of TOF and CAT.

MP3-2
Persistent Nuchal Translucency And The Fetal Heart

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Background: It has been reported that 6% of nuchal translucency (NT) > 99th centile are persistent into the second trimester. The objective is to describe the outcome of a cohort of cases with persistently elevated nuchal translucency (NT) in the second trimester who underwent fetal echocardiography.
Methods: A retrospective review of cardiac and genetic findings in cases identified with a persistently elevated NT between 2003-2014.
Results: Fifteen cases were identified with a NT >99th centile in the first trimester and persistently elevated to mid-gestation. A third trimester scan was performed in 10 of these cases of whom 8 had persistently elevated NT. In the second trimester, pleural effusions were seen in 6 cases and hydronephrosis in 4. Polyhydramnios was present in 9 cases in the third trimester. Congenital heart disease was diagnosed prenatally in 10/15 cases: pulmonary stenosis (n = 5), valve dysplasia (n = 2), hypertrophic cardiomyopathy (n = 6), aortic stenosis (n = 3), ventricular septal defect (n = 2), partial atrioventricular septal defect (n = 1), aberrant right subclavian artery (n = 1). Absence of the ductus venosus was evident in four.
Ventricular hypertrophy, subaortic stenosis and pulmonary stenosis were not present during the first trimester, but manifested in the second or third trimester or in the postnatal period. (Table 1) On the basis of intention to treat 11/12 (92%) survived to delivery, 9/12 (75%) survived to 28 days and 4/12 (33%) through one year. There was one further death at 14 months. Pulmonary valvotomy was required in three cases and septal myomectomy was performed in two cases.
Eleven out of fifteen cases were genetically confirmed to have Noonan syndrome, one had a normal genetic result and results were not available in three. Mutations identified were PTPN11 (n = 6), RAF1 (n = 2), RIT1 (n = 1) and SOS1 (n = 2).
Conclusions:

1. All fetuses with persistently raised NT had evidence of congenital heart disease at birth.
2. There is a high prevalence of Noonan syndrome in cases of persistently elevated NT in the second and third trimester.
3. The outcome in these cases is extremely guarded.

Table 1. Prenatal and postnatal ultrasound findings.

	1St Trimester	2nd Trimester	3rd Trimester	Infancy
Elevated nuchal translucency	15/15	15/15	8/10	-
Pleural effusion	-	6/15	4/10	-
Hydronephrosis	-	4/15	1/10	-
Polyhydramnios	-	-	9/12	-
Valvar stenosis	0/7	3/14	5/8	6/9
Ventricular Hypertrophy	0/7	1/14	3/8	8/9

MP3-3
Changing spectrum and outcome of 1281 congenital heart disease diagnosed in utero in 21 years of activity

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Introduction: Congenital heart disease (CHD) are the most common malformations pre and postnatally. Over the years, fetal echocardiography has become a widely practiced technique, and the proportion of prenatally detected cases is increasing, with more-and-more mild CHD detected.
Objective: Assess the changes in the spectrum and the outcome of prenatally detected congenital heart disease in our tertiary care center in 21 years of activity (1995-2015).
Methods and Results: We detected 1281 congenital heart diseases: 25% (315/1281) were associated with extracardiac and/or chromosomal anomalies and 75% (966/1281) were isolated. Termination of pregnancy was chosen in 48% (151/315) for associated anomalies and 19% (183/966) for isolated ($p < 0.001$). Of these, more than one half (96/183) occurred for hypoplasic left heart cases. The general survival rate is 72%, it is significantly lower for the group of associated heart diseases (75/164 vs 583/783; 46% vs 75%, $p < 0.001$).
Since 1995 to 2005 we diagnosed 678 CHD, the remaining 603 were detected since 2006.
Over 21 years we noticed a significative reduction of the multimalformated foetuses, complex CHD and of the hypoplasic left heart cases and an higher number of aortic arch anomalies and milder anomalies detected, such as ventricular septal defect and pulmonary valve stenosis (203/678 vs 120/603; 30% vs 20%). During the last ten years of activity the survival rate resulted has significantly increased (248/451 vs 435/519; 55% to 84%, $p < 0.05$), the termination rate has significantly decreased (227/678 vs 84/603; 33% to 14%, $p < 0.001$) and the number of neonatal deaths has significantly reduced (176/451 vs 52/519; 39% to 10%, $p < 0.001$).
Conclusion: Over 21 years the spectrum of fetal congenital heart disease has changed and their outcome is significantly improved. The survival and the voluntary termination of the prenatally detected congenital heart disease are strongly influenced by their severity and by the associated extracardiac and/or chromosomal anomalies.

MP3-4
The Impact Of Training And Protocols On The Increased Detection Rate Of Cardiac Anomalies In The Fetus: The Welsh Experience

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Background: Congenital heart disease is a leading cause of congenital-defect related death in childhood with improved antenatal detection of cardiac abnormalities, especially those of the

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outflow tracts, being key in improving outcomes. Due to wide acceptance that additional views of the heart improve diagnostic accuracy, from 2001 various training programmes were undertaken in Wales culminating in the mandatory inclusion of outflow tracts as part of the fetal anomaly scan in 2010. This study was undertaken to assess detection rates of outflow tract anomalies in each period, and the current status of training requirements.
Method: Retrospective analysis of cases in South Wales from 2001–2013 was undertaken via Departmental (n = 2958) and national (CARIS - n = 5420) databases. A training needs analysis questionnaire was undertaken in 2015 to assess current sonographer fetal echo training requirements.
Results: During 2001-2009 the mean number of outflow tract anomalies detected per year was 14.4 whereas during the period 2010-2013 it was 21.2 ($p = 0.008$). The mean referrals per year for these two periods was 196.2 and 208 respectively ($p = 0.65$).
The rate of antenatal detection of outflow tract abnormalities during these periods also increased (47% vs 70%, $p = 0.0005$). Detection rates peaked in 2011 reaching 80.6% before dipping to 70% and 66% in 2012 and 2013 respectively. 60% (n = 41) of sonographers described themselves as very confident at the examination of outflow tracts, however, of these, 77% (n = 30) still professed a need for further training updates.
Conclusion: The detection rate of outflow tract anomalies throughout South Wales has improved substantially. A major increase is seen after the initial training program, and continues with the All Wales training programme and subsequent mandatory implementation of outflow tract examination policy. It has, however, shown a decreasing trend since the cessation of training, indicating that successful screening policies must be combined with continuing structured training programs.

MP3-5
Coarctation Long-term Follow-up and Quality of Life: Predictive Value of Clinical Variables
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Objective: Long-term sequelae and events after coarctation repair are well described. However, the predictive value of variables from clinical follow-up investigation for late events has rarely investigated.
Methods: All patients, who have participated in the prospective cross-sectional COALA Study in 2000 with a structural clinical investigation including blood pressure measurement and symptom-limited exercise test were contacted for reevaluation of current clinical status, medical treatment, major cardiovascular events, office and ambulatory blood pressure measurement and the health-related quality of life questionnaire SF-36.
Results: From 273 eligible patients, we received data from 151 patients. Fifty -seven patients denied participating in the study. Nine patients died during the follow-up time with the age of 46 years (range 30-64 years), 25 patients had a cardiovascular event (12 procedures at the aortic valve or aortic arch, 8 procedures for recoarctation, 2 endocarditis, 2 cerebrovascular insults, one aortic dissection). Thirty-five patients (30%) showed normal ambulant blood pressure, 56 (48%) were on antihypertensive medication and 25 (22%) had arterial hypertension without receiving medication. Quality of life was good in the fields of physical role and pain. However, patients reported a significant impairment in general health, depending on the age. Arterial hypertension, echocardiographic measurements or exercise capacity from the COALA study were not predictive on functional health status.
Conclusion: Repaired coarctation of the aorta shows fairly low mortality on long-term follow-up. Important events are not only recoarctation, but also procedures at the bicuspid aortic valve. The rate of arterial hypertension is progressively increasing. However, the predictive value of clinical variables is limited, except the presence of a bicuspid aortic valve and the presence of aortic aneurysm for aortic valve procedures.

MP3-6
Supervised exercise training improves exercise capacity and chronotropic competence in children and adolescents with pulmonary arterial hypertension
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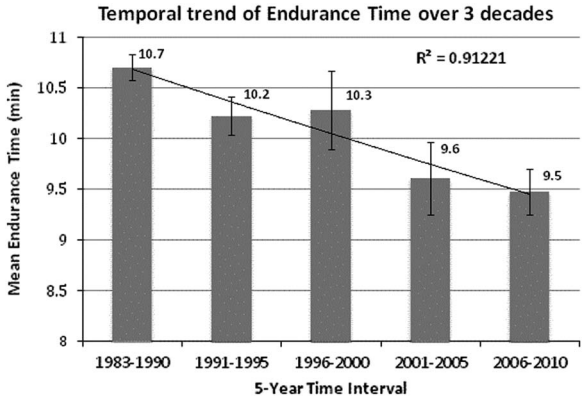
Background: Pulmonary arterial hypertension (PAH) is often associated with impaired exercise capacity and cardiac autonomic dysfunction. It has been shown that supervised training can improve exercise capacity in adult patients with PAH, which is usually associated with an improved peak oxygen uptake. So far, there is only limited experience of supervised exercise training in the pediatric age group. The objective of this prospective study was to assess the feasibility and efficacy of an exercise training program in children and adolescents with PAH.
Patients and Methods: Nine children with moderate PAH (six female, mean age 15.2 ± 3.8 years; mean pulmonary to systemic arterial pressure ratio 0.51 ± 0.19) performed homebased supervised exercise training (ergometer and theraband) for 16 weeks. Cardiopulmonary exercise testing and health-related quality of life was evaluated before and after 16 weeks of training. PAH-specific targeted medication remained unchanged during the study period.
Results: Exercise training was well tolerated in all patients. After 16 weeks of supervised training, patients significantly improved their exercise capacity (treadmill running distance increased from 589.5 ± 153.9 to 747.9 ± 209.2 meters ($p = 0.036$)). Peak oxygen uptake remained unchanged. Chronotropic index, a parameter that defines chronotropic response independently of age, resting heart rate, and functional state, improved from 0.77 ± 0.12 to 0.82 ± 0.11 ($p = 0.004$), while the increase in chronotropic index was related to the increase in running distance ($r = 0.62$; $p = 0.07$).
Conclusions: According to our experience, supervised exercise training is effective in children and adolescents with moderate PAH. The observed increase in exercise capacity was accomplished by an improved chronotropic competence rather than an increase in peak oxygen uptake as in the adult population. Therefore, chronotropic competence may serve as a physiologically important therapeutic target specifically for training programs in children and adolescents with PAH alongside the usual measurements of oxygen consumption. The prognostic impact of chronotropic competence needs to be assessed in future studies.

MP3-7
Trend in endurance level in healthy inner city children over a three decade period
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Introduction: With the increasing incidence of obesity and type II diabetes amongst youth in the past three decades, it is unclear how

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physical fitness and endurance has correlated with these trends. Understand how mean endurance time, a proxy for physical fitness, has changed in healthy individuals in the past 3 decades.
Methods: We reviewed the health records of consecutive apparently healthy Chicago city children who underwent an endurance test (Bruce Protocol) from 1983 – 2010. Patients with known cardiovascular conditions were excluded. Patients were divided in 5 groups in 5 year intervals based on the date of testing [figure1]. Endurance time, gender, race, age, and body mass index (BMI) data were collected.
Results: We identified 436 children (mean age 12.6 ± 3.2 yrs, 57% male). There was a significant difference in the mean endurance time between groups of 5-year intervals ($P < 0.001$) with endurance time being shorter at later testing years [figure1]. The endurance time was inversely correlated with the year of testing (Spearman's $r = -0.274$; $P < 0.001$). In contrast, there was no significant difference in the distribution of BMI between testing date 5-year intervals ($P = 0.205$). Multivariate linear regression model demonstrated that the date of testing, in 5 year intervals, was independently predictive of endurance time adjusting for BMI, race and gender and age ($P < 0.001$). BMI was the strongest independent predictor of endurance time ($P < 0.001$), followed by race ($P < 0.001$), age ($P = 0.001$), and gender ($P = 0.01$).
Conclusions: There is a downward trend in endurance time over the 27 years period among inner city kids. Temporal decline in endurance time was independent of factors known to be associated with endurance time such as BMI, age, gender, and race. BMI alone cannot fully explain the downward trending exercise time. Thus, factors such as deconditioning due to sedentary lifestyle and lack of motivation to endure on the treadmill among later generations may play a role in such decline.



MP3-8
Short-Term and long-Term Outcomes of Dilated and Non-compaction Cardiomyopathy Presenting during Childhood in West Sweden 1990-2015

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Objectives: To study the incidence, mortality and morbidity of dilated cardiomyopathy (DCM) and non-compaction of the left ventricle (LVNC) in children in West Sweden.
Materials and Method: Hospital records of children and adolescents, 0–18 years, diagnosed with DCM and/or LVNC over a 25-year period in Gothenburg region, were reviewed. Inclusion criteria for DCM were LV shortening fraction (LVFS) < 27% and symptoms of congestive heart failure. In LVNC inclusion criteria were according to international definition of the disease. Clinical data including echocardiography findings were registered. Survival was cross-checked against the Swedish Population Registry in October 2015. Standard statistical measures determined survival, normalization of cardiac function and morbidity in those with and without transplant or death.
Results: Sixty-nine patients were identified; 42 (61%) males and 27 (39%) females. The combined incidence of DCM and LVNC in children and young adults during this time period was 0.55 per 100 000 per year. Mean age at diagnosis was 6.7 years (1 day – 17.9 years). Eleven children (16%) presented before 4 weeks of age, 7 children (10%) between 1 month and 1 year, 21 children (30%) between 1 and 10 years and 30 children (43%) presented between 10 and 18 years. In the latter group most children also had a neuromuscular disease. Most patients had reduced ventricular function with a left ventricular shortening fraction (LVFS) < 27% at diagnosis (n = 43; 62%). The remaining patients with a LVFS > = 27% (n = 26; 38%), mainly had LVNC (n = 14) or were patients with left ventricular dilatation detected during screening for familial DCM. Transplant-free survival was 88.5%, 83.6% and 80.2% at one, two and five years after diagnosis, respectively. Combined mortality and transplant rate was 23.1% over the studied period.
Conclusion: The incidence of DCM and LVNC (0.55/100 000/year) in West Sweden was similar to reports from other countries. The majority of children with idiopathic DCM presented during the first year of life. Mortality was highest during the first year after diagnosis. The majority of children with DCM associated with other diseases were diagnosed beyond 10 years of age.

MP3-9
The Determination of Hypertension Persistence in Children Whose Aortic Coarctation Were Treated with CP Stent

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Objective: The aim of this study is to address presence of persistent hypertension in Aortic Coarctation (CoA) patients who were treated with endovascular stent, by using ambulatory blood pressure monitorization (ABPM), echocardiography, and biochemical evaluations.
Material and method: 20 patients (mean age: 14.2 ± 3.9 years) with CoA and 20 age- and sex-matched healthy controls were included and all results were compared between groups. Physical examination findings and blood pressures were recorded. Structure and functions of left ventricle, elastic functions of aorta, aortic diameters and carotid-intima media thickness (CIMT) were measured by echocardiography. Patients were assessed for hypertension by using ABPM. As indirect marker of arterial stiffness, pulse wave velocity (PWV) and augmentation index (aix@75) parameters were recorded.
Findings: Systolic and diastolic interventricular septum (IVSs, IVSd), diastolic left ventricular posterior wall (LVPWd), systolic left ventricular posterior wall (LVPWs) thicknesses, left ventricular mass (LVMass) and left ventricular mass index (LVMI; g/m2.7) values were found to be significantly higher in patient group. ABPM revealed, systolic pressure, daytime systolic pressure, mean arterial pressure and daytime mean arterial pressure values were found to be significantly higher in the patient group when compared to controls. Based on percentile values, 15% and 5% of

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the patients were pre-hypertensive and hypertensive, respectively in study group. PWV and cardiac output (CO) values were found to be significantly higher in the patient group. Based on PWV percentiles, PWV values were found to be above 95 percentile in 30% of the patients. No significant difference was found in aix@75 values and aortic stiffness, distensibility and strain values between groups. CIMT was found to be significantly higher in the patient group. No significant correlation was detected among CIMT, LVMI, aix@75 and PWV values (summarized in table).
Conclusions: It was shown that hypertension incidence, CIMT and PWV were greater than those in healthy population even CoA was corrected. This suggests that CoA is a part of generalized vasculopathy rather than being a localized narrowing. The use of new measurements in follow-up of patients with CoA can be helpful to predict risks. Further prospective studies are needed on this issue.

Parameters	Study Group	CONTROL	*P*
IVSd (cm)*	1.06 ± 0.24	0.8 ± 0.14	0.014
IVSs (cm)*	1.41 ± 0.26	1.14 ± 0.9	0.011
LVPWd (cm)**	0.94(0.78-1.02)	0.71(066-0.88)	0.009
LVPWs (cm)*	1.38 ± 0.23	1.10 ± 0.9	0
LVMI (g/m2.7)*	46 ± 10	32.3 ± 7.4	0
24 Hour SBP (mmHg)**	116.5 (106.5-126)	107.5 (105-111)	0.017
24 hour MAP (mmHg)**	88.5 (81-96)	83 (80-86)	0.043
Daytime SBP (mmHg)**	119.5 (108.2-130.2)	110.5 (106-114)	0.013
Daytime MAP (mmHg)**	91 (84-99)	84 (81-88)	0.021
CO (l/min)*	5.12 ± 0.43	4.8 ± 0.39	0.019
PWV (m/sc)**	4.8 (4.43-5.1)	4.45 (4.4-4.6)	0.009
CIMT (cm)**	0.40 (0.37-0.47)	0.34(0.31-0.41)	0.013

*mean ± standart deviation (SD), ** median (25p-75p).

MP3-10
A Retrospective Study In Children With Pulmonary Arterial Hypertension: A Single Center Experience

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Objectives: To determine the epidemiology, quality of life and management and effectiveness of the treatment of pediatric pulmonary arterial hypertension (PAH).
Methods: Patients who had diagnosis of PAH and treated at Department of Pediatric Cardiology, Gazi University Faculty of Medicine from February 2006 until October 2015, were evaluated retrospectively.
Results: Forty-one patients were included with a mean age of 83 ± 68 months (21 Female, 20 Male). 85.4% of patients had PAH associated with congenital heart disease and the ventricular septal defect was the most frequently defect. Four patients had primary PAH (9.8%), 1 patient had PAH caused by chronic lung disease and 1 patient had PAH caused by obstructive sleep apnea (%2.4). The most common clinical symptom reported was fatigue (65%). Telecardiography, electrocardiograpy (ECG) and echocardiography (ECHO) were performed in all patients. 6-minute walk test (6 MWT) were performed in twenty-five patients (61%). Cardiac catheterization was performed in 40 patients (97.6%). Only one patient couldn't be performed catheterization due to her bad clinical condition. Sixty percent of the patients, who had catheterization, were performed vasoreactivity test and 39% of them had positive response. There is no significantly difference between measured mPAB values in ECHO and catheterization (60 ± 53.5 mmHg vs. 65 ± 20 mmHg respectively). While the most frequently prescribed monotherapy was bosentan 48.8%, for the combined therapy it was bosentan plus iloprost with 22%. Eleven patients (26.8%) received monotherapy, 19 (46.3%) received dual therapy and 3 (14.3%) received triple therapy. During the follow up, the combined therapy was increasingly prescribed (43.9% versus 66.9%). The mean time of adding second drug was 27 months. During the follow up, 4 patients died. One– and five- year survival rates were 94% and 86%, respectively.
Conclusions: At the experienced centers, the positive results on the survival rate and life quality of patients with PAH, obtained with the current treatment options. As seen in this study, PAH is a progressive disease and the combined therapy is inevitable. Owing to this reason, the combined therapy should be considered in the early stage of the disease.

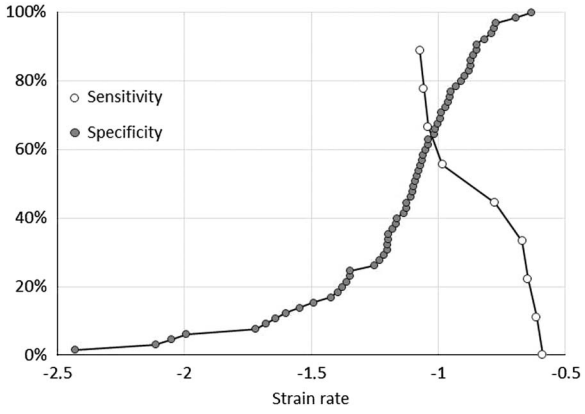
MP3-11
Longitudinal myocardial deformation as a predictor of impaired ventricular function in children after the Fontan operation

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Introduction: The short term survival of children with single ventricle (SV) heart defects after the Fontan operation has improved over the last decades, but impaired cardiac function is a major cause of morbidity and mortality over time. Cardiac magnetic resonance imaging (cMRI) is the golden standard in assessing SV function, but high costs and limited availability hamper it's routine use. A cheaper and more available alternative for assessing SV function is echocardiography. *Study aim:* To describe the association between myocardial deformation parameters assessed by speckle tracking echocardiography (STE) and SV function assessed by cMRI. *Methods:* Cross-sectional, multicenter study in 109 children after completion of the Fontan circulation. Echocardiography and cMRI were performed on the same day. STE peak longitudinal strain (ε) and systolic strain rate (SR) of the lateral wall of the dominant ventricle were measured off-line using the apical SV echocardiographic view. Impaired SV function by cMRI was defined as ejection fraction (EF) < 45%. Pearson correlation was used to assess associations between methods and independent T test was used to compare groups. *Results:* Mean age at study was 12.0 (range 9.7-14.6) years. Feasibility of STE was 80%. cMRI was performed in 70/109 participants. Mean cMRI EF was 53% (range 34-75%). Pearson R for cMRI EF versus global lateral peak longitudinal ε and SR was -0.30 (p = 0.01) and -0.21 (p = 0.07) respectively. Children with impaired EF by cMRI (n = 8) had reduced ε and SR values compared to children with normal EF (n = 61): -15.5 ± 3.7% versus -18.9 ± 3.5% (p = 0.01) and -0.85 ± 0.21 1/s. versus -1.13 ± 0.30 1/s (p = 0.01) respectively. The figure shows the sensitivity and specificity of SR to predict abnormal SV EF by cMRI. *Conclusions:* Although SV systolic function is generally well preserved in contemporary pediatric

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patients after the Fontan operation, some patients have impaired SV function assessed by cMRI. STE myocardial deformation parameters are moderately correlated with SV EF by cMRI but might be of use in predicting impaired EF in children after the Fontan operation.



MP3-12

Arterial functions and the effect of valve morphology in children with bicuspid aortic valve

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Introduction: We investigated aortic elasticity and flow mediated dilation (FMD) of brachial artery in children with bicuspid aorta and the effect of valve morphology on aortic elasticity.
Methods: The study included 39 patients (mean age = 9.7 ± 5.1; range: 1.5-18) with bicuspid aorta (BAV group) and 39 healthy children (mean age = 9.8 ± 3.82; range: 4-16) as control group. None of the patients had more than mild stenosis (gradient > 20 mmHg), mild regurgitation or associated congenital heart disease. Echocardiography was performed using Philips IE33 Echocardiography machine equipped with 5 MHz and L-11 Mhz linear transducer. LV systolic and diastolic functions, morphology of aortic valve, systolic and diastolic diameters of aortic annulus, sinus valsalva (SV), sinotubuler junction (STJ), ascending aorta (AscAo) were obtained and aortic strain (AS), aortic distensibility (AD), aortic stiffness index (ASI) were calculated. Flow mediated dilatation (FMD) of brachial artery was measured. The study group was divided into two subgroups as horizontal opening (AP; fusion between right and left coronary cusps) and vertical opening (LR; fusion between right and noncoronary cusps).
Results: Valve morphology was AP in 27 and LR in 12 patients. Aortic root and AscAo of the patients were larger than the controls according to Z-scores (Annulus, SV, STJ and AscAo were p: 0.001, p = 0.001, p = 0.02, p = 0.001, respectively). Stiffness index of SV, STJ and AscAo were higher than controls (p = 0.015, p = 0.001, p = 0.02, respectively). AS and AD has increased at SV and STJ levels in patients (p = 0.01, p = 0.04; p = 0.001, p = 0.001 respectively). FMD did not differ between groups. Z scores of aortic root were not significantly different between AP-LR subgroups. Asc Ao Z score was over 2 SD in 14 patients (10 of them had AP and 4 LR morphology, which was statistically significant, p = 0.05). AS (p = 0.02) and AD (p = 0.001) were lower and ASI (p = 0.001) was higher in the AP subgroup than LR subgroup in AscAo. Valve morphology had no effect on valve function. FMD was not different between two valve morphologies.
Conclusion: We showed that bicuspid aortic valve, especially with AP valve morphology, causes stiffer and less distensible aorta and dilation of AscAo is more likely with AP valve morphology unrelated to valve function.

MP3-13

Long-term functional capacity in patients with Extracardiac Fontan Circulation

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Objectives: thanks to advances in cardiac surgery life expectancy of patients with congenital heart disease has considerably grown. Aim of our study was to evaluate functional capacity in two groups of patients who underwent an extracardiac Fontan operation in the last 25 years (N = 135). There are few available data on long-term results of these patients.
Methods: we have divided the sample into two subgroups on the basis of months of follow-up from Fontan procedure. All patients were tested with maximal cardiopulmonary exercise test (CPET) with maximal oxygen uptake (VO2 max). Data from time of effort (TE),maximal heart rate (HR), maximal blood pressure (BP)and VO2max were also calculated as a percentage of normal predicted values. Lung function test to measure forced vital capacity (FVC), forced expiratory volume in 1 sec (FEV(1), and peak expiratory flow (PEF) was also performed.
Results: Group A (N = 77, male = 48, female = 29) months after surgery:180-60. Group B (N = 58, male 32, female 26) months after surgery:180-300. No statistically significant differences were found in TE% (58% group A vs 56% group B). We found a tendency, although not statistically significant, in decresing in VO2 max and in PEF between groups (group A: VO2 max 28.5 ml/kg/min + 5.6. PEF 84%. Group B VO2 max 25. ml/kg/min + 5.3. PEF 79%).
Conclusions: We can speculate that, although exercise tolerance in patients underwent extracardiac Fontan is greatly reduced compared with healthy controls, there is no evidence, however, of a significant deterioration of functional capacity over the years. It seems rather possible that the completion of this surgical procedure is leading to a gradual improvement of cardiorespiratory parameters of these patients. Further studies are needed to confirm our hypotheses.

MP3-14

Experience with Coarctation of the abdominal aorta (Mid-Aortic Syndrome) in children

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Objective: The aim of this study was to describe and analyze the features and outcomes of children and adolescents diagnosed with coarctation of the abdominal aorta.
Methods: Demographics, clinical data echocardiographic measurements, MRI and/or CT-scan imaging, angiography if available, surgical and/or medical treatment data were analyzed retrospectively.

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Results: Thirteen patients (6 males) were diagnosed with mid-aortic syndrome (MAS), at the mean age of 7.6years (9 days to 20years). Among them, 11 (85%) had a genetic disorder Williams Beuren syndrom in 2, neurofibromatosis in 1, metabolic disease in 1, Takayashu in 1, arterial dysplasia in 1, diabetes in 1 , deletion 1p36 and cardiomyopathy in 2, severe statural and neurological impairment in 1 and short-gut syndrome in 1. High blood pressure was observed in all cases, except one neonate and 2 infants with cardiomyopathy and severe LV systolic dysfunction. Hyppoplasia of aorta was supra-renal in 2 cases, infra-renal in 5 and global in 6; renal arteries were involved in 5 cases and mesenteric arteries in 5. Eleven patients received medical antihypertensive therapy (85%), 3 underwent percutaneous (23%) and 5 surgical (38.5%) aortic and renal angioplasty; only one case had neither medical nor interventional therapy. Median follow-up is 5 years (mean 8.8 years). Outcome was favourable in 8 cases, while 2 had uncontroled high blood pressure and one right heart failure. Two patients died (15.3%) because of renal and cardiac severe impairment.
Conclusion: MAS is frequently associated with genetic disorders and mainly complicates with HBP. Angioplasty is required in more than half of the cases. Renal involvement probably impacts the most on prognosis.

MP3-15
Acute Rheumatic Fever in South – East of Turkey: Clinical Features and Epidemiological Evaluation of the Patients over the Last Twenty Years
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Objective: The aim of this study was to evaluate retrospectively clinical and epidemiological features of rheumatic fever and rheumatic heart disease using last twenty years of data of our hospital, to investigate incidence and seasonal prevalence, to detect changes over time in clinical and epidemiological features by comparing two periods (first 7 and second 13 years).
Materials and Methods: The medical records of 396 patients with rheumatic fever (first attack or recurrence), rheumatic heart disease or previous rheumatic fever who admitted to Pediatric Cardiology Department of the Çukurova University between January 1993 and January 2013 were analyzed retrospectively. First period was between 1993 and 2000, and the second 2000–2013.
Results: There were 206 patient in first period and 190 patients in second period. The mean age of patients was 10.8 years. No significant difference was found between the first and second periods in distributions of age and gender. The peak season for the initial presentation was winter in both period. The estimated incidence rates of acute rheumatic fever were 2.7/100,000 in 2000, 0.7/100,000 in 2012. Among the major findings, the most common included carditis at 81%, arthritis at 75.2%, Sydenham's chorea at 10%, and erythema marginatum at 0.5%, at first period and carditis at 86%, arthritis at 79%, Sydenham's chorea at 18%, and subcutaneous nodules at 1.6%, at second period respectively. There were no significant differences about major findings between two periods. At the follow up, three patients had died and 17 patients had underwent to surgery.
Conclusion: Although the incidence of acute rheumatic fever has decreased, it still continues to be an important disease can lead to serious morbidity and mortality in our country. In future although genetic factors can not be changed, changes in environmental factors and healthy policy will decrease the frequency of the disease and its complications.

MP3-16
Treatment strategies for protein-losing enteropathy in Fontan patients
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Objectives: Protein-losing enteropathy (PLE) is a rare but severe complication occurring after Fontan procedure. Reduced cardiac output, chronic venous congestion and intestinal inflammation are believed to play a role in PLE development. However, given the multifactorial and largely unknown pathogenesis, no single proposed treatment strategy has proven universally successful. We thus evaluated the use of several surgical, interventional and medical therapeutic strategies and outcome in our PLE patients.
Methods: In a retrospective review of the entire cohort of 351 Fontan patients treated in our institution ($n = 272$ originally operated in our institution) we identified 24 patients (6.9%) diagnosed with PLE. Diagnosis was established when clinical criteria as diarrhoea and/or recurring edema, pleural effusions or ascites and abnormal laboratory values suggesting intestinal protein loss as low serum albumin and protein and elevated faecal alpha-1-antitrypsin were present. Data from clinical history, treatment and outcome was extracted and analysed.
Results: Freedom from PLE was 89.9% and 87.9% at 10 and 20 years. PLE developed at a median of 3.0 [0.1-16.5] years after Fontan. Haemodynamic issues as Fontan pathway obstructions, aortopulmonary collaterals, dysrhythmias or phrenic palsy were identified and addressed in 22 patients (91.7%). Treatment with one or more medication was initiated in 14 patients (58.3%) and included Sildenafil in 12 (50%), Bosentan in 4 (16.7%) and Budesonid in 8 patients (33.3%). Surgical and interventional procedures alone yielded stable clinical remission in 4 patients (16.7%). Additional medication achieved stable remission in another 6 patients (25.0%) and relapsing remission in 4 (16.7%). PLE-associated mortality is substantial, 10 patients (41.7%) died during follow-up. In Fontan patients with PLE, freedom from death or transplantation was significantly decreased compared to those without PLE (33.0% vs. 87.1% at 20 years after Fontan, $p < 0.001$).
Conclusions: Given the small numbers of Fontan patients developing PLE, studies investigating therapeutic strategies are difficult to perform. Treatment has to adjust to the individual patient's findings and may thus include elimination of Fontan pathway obstructions, optimizing ventilatory conditions, reducing pulmonary vascular resistance and anti-inflammatory therapies. Nevertheless, despite individually focussed therapy, reoccurrence is frequent and mortality in persistent PLE remains substantial. Cardiac transplantation should be considered early.

MP3-17
Intramyocardial injection of autologous bone marrow stem cells in children with Hypoplastic Left Heart Syndrome: the THABY Phase I trial
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Introduction and Objectives: Treatment of heart failure in Hypoplastic Left Heart Syndrome (HLHS) remains one of the biggest challenges among congenital heart diseases. Cell therapy has shown encouraging results in the field of heart failure. The aim of this trial was to determine the feasibility and safety of intramyocardial injection of mononuclear stem cells (MSC) in children with HLHS.

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Methods: We designed a nonrandomized trial to prospectively include 9 patients with diagnosis of HLHS in Norwood stage I or II. At the time of subsequent palliative surgery we performed intramyocardial injection of autologous bone marrow MSC derived under GMP conditions.
Primary end point was to examine the feasibility and safety of the procedure. Security was defined as absence of clinical events, arrhythmias on Holter ECG or focal lesions on echocardiography and cardiac MRI. As secondary end point we tried to assess improvement on cardiac function evaluated by MRI.
Results: Between November 2013 and April 2015, 9 patients (3-9 years) at Norwood stage II received MSC (339 × 106 cells ± 191 × 106) during Fontan surgery. Bone marrow extraction, cell processing and delivery were feasible and did no interfere with surgery. There were no incidences related with the injection (no more bleeding, no arrhythmias) We found no adverse effects on a 12 months follow-up. Cardiac function improved, although this did no reach statistical significance (FE 46% ± 5% vs 54% ± 13%; p = 0.253).
Conclusions: Intramyocardial injection of autologous bone marrow MSC is feasible and safe in children with HLHS during Fontan surgery. Phase 2 trials are needed to determine the potential benefits on cardiac function and clinical outcomes.

MP3-18
Evaluation ventricular arrhythmogenesis in Down syndrome patients with congenitally normal hearts

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Objective: Previously reported studies have shown that patients with Down syndrome (DS) with congenitally normal hearts (CHD) may exhibit cardiac functional abnormalities, valvular dysfunction , bradycardia, AV block and increased QT dispersion (QTd) and corrected QT dispersion (QTcd) that represent the heterogeneity of ventricular repolarization. Thus, due to increase in life expactancy in persons with DS, these persons are nedeed long-term follow-up in cardiovascular field. Recently, new ECG-derived indexes such as Tp-e which is the measurement of the interval between the peak and the end of the T-wave has emerged as a marker of transmural dispersion of repolarization (TDR). As Tp-e interval, Tp-e/QT ratio is also used as an index of ventricular repolarization. Prolongation of Tp-e interval and increased Tp-e/QT ratio have been found associated with malignant ventricular arrhythmias. The novel repolarization indexes Tp-e and Tp-e/QT have not been studied among these patients previously.The aim of this study was to evaluate Tp-e interval and Tp-e/QT ratio in DS patients without congenital heart defects.
Method: The standard 12-lead electrocardiograms of 134 children with Down's syndrome without congenital heart defects and 110 age-and-sex matched healthy children were assessed by a blinded specialist.
Results: Tp-e interval,Tp-e dispersion, Tp-e/QT and Tp-e/QTc ratios were found significantly higher in DS group compared to without DS group. Similarly reported study we found significantly higher QT and QTc dispersion in children with DS with congenitally normal hearts. We could not find any association between the Tp-e interval and Tp-e/QT ratio and age in DS group. And also any correlation were not determined between P, QT and QTc dispersions and gender.
Conclusions: In conclusion, our results showed that children with DS are more prone to ventricular and arryhtmias due to the prolonged durations of QT dispersions, Tp-e interval,Tp-e dispersion, Tp-e/QT and Tp-e/QTc ratios. Thus, all children with DS should be carrefully assess with electrocardiography according to the possible ventricular arryhthmias during the cinical follow up even in the absence of concommitant congenital heart disease.

Table 1. Features and electrocardiographic findings for Down syndrome (DS) patients and control subjects

	Down (n:134)	Control (n:110)	P value
Gender (M/F)	61/49	69/65	0.53
Median age (year)	6	5	0.005
Mean ± sd age (year)	6.8 ± 4.9	5.2 ± 3.5	0.005
Heart rate	108 ± 28.8	98 ± 23.5	0.002
Tpeak-end max (ms)	73.7 ± 24.4	64.7 ± 23.4	0.004
Tpeak-end min (ms)	43.3 ± 18.2	49.0 ± 19.0	0.018
Tpeak-end dispersion (ms)	30.4 ± 17.0	15.6 ± 7.8	0.000
QTmax (ms)	334 ± 32.4	328 ± 27.9	0.14
QTmin (ms)	291 ± 33.7	296 ± 26.0	0.14
QT dispersion (ms)	43.4 ± 14.7	31.9 ± 9.4	0.000
QTcmax (ms)	397 ± 23.1	387 ± 24.0	0.001
QTcmin (ms)	346 ± 25.6	361 ± 27.6	0.000
QTc dispersion	50.8 ± 18.2	25.4 ± 11.4	0.000
Tpeak-end / QT	0.22 ± 0.10	0.19 ± 0.06	0.013
Tpeak-end / QTc	0.18 ± 0.06	0.16 ± 0.05	0.015

MP3-19
Outcome of symptomatic partial atrioventricular septal defect necessitating repair during infancy

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Background: Patients with partial atrioventricular septal defect are typically asymptomatic and electively repaired at pre-school age. We sought to analyse the mortality, re-operation rate and risk factors with adverse outcome of symptomatic infants with partial atrioventricular septal defect necessitating repair during infancy in one UK centre in recent era(2000-15).
Methods: The inclusion criteria were as follows:1. partial atrioventricular septal defect, 2. heart failure unresponsive to medical treatment, and 3. bi-ventricular repair during the first year of life. Thirteen patients (6males), who had undergone corrective surgery at a median age of 197(16-346) days, were included. Seven patients had an associated syndrome: Down(n = 3), trisomy 47XXX(n = 1), Noonan (n = 1) and VACTERL (n = 1). Five patients required either surgery (4 coarctation repair) or transcatheter intervention (1balloon angioplasty of pulmonary artery) before primary repair. One patient with tracheobronchomalacia required cardiopulmonary resuscitation before repair.
Results: Median weight at primary repair was 5.1 kg(3.3-7.5 kg), median hospital stay 19 days (4-128 days). Five patients had unfavorable anatomy of the left atrioventricular valve: the leaflets were severely dysplastic in 2 cases, mildly dysplastic in 1 case, 1 patient had unusual orientation of the zone of apposition and 1 patient small left atrioventricular valvar annulus.
One patient died 68 days after primary repair from a non-cardiac cause.

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Five patients (42%) were reoperated at a median interval of 2.8 years (33days–8.3years), including left atrioventricular valvar repair for severe regurgitation in 3 cases, relief of subaortic (n = 1) and pulmonary vein stenosis (n = 1). One patient required repetitive left atrioventricular valve repair, followed by mechanical valve replacement and pacemaker implantation.
The probability of developing severe left atrioventricular valvar regurgitation 10 years after primary repair in patients with unfavorable anatomy was 100% and 0% in patients with favorable anatomy ($p < 0.01$).
After a median follow-up of 3.8years, all patients were asymptomatic from the cardiac point of view and no patient had a severe atrioventricular valvar regurgitation.
Conclusion: Although the in-hospital mortality is not negligible in patients necessitating repair of partial atrioventricular septal defect in infancy, the long term outcome is favorable with no reported mortality and good functional status. The rate of re-operation mainly due to severe left atrioventricular valvar regurgitation is high in patients with dysplastic valve.

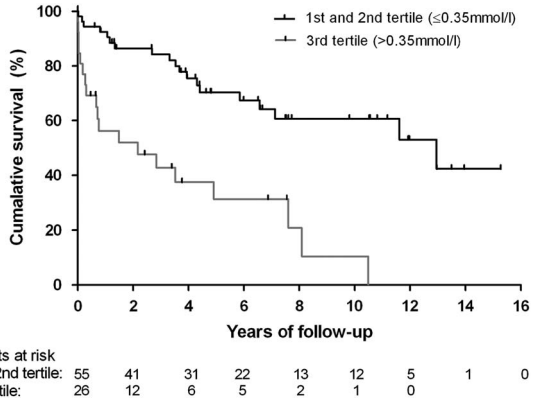


Figure 1
Transplantation-free survival stratified by uric acid levels measured at baseline. The log-rank test showed a significant difference ($p < 0.001$) between the groups.

MP3-20
Uric Acid As A Biomarker in Paediatric Pulmonary Arterial Hypertension
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Introduction: For optimal clinical decision-making in the treatment of paediatric pulmonary arterial hypertension (PAH), it is important to have a reliable, non-invasive and inexpensive biomarker that can be used to monitor disease severity throughout the course of the disease. The aim of this study was to evaluate the association of uric acid levels, at baseline as well as during the course of the disease, with disease severity and outcome in children with PAH.
Methods: This longitudinal study included data of 81 consecutive children from the Dutch National Network for paediatric PH which were prospectively followed according to a standardised protocol, including serum uric acid measurements. Uric acid values at baseline as well as during the course of the disease were correlated with disease severity markers. In addition, the association of uric acid with death or lung-transplantation at baseline and during follow-up was determined.
Results: Higher serum uric acid levels were associated with higher WHO-FC ($\beta = 0.234$, $p = 0.003$) and NT-proBNP ($\beta = 0.236$, $p = 0.014$) and lower TAPSE Z-scores ($\beta = -0.663$, $p = 0.027$) at baseline. Longitudinal analysis demonstrated that this association remained stable throughout the course of the disease. Lung-transplantation free survivors had a significantly ($p < 0.001$) lower value of uric acid at baseline. They also showed a significantly ($p < 0.001$) smaller increase in uric acid during follow-up compared to non-survivors (0.004 mmol/l/year vs. 0.014 mmol/l/year). A 50% deterioration of uric acid during follow-up was associated with a 3.9 times higher risk of death or lung-transplantation.
Conclusions: This study demonstrates that higher serum uric acid levels are associated with worsening disease severity and increased mortality risk in children with PAH, an association that remained stable throughout the course of the disease. Monitoring serum uric acid provides valuable information, which could help guide decisions in the management of paediatric PAH.

MP4-1
Ventricular Noncompaction and the association with congenital heart defects
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Introduction: Noncompaction of the Ventricular Myocardium (NVM) is a rare condition with debatable clinical significance and no clear-cut morphological diagnostic criteria. Non-systematic reviews show that some congenital cardiac defects can be associated with NVM. The goal of this study was to evaluate the incidence of NVM in heart specimens with congenital defects.
Methods: The compacted and noncompacted myocardial layers were grossly measured in three different heart walls: inlet, apex and outlet of the left ventricle (LV). Coefficients of non-compaction were calculated according to the criteria described by Jenni and Chin.
Results: Five types of cardiac defects were studied: isolated ventricular septal defect (VSD, n = 51), isolated atrial septal defect ASD, n = 15), atrioventricular septal defect (AVSD, n = 41), transposition of the great arteries (TGA, n = 54), isomerism of the atrial appendages (n = 29) and Ebstein malformation (n = 24) (total = 214 heart specimens; 46% from male patients, 64% under 12 months of age). According to the Chin's criterium the percentages of non-compaction at the LV apex were respectively: 50.98% in VSD specimens; 25% in Ebstein malformation, 22.2% in TGA, 6.7% in ASD, 6.1% in Isomerism, and 2.4% in AVSD. According to Jenni's criterium the percentages were: 33.3% in VSD, 31.0% in Isomerisms, 16.7% in TGA, 8.33% in Ebstein, 6.7% in ASD and 2.4% in AVSD. Considering all hearts, the apex region showed the largest percentage of non-compaction (Chin = 24.5%; Jenni = 16.3%). Concomitance of non-compaction in different regions of the same heart was low (4 hearts according to Chin's criterium and 2 according to Jenni's). In the TGA group, there was no association of MVN and the presence of VSD ($p = 0.067$). The Kappa test was used to compare the diagnosis of non-compaction according to both criteria. In ASD, AVSD, TGA and isomerism there was an almost perfect agreement ($p < 0.001$; Kappa > 0.8), in Ebstein there was a moderate agreement ($p = 0.023$; Kappa = 0.412), and in VSD there was a fair agreement ($p = 0.05$; Kappa = 0.289).
Conclusion: Predominance of non-compaction at the LV apex of VSD heart specimens has not been previously described. The true significance of regional myocardial non-compaction in congenital

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heart defects is unknown, and needs correlation with clinical and outcome data.

MP4-2
Prevalence of and performance of echocardiography in the detection of asymptomatic pulmonary vein obstruction in children after pulmonary vein repair

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Objectives: Pulmonary vein (PV) repair is burdened by a significant risk of (re-)stenosis. A proportion of patients with PV stenosis is asymptomatic. Echocardiography is routinely performed to screen for PV stenosis while cardiac magnetic resonance (CMR) is the gold standard for the detection of PV obstruction.
The aims of this study were a) to determine the prevalence of postoperative PV stenosis in a cohort of patients without clinical suspicion of obstruction, and b) to evaluate the ability of echocardiography to identify PV stenosis in these patients.
Methods: Echocardiography and CMR in asymptomatic pediatric patients after PV repair were retrospectively compared.
Results: Thirty-five patients were included: 30 had had total anomalous pulmonary venous connection (APVC), 3 had had partial APVC, and 2 had been operated for primary PV stenosis. Repair techniques included conventional (n = 13) and sutureless (n = 22). For 4 out of 22 sutureless patients, the indication was recurrent PV stenosis. The time between the surgery and CMR was a median of 83 months (6 days to 17 years).
CMR revealed significant stenosis in at least one pulmonary vein in a total of 11 children (31%), two following conventional repair and 9 after sutureless repair. The time between echocardiography and CMR was a median of 1.6 months (0–18). Echocardiography was unable to visualize all PVs in 16 patients (46%). In 8 patients (23%) the results regarding PV patency by echocardiography were incorrect, as compared to CMR. In 26% (9/35) a significant stenosis of at least one PV was missed by echocardiography. Compared to CMR echocardiography diagnosed PV stenosis correctly with a sensitivity 0.18, a specificity of 0.88, a positive predictive value of 0.40, and a negative predictive value of 0.70. The Kappa analysis between echocardiography and MRI showed poor agreement (0.11).
Conclusions: A third of this, albeit selected, cohort of patients who are asymptomatic after PV repair had significant obstruction of at least one PV. Echocardiography frequently missed obstruction in these patients.
CMR should be considered as an additional screening test in children after PV repair, irrespective of symptoms or echocardiographic findings.

MP4-3
Echocardiographic arterial measurements in complex congenital diseases before bidirectional Glenn: Comparison with cardiovascular magnetic resonance imaging

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Introduction: This study sought to investigate diagnostic accuracy of echocardiographic measures of great vessels in patients before bidirectional cavopulmonary connection (BCPC) compared to cardiovascular magnetic resonance (CMR).
Methods: Seventy-two patients (61% after Norwood operation) undergoing BCPC between 2007 and 2012 were assessed pre-operatively using echocardiography and CMR. Bland-Altman analysis and correlation coefficients were used for comparison of echocardiography and CMR measurements. Sensitivity, specificity, positive and negative predictive values were calculated to assess the ability of echocardiography to detect vessel stenosis.
Results: Twenty-four percent of all vessel measurements could not be made by echocardiography due to poor image quality. Acquisition of unsatisfactory images was higher in non-sedated patients. Although there was reasonable correlation (0.68-0.90) and low bias (-0.8 to 0.5), there were wide limits of agreement between echocardiography and CMR demonstrating poor agreement. Sensitivity and specificity for pulmonary branches were moderate (sensitivity for right pulmonary artery (RPA) 67%, left pulmonary artery (LPA) 54%, specificity for RPA 65%, LPA 72%) with low levels of accuracy (RPA and LPA 42%). Sensitivity, specificity and accuracy were better for aorta (82%, 86%, 63% respectively).
Conclusion: This study demonstrates modest agreement between echocardiographic and CMR measures of vessel diameter and stenosis detection. Approximately a quarter of all vessel segments could not be measured using echocardiography due to poor image quality, which was significantly lower in non-sedated patients. These findings suggest that in complex patients echocardiography cannot be used as the only method of comprehensive assessment of the vasculature before BCPC.

MP4-4
Right and left atrium volumes in Repaired Tetralogy of Fallot: impact on ventricular dysfunction and adverse cardiac event

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Introduction: Left atrium (LA) enlargement is directly proportional to the severity of the diastolic left ventricle (LV) filling abnormality, reflecting the severity of underlying cardiovascular burden. The meaning of right atrium (RA) and LA enlargement in Repaired Tetralogy of Fallot (repaired-TOF) has not been fully studied. The aim of the study is to evaluate the RA and LA atriums volumes in a large cohort of repaired TOF, to determine if they reflect conventional indexes of ventricular dysfunction and to assess their association to adverse cardiac event.
Methods: Consecutive repaired-TOF with age $\geq$ 12 years referred to our center have been evaluated by a protocol comprehensive of clinical evaluation, electrocardiogram, trans-thoracic Echo (TTE), CMR comprehensive of atrii volumes, cardio-pulmonary-exercise test (CPT), NT-Pro BNP.
Results: the population study was constituted of 170 repaired-TOF (28 ± 13 years). RA indexed volume (RAVi) correlates with LA indexed volume (LAVi) (r: 0.59, $P < 0.01$), with indexed right ventricular end-diastolic volumes (RVEDVi) (r: 0.49, < 0.001; r: 0.7, $p < 0.001$) and inversely with RVEF (r: -0.21, $p = 0.005$) and LVEF (r: -0.22, $p = 0.003$). RAVi and LAVi increase with age: respectively r: 0.52, $P < 0.001$, r: 0.59 $p < 0.001$; and correlate

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with age at primary repair (r: 0.45, $p < 0.001$; r: 0.7, $p < 0.001$), indexed LV end-diastolic volumes (LVEDVi) (r: 0.32, $p < 0.001$; r: 0.38, $p < 0.001$). RAVi was higher in patients with significant tricuspid regurgitation: 51 ± 16 ml/m2 vs 63 ± 19 mL/m^2, $p < 0.001$. LAVi and RAVi resulted less dilated in patients classified NYHA I in comparison with patients in NYHA II/III: 29.6 ± 7.8 mL/m^2 vs 39.6 ± 22 mL/m^2 $p < 0.001$ and 52.5 ± 16.3 mL/m^2 vs 62.3 ± 23 mL/m^2 $p < 0.001$, with Ln Nt-ProBNP r: 0.45, $p < 0.001$ and r: 0.45, $p < 0.001$ and RA correlate inversely with Vo2/kg/min at CPT: r: -0.22, $P = 0.006$. LAVi and RAVi were associated to adverse cardiac event before and during Follow-up ($p < 0.01$), mainly for atrial arrhythmias for RAVi, while LAVi was associated to both atrial and ventricular arrhythmias.
Conclusion: In our population of repaired-TOF, RAVi correlates with LAVi. Both of them correlate with conventional indexes of ventricular dysfunction and were associated to adverse cardiac event before study and during follow-up. Further longitudinal studies are required to shed light on the prognostic value of the atrii.

MP4-5
Patients after Fontan with a 'total cavopulmonary connection' Fontan modification develop more collateral flow compared to 'old-fashioned' Fontan modifications

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Objectives: Fontan palliation has evolved over time. Among many modifications creation of 'total cavopulmonary connection (TCPC) ' Fontan such as intracardiac tunnel or extracardiac conduit has been proposed as a superior alternative to 'old-fashioned' Fontan modifications such as RA-PA, or RA-RV tunneling. Collateral flow is a well-known residuum in patients with Fontan. However, no studies have examined collateral flow between different types of Fontan modifications. So the aim of this study was to compare collateral flow of patients with a TCPC to collateral flow of patients with 'old-fashioned' Fontan modifications.
Methods: We conducted a retrospective study on patients with any type of Fontan, who underwent a routine clinical CMR at our institution over a 6-year period with collateral flow quantification. The entire cohort was divided into two groups: 32 patients with TCPC, (median age 15(2-60)yrs); 7 patients with 'old-fashioned' Fontan modifications, (median age 31(21-35)yrs). Patients with a fenestration were excluded. We determined collateral flow by using the commonly used equation: flow volume aorta ascendens – (flow volume superior vena cava + flow volume inferior vena cava).
Results: Collateral flow was larger in patients with a TCPC compared to patients with 'old-fashioned' Fontan modifications [median 0.6((-0.1)-1.7) vs. 0.2((-0.04)-0.5) L/min/m^2, $p < 0.006$]. Additionally, cardiac index measured as the flow volume in the aorta ascendens was also larger in patients with a TCPC compared to patients with 'old-fashioned' Fontan modifications [median 3.2(2.2-4.9) vs. (2.6(1.9-3.3) L/min/m^2, $p < 0.01$].
Conclusions: Patients years after any type of Fontan modifications have substantial collateral flow. However, patients with a TCPC have significantly more collateral flow compared to patients with 'old-fashioned' Fontan modifications. This leads to a significantly higher work load of the heart as demonstrated by the larger cardiac index of patients with a TCPC compared to patients with 'old-fashioned' Fontan modifications. We suggest that the larger collateral flow in TCPC may be due to different flow characteristics in the superior vena cava compared to 'old-fashioned' Fontan modifications.

MP4-6
3D Printed models for Interventional Planning - Assessment in patients with Coronary Fistula

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Introduction: Coronary fistulae represent one of the most challenging anatomical defects to define accurately. We investigate the added benefit of three dimensional (3d) volume rendering (VR) of cross-sectional images over 3d printing for diagnosis and interventional planning.
Methods: Four cases of coronary fistula were considered for transcatheter closure. Multidetector computed tomography (MDCT) with retrospective gating (3 cases) or MRI (3d whole-heart balanced steady-state-free-precession) images were used. Segmentation was performed using Mimics v18.0, Materialise, Leuven, Belgium. Printed models used polyjet technology (Tango plus material) or fused deposition modelling (polyurethane).
Two cardiologists, independently reported source images. They were then given access to 3D VR and allowed to review source images and revise the report. The case was then discussed with the interventional team to plan device closure. Finally, the team was given access to the printed model and allowed to review all images and reconsider the plan.
Results: Case 1 had an RCA (right coronary artery) to LV (left ventricle) fistula. Case 2 had a fistula to the CS (coronary sinus) communicating with RCA and LCx (left circumflex). Case 3 had a LCx artery fistula draining to both the CS and LAA (left atrial appendage) with two aneurysms. Case 4 had a fistula from the left main stem (LM) to the SVC–RA junction.

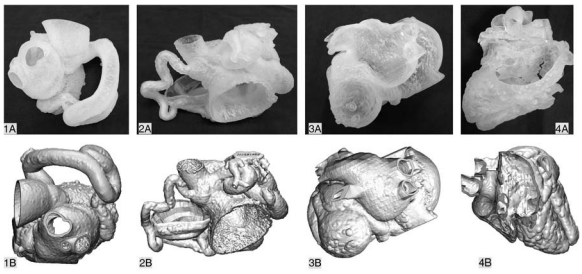


Figure. 1) RCA to LV fistula. 2) Coronary fistula to CS involving the RCA and LCx. 3) Left circumflex fistula draining to CS and LAA (with 2 aneurysm). 4) Fistula from left main stem to SVC-RA junction.
A) 3D printed model on Tango Plus. B) 3D Mesh performed on Mimics. RCA = right coronary artery. LV = left ventricle. CS = coronary sinus. LCx = left circumflex. LAA = left atrial appendage. SVC = superior vena cava. RA = right atrium

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Using source images alone, both cardiologists were able to correctly describe the course and drainage of 2/4 cases. The use of volume rendering increased this to 4/4 for both cardiologists. However, the interventional plan made after review of source imaging and VR, was changed for two cases after reviewing the 3d printed models alongside conventional images. In case 3, the initial approach and device sizing was changed and in case 2, the decision to proceed was changed after the extent of bilateral coronary involvement was fully appreciated.
Conclusions: Coronary artery fistulae require detailed 3D geometric analysis using VR in addition to MDCT or MRI source images for correct diagnosis. In these cases, 3D printing adds incremental value to the determination of feasibility and approach to intervention.

MP4-7
Increased diffuse myocardial fibrosis on cardiovascular magnetic resonance T1-mapping and intraventricular conduction delay on electrocardiogram on long-term follow-up after surgical septal myectomy in children with obstructive hypertrophic cardiomyopathy

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Introduction: The long-term effect on myocardial remodeling of surgical septal myectomy performed in children with severe obstructive hypertrophic cardiomyopathy (HCM) and its influence on cardiac conduction system disease or arrhythmias is unclear.
Methods: Patients with childhood HCM were evaluated by electrocardiogram (ECG), Holter ECG, cardiopulmonary exercise test, transthoracic echocardiography (TTE), genetic testing, and cardiovascular magnetic resonance (CMR). Diffuse myocardial and focal fibrosis was assessed by myocardial extracellular volume fraction (ECV) with T1-mapping and late gadolinium enhancement, respectively, on CMR. Patients with were compared to patients without prior myectomy.
Results: Age, gender, and genetic etiology were similar between groups. Patients with prior myectomy had greater ECV on CMR T1-mapping and longer QRS intervals on ECG as compared to patients without prior myectomy. Myocardial hypertrophy on TTE or CMR, focal fibrosis by late gadolinium enhancement CMR, atrioventricular conduction time on ECG, and arrhythmia vulnerability on Holter ECG or exercise testing were not different between groups (Table).
Conclusion: Long-term follow-up demonstrates increased diffuse myocardial fibrosis and intraventricular conduction delay after surgical septal myectomy performed during childhood for obstructive HCM. This might be due to intrinsic general myocardial remodeling in HCM after severe left ventricular outflow tract obstruction or secondary to inflammatory triggers during extracorporeal circulatory bypass in patients requiring cardiac surgery during childhood.

	Septal myectomy (9)	No septal myectomy (5)	p-value
Male / Female (N)	7/2	4/1	NS
Noonan syndrome / non-syndromic HCM (N)	5/4	2/3	NS
Age at surgery (yrs)	2.5 (0.2/16.1)	-	-
Age at study (yrs)	15.3 (5.8/30.6)	14.8 (11.1/18.6)	NS
ECG: QRS (ms)	114 ± 22	82 ± 15	0.014
ECG: PQ (ms)	140 ± 49	128 ± 30	NS
CMR: ECV total (%)	30.5 ± 4.4	26.6 ± 1.7	0.026
CMR: late gadolinium enhancement (N)	4/9	2/5	NS
CMR: Left ventricular mass (g/m^2)	123 ± 97	91 ± 36	NS
TTE: IVSd Z-score	3.0 ± 2.7	3.1 ± 2.2	NS
TTE: LVPWd Z-score	2.5 ± 1.9	3.3 ± 1.5	NS
TTE: Ejection fraction (%)	74 ± 9	82 ± 11	NS
Rhythm disturbances (N)	1/9	0/9	NS

Data expressed in mean ± STD or median (min/max) according to sample distribution; N: number of patients; NS: no statistical significance; yrs: years; ms: milliseconds; IVSd: enddiastolic interventricular septum thickness ; LVPWd: enddiastolic left ventricular posterior wall thickness.

MP4-8
The role of speckle-tracking echocardiography in early detection of cardiotoxicity in asymptomatic cancer survivors children

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Introduction: Advances in cancer treatment have led to 5-year survival rates approaching 80% for the majority of paediatric malignancies; however, late effects of therapy on cancer survivors are now becoming a relevant concern. Although cancer treatment-related heart damage often becomes apparent years later, conventional echocardiography cannot identify patients that will develop toxicity in a pre-clinical stage. Consequently, we are not able to apply early intervention measures preventing damage progression. The aim of this study is to evaluate the role of speckle-tracking echocardiography in early detection of cardiotoxicity in asymptomatic cancer survivors children.
Patients and Methods: We performed an echocardiographic study in a cohort of 32 cancer survivors children who received anthracyclines for a haematological malignancy and completed cancer therapy at least 2 years earlier. Patients were stratified into two groups (low/high risk) according to the existence of known risk factors for cancer treatment-related cardiotoxicity - e.g. anthracyclines cumulative dose, total mediastinal irradiation dose, number of haematopoietic stem-cell transplantation (HSCT) procedures. Cardiac function was assessed by evaluating parameters obtained from both conventional and speckle-tracking echocardiography. We compared measures to reference values of an age-balanced population and correlated with the presence of risk factors.
Results: After a median follow-up of 7 years, at conventional echocardiography none of the patients manifested an evident LV systolic or diastolic dysfunction; however, 13/32 survivors (41%) showed an alteration of either LV diastolic or dimensional parameters or both. At speckle-tracking echocardiography, all patients showed a significant reduction in global longitudinal strain (GLS) systolic peak (mean difference: -7.5%, p-value < 0.001), with the highest decrement observed in the high-risk group (z-score mean difference between groups: 1.1; p-value < 0.025).

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We observed a statistical correlation between GLS reduction and the number of HSCT procedures (z-score mean difference between ≥ 2 transplantation and no transplantation: 1.4; p-value < 0.025), but not with anthracyclines or mediastinal irradiation doses.
The present study shows that speckle-tracking echocardiography can detect early heart dysfunction in cancer survivors, allowing us to reveal subclinical toxicity in the entire study population. We observed that the number of HSCT procedures is the only independent risk factor correlating with the extent of strain alteration.

MP4-9
Feasibility of lung ultrasound in pediatric cardiac surgery: a new basic tool for pediatric cardiologist. Preliminary experience
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Background: Lung ultrasound (LUS) is gaining consensus as a non invasive, easy, fast and low cost technique for the diagnosis of pulmonary disease. Pulmonary complications are very common in pediatric cardiac surgery. Despite this of LUS remain limited. Aim: to test the feasibility of LUS in pediatric cardiac surgery.
Methods: From June 2015 to November 2015, 380 LUS examinations have been performed examination in 103 pts (0-17 years). Of these 62 children (median age 6 months, range 1 day-16,7 years) who had one or more examinations at fixed post-operative times (12-24 hours, at 5-7 days and before discharge) were selected. According to standardized protocols for each hemi-thorax 3 major areas (anterior/lateral/posterior) have been evaluated separately and every area has been further divided into the upper/lower half. We mainly evaluated the presence/degree of B lines, the presence/severity of pleural effusion/atelettasia.
Results: lateral examination was always feasible while anterior and posterior areas were approachable only in 67% and 85% respectively (due to medications and difficult in mobilization). B lines were present in all post-operative patients. The percentage of B lines did not vary from 12-24 hours versus 5-7 days (respectively $90 \pm 7\%$ vs $90 \pm 10\%$) while reduced significantly at discharge ($51 \pm 13\%$ $p < 0.0001$).
Pleural effusion (from mild to severe) were diagnosed in 58% at 12-24 hours, in 50% at 5-7 days, and in 25% before discharge. Atelectasis (from trivial to severe) were present in 73% at 12-24 hours; in 80% at 5-7 days, and in 30% before discharge. Atelectasis were seen in isolation in 29% while in the remaining were associated to pleural effusion There were no difference in distribution and severity of effusion and atelettasia among the two lungs.
The posterior approach was much more sensible than anterior/lateral in the diagnosis and severity estimation of effusion/atelettasia (Kappa coefficient ranging from 0.00 to 0.09).
Conclusions: LUS is feasible after pediatric cardiac surgery at all the ages. LUS allows differential diagnosis and severity estimation of effusion/atelectasis and the posterior approach is much more accurate than anterior/lateral for this setting. Wider studies are necessary to reinforce these data and provide new information.

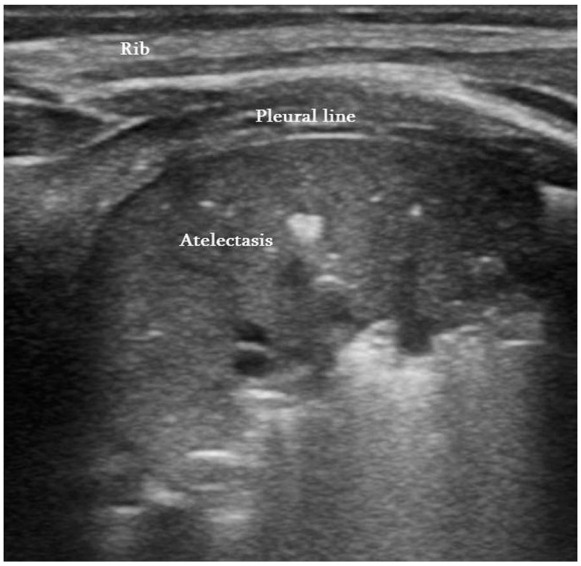


Figure 1. Example of moderate atelectasis.

MP4-10
Real-Time 3D-Echocardiography of the Left Ventricle - Pediatric reference-values for left ventricular volumes using knowledge based reconstruction
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Background: Determining left ventricular (LV) 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 LV assessment, however pediatric reference values are lacking. The aim of the study was to establish pediatric percentiles.
Methods: In a multicenter prospective design, 634 healthy children and adolescents (range, 1 day- 216 months) underwent RT3DE imaging of the LV. After initial training, echocardiography was performed 5 by different operators with 5 different ultrasound machines of 2 vendors (ie 33, Philips; Vivid 7, GE) in 3 different centers. 583/634 (91.9%) RT3DE data sets were quantified using a vendor-independent software (TomTec LV2.7, Unterschleissheim, Germany). In a core lab, all analysis were done using a contour finding sensitivity of 30, depending on our previous correlation with CMR-data (1). Reference centile curves were computed using Cole's LMS method.
Results: Percentiles for LV enddiastolic, endsystolic and stroke volumes (EDV, ESV, SV) were gender dependent and are presented as volumes indexed to body surface (Haycock) in Figure 1. Intra- and interobserver-variation for all parameter were excellent with intraclass correlation coefficients (ICC) between 0.935-0.998.
Conclusions: In children, calculation of LV volumes by vendor independent software is reproducible and accurate (1) if agreement on data acquisition and data analysis has been defined. The percentiles provided are based on a large sample size using different ultrasound machines and a vendor-independent software and may be useful for clinical practice and research.

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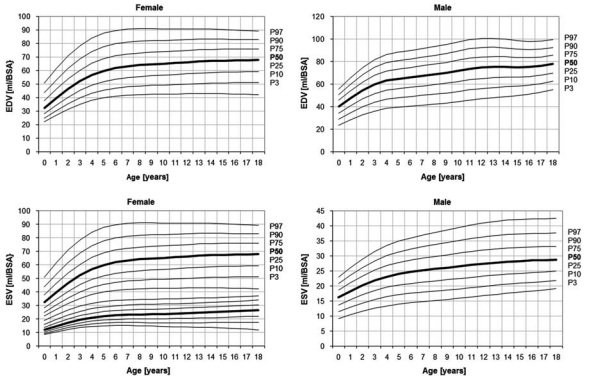


Figure 1
Gender-dependent percentiles for LV volumes indexed to body surface (Haycock).

1. Laser KT, Bunge M, Hauffe P al. Eur J Echocardiogr. 2010 Mar; 11(2):138-48.

MP4-11
Additional Value of Three-Dimensional Rotational Angiography in the Diagnostic Evaluation and Percutaneous Treatment of Children With Univentricular Hearts

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Introduction: Children with univentricular hearts undergo staged palliation to a Fontan circulation. In the interstages, detailed anatomical information is required to proceed. Anatomical information acquired by conventional 2D angiography (CA) is often difficult to interpret. Three-dimensional rotational angiography (3DRA) is a promising imaging technique that makes real time 3D images and is suitable for displaying cardiovascular anatomy. The aims of this study are to evaluate the diagnostic value, percutaneous treatment and additional value of 3DRA in children with univentricular hearts and to design a 3DRA image protocol.
Methods: A retrospective study was performed in which demographic data, clinical data and catheterization details (amount of contrast, radiation doses, interventions and complications) of both imaging techniques were collected. Image quality, interventional success and additional value of 3DRA were reviewed and scored. Data of the two techniques were compared to each other. Technical settings of all 3DRAs (rapid pacing, number and location of contrast injection, amount of contrast and contrast dilution) were collected.
Results: Between 2003 and 2014 128 patients underwent 186 CAs and 56 3DRAs. Image quality was good with both techniques, with better imaging of shunt and coronary arteries with 3DRA ($p = 0.000$). More interventions were performed in the 3DRA versus CA group: 44/56 3DRAs compared to 84/186 CAs ($p = 0.000$). Intervention success rate ($p = 0.087$) and complication rates ($p = 0.949$) were similar. Amount of contrast, number of angiographies and radiation used did not significantly differ. The additional value of 3DRA arises from the good imaging capacities and 3D reconstruction of cardiovascular anatomy, collaterals, stenosis, vessel-vessel and vessel-bronchi interactions.
Conclusion: 3DRA is superior to CA in displaying the shunt, coronary arteries, collaterals, stenoses, vessel-vessel and vessel-bronchi interactions in children with univentricular hearts, with comparible radiation dosages, amount of contrast used, intervention success rate and complication rate.

MP4-12
Fontan circulation shows deranged relative intra-cardiac blood flow components but normal kinetic energy profile

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Introduction: Ventricular function assessment is an important part of the assessment of the Fontan circulation. Recent advances in 4D flow cardiovascular magnetic resonance imaging now allow detailed assessment of left ventricular intra-cardiac flow patterns and kinetic energy (KE). A previous study has shown abnormal flow parameters in adults with dilated cardiomyopathy and low normal ejection fraction. We hypothesised that flow changes may already be apparent in the ventricle in Fontan patients prior to a decline in ejection fraction.
Methods: We prospectively enrolled 16 participants (8 Fontan patients and 8 age and sex matched controls) who underwent 3 T CMR assessment. For analysis the ventricular volume was divided into 4 functional components; direct flow (DF), delayed ejection flow (DEF), retained inflow (RI) and residual volume (RV). For each components the volume was calculated and expressed as percentage of end diastolic volume and the kinetic energy was calculated over the cardiac cycle and measured at end diastole.
Results: The mean age was 22 years (range 14-35). Mean ejection fraction was 57% in the Fontan patients and 67% in the healthy volunteers. 6 Fontan patients had an atrio-pulmonary Fontan connection and the remaining 6 had a total cavopulmonary connection.
Compared to healthy volunteers, Fontan patients had significantly reduced direct flow (28% vs 40%; $p = 0.018$) and increased residual volume (38% vs 28%; $p = 0.019$). This was less apparent in the 2 patients with a single ventricle and intact septum.
The kinetic energy was comparable in both groups (KE direct flow in Fontan $2.5 \times 10\text{-}4$ vs healthy volunteers 2.7×10^{-4}mJ; $p = 0.84$).
Conclusions: This study shows that ventricular 4D flow assessment is feasible in complex congenital heart disease.
Fontan patients show less efficient blood flow patterns than healthy volunteers. Unlike adults with cardiomyopathies, the Fontan patients had normal kinetic energy values. This may suggest that ventricles in the Fontan circulation are different from adults with ventricular dysfunction; the anatomical ventricular geometry may impact on the efficiency of blood flow but less on kinetic energy profiles. This new technique may allow novel insights into the pathophysiology of ventricular dysfunction in the Fontan circulation and may provide novel imaging biomarkers.

MP4-13
Use of a Semi-Automated Cardiac Segmentation Tool Improves Reproducibility and Speed of Segmentation of Contaminated MRA Datasets

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Introduction: Three-dimensional printing has increasing clinical applications in paediatric cardiology. Time required for segmentation and conversion of image data into a printable stereolithography (STL) file remains a significant limitation to widespread use.

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We investigated the impact of semi-automated cardiovascular-specific segmentation software on time and reproducibility of segmentation.
Methods: Magnetic resonance angiograms (MRAs) of 19 patients undergoing intervention for right ventricular outflow lesions were segmented to demonstrate the right heart. STLs were created by two independent clinicians using semi-automated cardiovascular segmentation (SAS; inPrint 1.0.0.156 Beta, Materialise) and traditional manual segmentation (MS; Mimics 18.0.0.524, Materialise). Time taken was recorded and geometric STL disagreement was determined (0% = exact overlap, 100% = complete disagreement). MRAs were categorised as clean when only right heart structures were present in the MRA, or contaminated when left heart structures were also present and required removal.
Results: 18 (7 clean and 11 contaminated) cases were successfully segmented with both methods. Time to STL for clean datasets was faster with MS than SAS (median 209 s (IQR 192-252) vs. 296 s (272-317), $p = 0.018$) while contaminated datasets were faster with SAS (455 s (384-561) vs. 866 s (310-1429), $p = 0.033$). MS yielded STLs that were always larger than SAS by $4.9 \pm 0.89\%$ (Fig. 1B&D). Interobserver STL geometric disagreement was significantly lower using SAS than MS overall ($0.70 \pm 1.15\%$ vs. $1.31 \pm 1.52\%$, $p = 0.030$), and for the contaminated subset ($0.81 \pm 1.08\%$ vs. $1.75 \pm 1.57\%$, $p = 0.036$, Fig. 1). However, there was no significant difference between techniques for clean datasets ($0.53 \pm 1.40\%$ vs. $0.60 \pm 1.36\%$, $p = 0.24$). Most geometric disagreement occurred at areas where left heart contamination was removed.

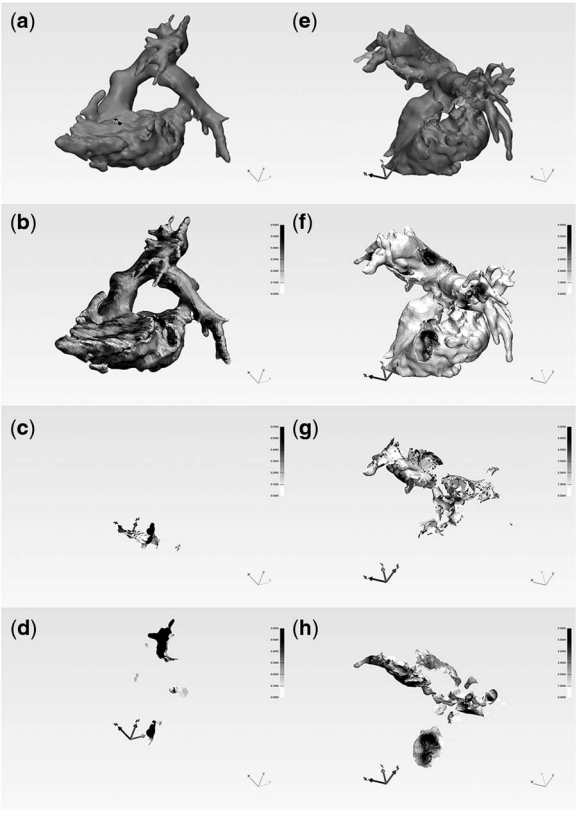


Figure 1.
A clean (a-d) and contaminated (e-h) dataset. a&e show the anatomy; b-d and f-h show the thickness of any geometric disagreement (if present) for SAS and MS (b&f), interobserver SAS (c&g), and interobserver MS (d&h), scale 0-0.6 mm for b-d, 0-6mm for f-h.

Conclusions: Semi-automated segmentation was faster and more reproducible for contaminated datasets, while manual segmentation was faster and equally reproducible for clean datasets. Semi-automated segmentation methods are preferable for contaminated datasets and continued refinement of these tools should be supported in conjunction with improved image acquisition methods to avoid arterial phase contamination.

MP4-14

Flow pattern and vascular distensibility of the pulmonary arteries in patients after repair of tetralogy of Fallot. Insights from 4D flow CMR

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Objectives: Pulmonary regurgitation is common after repair of tetralogy of Fallot (TOF). The regurgitant flow may lead to changes in flow profile, size and distensibility of the pulmonary arteries (Pas). We sought to assess Pa flow and distensibility in TOF patients (pts) by cardiac magnetic resonance (CMR) and to correlate them with flow patterns provided by 4D flow CMR.
Methods: 18 TOF pts (mean age 28 ± 11yrs, weight 63 ± 12 kg) and 9 controls (age 17 ± 7 yrs, weight 63 ± 24 kg) underwent CMR. 2D Phase-contrast (PC) images were acquired through-plane in the main (MPA), right (RPA) and left pulmonary artery (LPA). A 4D PC dataset was acquired covering all great arteries. Vessel areas and quantitative flow were measured on the 2D PC images. Flow patterns in Pas were qualitatively assessed for presence of helix or vortex on the reconstructed 4D images. Flow parameters, size and distensibility of the Pas were compared between TOF pts and controls and in the TOF group between RPA and LPA with regard to helix/vortex.
Results: In TOF pts, MPA mean regurgitant fraction (RF) was $25 \pm 17\%$. Compared to controls, both Pas were larger and distensibility was higher in LPA. RF was greater in LPA than in RPA (p 0.001) and LPA was larger than RPA (p 0.0342). Net flow was lower in LPA than in RPA (p 0.0005). Distensibility was similar in LPA and RPA and correlated with RF, regurgitant flow and minimum area.
Vortex was observed in LPA in 72% of TOF pts, but not in normals. Helix was present in 44% of pts and in 11% of normals and was correlated with higher distensibility (p 0.04). Presence of vortex was independent from any other parameter.
RPA presented helix in 77% of pts and in 55% of controls. Vortex was only detected in 11% of TOF pts.
Conclusion: After TOF repair, Pas size and distensibility are mainly determined by the amount of regurgitant flow and less by flow patterns, such as vortex or helix. Characteristic flow patterns are found in LPA and RPA, which seem to be more related to the geometry of the pas than to quantitative flow parameters.

MP4-15

Rigid body rotation is a robust and highly reproducible marker of cardiac dysfunction in children and young adults with heart disease

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Objective: Reversed left ventricular (LV) twist, so-called rigid body rotation (RBR), is a marker of cardiac dysfunction. Speckle tracking echocardiography (STE) is commonly used for the assessment of LV twist mechanics. Region of interest (ROI) width variability affects the reproducibility of strain measurements. The aim of this study was to investigate the effect of ROI width variability on the reproducibility of RBR patterns in children and young adults.
Methods: Our echocardiographic database was searched to identify all patients with a RBR pattern who underwent STE between January 2010 and August 2014. LV twist was assessed using STE. A predetermined protocol of ROI width variability was applied. Blinded review of the RBR pattern was performed independently by two investigators for all patients.
Results: A total of 374 STE datasets were reviewed. A RBR pattern was recognized in 21 patients (5.6%; mean age 11.2 ± 6.6; male 61.9%). Patients with a RBR pattern were diagnosed having dilated cardiomyopathy (n = 12, 57.1%), LV non-compaction (n = 6, 28.6%), hypertrophic cardiomyopathy (n = 2, 9.5%), restrictive cardiomyopathy (n = 1, 4.8%). RBR patterns included: reversed apical rotation (n = 12, 57.1%), reversed basal rotation (n = 7, 33.3%), and mirror-image rotation (n = 2, 9.6%). ROI width variability did not affect the RBR pattern in any of the patients, reproducibility was excellent. In addition, there was 100% agreement on the observed RBR pattern between investigators.
Conclusions: RBR is not affected by ROI width variability, suggesting that the presence of a reversed LV twist pattern is a robust marker of impaired cardiac function in children and young adults with heart disease.

MP4-16
Aortic arch anomalies associated with vascular ring: Clinical relevance of prenatal diagnosis
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Introduction: In the majority of cases aortic arch anomalies (AAA) associated with vascular rings are detected due to clinical symptoms resulting from tracheal compression. An increasing number of these anomalies are detected now by fetal echocardiography. The purpose of this study was to assess the incidence of postnatal airway obstruction in different types of prenatally detected AAA.
Methods: We analyzed all cases of prenatally detected AAA associated with vascular ring and compared findings of fetal echocardiography with postnatal echocardiography, MRI, CT-thorax and clinical symptoms.
Results: From 11/2010 –12/2015 14 cases of AAA associated with vascular ring were diagnosed by fetal echocardiography in our tertiary referral center. Only one patient had significant congenital heart disease (VSD). 3 patients had double aortic arch (DAA) with patency of both arches, while another 3 patients had DAA with atresia of the left arch between the left subclavian artery and left ductus arteriosus. The remaining 8 patients had less tight vascular rings formed by a right aortic arch (RAA), aberrant left subclavian artery (ALSA) originating from a left-sided diverticulum of Kommerell and a left sided ductus arteriosus. Chromosomal anomalies (trisomy 21) were present in 2/14 patients. The prenatal diagnosis of a vascular ring was confirmed in all cases by postnatal echocardiography, MRI (4 patients) or CT-thorax (3 patients). 4/14 patients (29%) developed tracheal obstruction and underwent surgery including one newborn and one infant with DAA, one infant with incomplete DAA and one infant with RAA, ALSA and left-sided ligamentum arteriosum. 10 patients (71%) remained asymptomatic without stridor or swallowing problems during a mean follow-up of 2.6 years.
Conclusions: Fetal echocardiography provides important new information on the clinical relevance and natural history of aortic arch anomalies associated with vascular ring: Patients with RAA, ALSA and left ductus arteriosus resulting in a loose vascular ring remained asymptomatic in the majority of cases during infancy and childhood. Although tracheal compression was present in half of the patients with DAA, 50% of these patients with complete vascular ring were asymptomatic. Since airway obstruction may develop later in life, patients with vascular ring should be offered long-term follow-up.

MP4-17
First Pass Perfusion Reserve Index in Paediatric Patients with Arterial Switch for Transposition of Great Arteries
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Background: Arterial switch operation is the treatment of choice for correction of transposition of the great arteries (TGA). Coronary anatomy is the most impacting early prognostic factor after surgery. We previously described potential mechanisms of late coronary complications. Data about late coronary outcome are lacking. Noninvasive first pass perfusion with cardiac magnetic resonance (cMRI) is sensitive technique for evaluation of myocardial perfusion in ischemic heart disease. We aimed to apply a comprehensive cMRI protocol to assess coronary artery complications in late follow up after this surgery .
Methods: Between 2010 and 2015, 66 patients were prospectively enrolled (median age 13.9 ± 5.5 years) into two groups according to the presence (9/66) or absence (57/66) of coronary arteries abnormalities at previous screening test at 5 ± 2 years. Coronary position and anatomy were analysed as previously described. Semi-quantitative evaluation of myocardial perfusion was performed by the analysis of myocardial First-Pass perfusion images at rest and during adenosine infusion and the segmental perfusion reserve index (PRI) was calculated. Late gadolinium enhancement (LGE) was studied.
Results: Eleven patients (11/66 = 16%) had perfusion defects (PRI < 1.5). In 9/11 perfusion defects were in anterior-anteroseptal or anterolateral segments vs 2/11 in inferior-inferoseptal-inferolateral segments. In 16 (16/66 = 24%) patients left coronary artery was reimplanted in clock position 11 or 12. These patients had more frequent perfusion defects than other patients (7 vs 2, $p = 0.004$). Moreover, considering only the myocardial segments irrigated by left coronary artery, semi-quantitative perfusion defects were found only in patients with left coronary artery in position 11 and 12 (7 vs 0, $p = 0.0002$). These patients had significantly lower PRI in myocardial segments irrigated by left coronary artery myocardial than other patients (2.0 ± 1 vs 2.7 ± 1, $p < 0.05$). LGE was positive in 2 cases. No patients with decreased PRI had LGE. All patients with decreased PRI were asymptomatic and did not show echocardiographic or functional signs of myocardial ischemia.
Conclusion: cMRI can provide a useful tool for detecting asymptomatic myocardial perfusion delay. PRI analysis results confirm the presence of a high-risk group of patients. Clinical implication of early detection of perfusion delay should be investigated.

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MP4-18

Cardiac valve annulus diameters in extremely preterm infants: a cross-sectional echocardiographic study

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Introduction: With the increasing incidence of births of preterm, very low birth weight infants, there is a demand for echocardiographic reference values of cardiac dimensions. The objective of this study is to provide reference values of cardiac valve annulus diameters in a cohort of extremely preterm very low birth weight neonates, and correlate these with patient characteristics.
Methods: Valve diameters of 376 infants were measured using two dimensional echocardiography. Correlations between valve diameters and patient characteristics (birth length, body surface area, birth weight, gestational age and sex) were assessed. Birth weight only was used to establish linear regression models. Inter- and intraobserver variability were assessed through intra class coefficient (ICC) analysis.
Results: Substantial variability was found for all valves (aortic valve mean (standard deviation; range): 5.0 mm (0.6; 3.7-6.5); pulmonic valve: 5.8 mm (0.8; 3.4-7.9); mitral valve: 8.0 mm (1.0; 5.5-10.5); tricuspid valve: 7.6 mm (1.2; 4.9-10.6)). Regression analysis showed moderate correlations between birth weight and valve diameter (R^2 aortic valve: 0.36; pulmonic valve: 0.20; mitral valve: 0.24; tricuspid valve: 0.24). Good intraobserver (ICC range 0.74-0.91) and interobserver agreement (ICC range 0.77-0.89) was found.
Conclusions: Our study provides ready-to-use reference values for cardiac valve annulus diameters for extremely preterm infants.

MP4-19

Ventricular Kinetic Energy in Young Fontan Patients

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Introduction: Four-dimensional (4D) flow magnetic resonance imaging (MRI) enables kinetic energy (KE) quantification of intraventricular blood flow. In this study we aimed to quantify the KE in in patients with univentricular heart and to assess the change in the KE after different types of interventions.
Methods: 4D flow MRI was acquired in patients with Fontan circulation (n = 12; median age 12, range 3-29 years) and in healthy volunteers (n = 8; median age 26, range 23-36 years). MRI was repeated after transcatheter embolization of significant aortopulmonary collaterals (APC; n = 1), after stenting of left pulmonary artery (n = 1) and after surgical replacement of hepatic flow tunnel with a Y graft due to significant central pulmonary artery stenosis with secondary formation of arteriovenous (AV) fistulas in the right lung (n = 1). Intraventricular KE was calculated throughout the cardiac cycle and indexed to stroke volume (SV).
Results: The systole/diastole ratio of KE in Fontan patients was similar to the ratio of the controls' left ventricle (LV) or right ventricle (RV) depending on the ventricular morphology (Cohen´s kappa = 1.0). Peak systolic KE/SV did not differ in patients compared to the LV in controls (0.016 ± 0.006 mJ/ml vs 0.020 ± 0.004 mJ/ml, $p = 0.09$). Peak diastolic KE/SV in Fontan patients was lower than in the LV of the control group (0.028 ± 0.010 vs 0.057 ± 0.011 mJ/ml, $p < 0.0001$). In the patient with intrapulmonary AV fistulas, the KE curve had two diastolic peaks before intervention, but after intervention the early diastolic peak was higher and more fused with the late diastolic peak. The patient with APC had a fused diastolic curve with a plateau before intervention. After intervention the curve showed two diastolic peaks. As expected, no change in KE curve was observed after LPA stenting.
Conclusions: In patients with univentricular heart, KE is dependent on the morphology of the ventricle and is decreased compared to controls. The results suggests that KE could be used for patient follow up, since KE might reflect early disruptions in the circulation.

MP4-20

The CURE: Quantification of left ventricular dyssynchrony in Ebstein´s Anomaly by cardiovascular magnetic resonance feature tracking and 4 dimensional volume analysis

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Introduction: Cardiovascular magnetic resonance (CMR) imaging of Ebstein's Anomaly (EA) suggests the presence of LV dyssynchrony of the basal septum. Feasibility of LV dyssynchrony assessment has recently been shown using CMR feature tracking (CMR-FT) derived circumferential and radial uniformity ratio (CURE and RURE) and 4D volume analysis derived Systolic Dyssychrony Index (SDI).
We sought to quantify intra-ventricular LV dyssynchrony in EA patients using CMR-FT and 4D volume analysis.
Methods: 31 EA patients and 31 matched controls were included. CMR-FT was performed on short axis SSFP cine stacks, whilst 4D-volume analysis utilized long and short axis orientations employing dedicated software (Diogenes and 4D-LV-Analysis, TomTec, Unterschleißheim, Germany). CURE, RURE and 4D-SDI as measures of dyssynchrony were calculated. QRS duration, BNP and Total R/L-Volume Index (R/L Index) were registered. CMR dyssynchrony parameters were evaluated for correlations with heart failure parameters by Spearman´s analysis.
Results: EA patients (mean age 26.3 years, controls 23.7 years) had significantly longer QRS duration (QRS 119 ms ± 32 (EA) vs. 97 ms ± 14 (controls, $p < 0.01$)). CMR derived measures of dyssynchrony were altered as follows: (4D-SDI $7.60\% \pm 4.58$ (EA) vs. $2.54\% \pm 0.62$ (controls, $p < 0.001$)), (RURE 0.72 ± 0.09 (EA) vs. 0.79 ± 0.06 (controls, $p < 0.001$)), (CURE 0.77 ± 0.05 (EA) vs. 0.86 ± 0.03 (controls, $p < 0.001$)), (ratio RURE:CURE 0.75 ± 0.05 (EA) vs. 0.83 ± 0.04 (controls, $p < 0.001$)).
Spearman's analysis revealed significant correlations of CURE with BNP (Spearman's Rho (SR)-0.508, $p = 0.005$) and R/L Index (SR -0.473, $p = 0.008$), 4D-SDI with R/L Index (SR 0.518, $p = 0.006$) and a trend towards a correlation with BNP (SR 0.363, $p = 0.068$). 4D-SDI and CURE correlated significantly with QRS duration (SR for SDI 0.588, $p = 0.001$; for CURE -0.425, $p = 0.017$).
Conclusions: EA patients exhibit pronounced intra-ventricular dyssynchrony of the LV as compared to a control group. Amongst

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the different dyssynchrony indexes CURE shows the strongest association with QRS duration and heart failure (BNP) and disease severity parameters (R/L Index). These novel markers can easily be quantified from routine CMR studies and may have a role in the assessment of deterioration of cardiac function, carrying potential value for management of EA.

P1-1

Analysis of Ventricular Tachycardia in Children with Structurally Normal Heart: Six Years Experience From Turkey

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Objective: The aim of our study was to review the clinical characteristics and outcomes of otherwise healthy children with ventricular tachycardia (VT), in our clinic.
Methods: A single-center retrospective review of patients with VT between January 2010 and December 2015 was undertaken. Patient work-up include presenting symptoms, 12-lead ECG, ambulatory ECG recording, exercise testing, echocardiography to rule out structural heart disease and electrophysiologic study. Patients with underlying structural or functional heart disease, inherited arrhythmias or with significant systemic illness were excluded.
Results: A total of 72 patients [40 M (55%), 32 F (45%)] were eligible for inclusion. The mean patient age was 11.6 ± 9.5 years (range: 8 days - 18.6 years), and the mean patient weight was 48.5 ± 30.4 kg (range: 3.5-95 kg).The most frequent initial complaint was palpitation in 38 cases (53%), syncope in 5 (6%), chest pain in 5 (6%), and one (1%) had fetal arrhythmia as well. In addition, one infant (1%) was diagnosed with the complaint of prolonged crying. All patients were evaluated with echocardiographic examination and 10 patients with magnetic resonance imaging. 12 lead ECG and 3-channel Holter ECG monitoring were performed in all patients. In addition, exercise testing and 12 lead Holter ECG monitoring were performed in 46 (63%) and 12(16%) patients, respectively.
Patients were grouped according to the origin of VT as follows; right ventricular tachycardia (right ventricle outflow tract(40) and para-Hisian(1) n = 41), left ventricular tachycardia (left ventricle posterior fascicular(13), anterior fascicular(1), coronary cusp (10), and purkinje fiber(2); n = 26). In 5 patients location of VT was not determined.
Antiarrhythmic treatment was instituted in 62 cases-1 (86%). Electrophysiologic study and ablation were performed in 40(55%) of 72 patients which were symptomatic and had frequent PVCs and tachycardia. One patient underwent cryoablation and others had radiofrequency ablation as well. The mean follow-up period was 22.9 ± 0.5 months (range:1 months–6 years).
Conclusion: Management and prognosis of idiopathic ventricular tachycardia in patients with an anatomically normal heart, differs from ventricular tachycardia associated with structural heart disease. Ablation should be kept in mind as a curative treatment option in these patients.

P1-2

The evaluation of rhythm and conduction disturbances in congenitally corrected transposition of the great arteries

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Background: In this study, we aimed to evaluate the rhythm problems and management of patients with congenitally corrected transposition of the great arteries (ccTGA).
Patients and Methods: The records of 93 ccTGA patients were revieweved retrospectively between 2009-2015 and 53 patients with regular ECG, ECHO, Holter monitorization and clinic follow-up data were found eligible for the study.
Results: 22 female, 31 male patients were included in the study. The median age at admission was 4.5 years (ranging 7 days to 47 years). Out of 53 patients, 13 (25%) were isolated ccTGA. Atrioventricular (AV) conduction problem in 19 (35%) of the patients, atrial arrhythmia in 12 (22%) of the patients were detected at initial ECG, ECHO and HOLTER evaluations. There was initially 7 (13%) patients who had pacemaker implanted due to complete AV block. No ventricular arrhytmia was detected (Table I). The median follow-up of patients was 34 months (ranging 7 months to 6 years). During follow-up, pacemaker was implanted to 2 patients with isolated ccTGA and 2 patients with postoperative AV block operated due to associated lesions. Electrophysiologic study and ablation was performed to 3 patients with Wolff-Parkinson-White and supraventricular tachycardia. Patients are still on clinical follow-up.
Conclusion: As there is an increase risk of spontaneous atrioventricular block and atrial arrhytmias in cTGA patients, 24 hours Holter monitorization should be a part of routine follow up of these patients even with normal preceding reports. Early diagnosis and treatment will decrease morbidity and mortality.

Table 1. The initial ECG and Holter findings of patients

The initial ECG and Holter findings of patients	*n = 53*
Normal	*24*
Conduction Abnormalities	*19*
First Degree Atrioventricular block	*4*
2:1 Second Degree Atrioventricular block	*3*
Complete Atrioventricular block	*3*
Bundle Branch Block	*2*
Pacemaker	*7*
Supraventricular arrhythmia	*14*
Wolff-Parkinson-White	*2*
Ectopic Atrial Rhythm	*6*
Premature atrial contaction	*3*
Focal atrial tachycardia	*3*
Ventricular arrhythmia	*0*
⋆*Total number of rhythm disorders*	*33*

⋆There were more than one rhythm problems in some of the patients.

P1-3

Cryoablation with an 8-mm-tip Catheter for Right-Sided Accessory Pathways in Children

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Introduction: Cryoablation is increasingly utilized in children because of its safety profile. Recently, larger-catheter tips have been more widely used to improve long-term success rates. The aim of this study was to assess the safety and efficacy of 8-mm-tip catheters for cryoablation of right-sided accessory pathways (APs)

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in children. Methods: Electrophysiological procedures were performed using the EnSite™ system (St. Jude Medical Inc., St. Paul, MN, USA). Results: Between July 2010 and July 2014, 54 patients (mean age: 13.1 ± 3.7 years) underwent cryoablation using an 8-mm-tip catheter. In 18/54 (33%) patients where an 8-mm-tip catheter was the first-choice catheter, the acute success rate was 18/18 (100%). There was a history of previous failed attempts or recurrence with radiofrequency ablation and/or 6-mm-tip cryoablation in 36/54 (67%) patients. The acute success rate in these patients was 24/36 (67%). No fluoroscopy was used in 34/54 procedures. The recurrence rate was 6/42 (14%) during a mean follow-up period of 32 ± 15 months. In 1 patient, transient atrioventricular block occurred. Conclusions: Cryoablation with an 8-mm-tip catheter for right-sided APs in children who weight over 40 kg appears to be safe and acutely effective in cases where conventional ablation methods fail and also as a first choice for ablation procedure. However, the recurrence rate still seems to be high.

P1-4
Cardiac Nodal and Cardiac Autonomic Functions in Children with Vasovagal Syncope

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Introduction: Vasovagal syncope (VVS) mainly exists as a result of autonomic imbalance in the patients. In the present study, cardiac nodal (sinoatrial node and atrioventricular node) and cardiac autonomic functions have been investigated in the children with VVS. Thus the effect of the existing autonomic status on the characteristics of cardiac impulse conduction has been demonstrated in the children with VVS.
Methods: The study included 51 pediatric patients (the mean age 14.01 ± 2.79 years, range 7 to 18 years; 30 females) who had been evaluated for syncope, and who existed with normal investigations of ECG, Holter, exercise testing, and echocardiography. All patients underwent head-up tilt testing (HUTT), and their parameters of heart rate variability (HRV) (SDNN, SDANN, SDANNi, r MSSD, p NN50, HF,LF,VLF) were evaluated by the analysis of 24-hour Holter testing. Corrected sinus node recovery time (CNRT), and Wenckebach point (WP) were measured in all patients by the transoesophageal atrial stimulation. The patients were grouped, and investigated according to the results of HUTT.
Results: All patients existed with normal cardiac nodal functions. In the patient group existing with positive HUTT results, the HRV parameters (HF, r MSSD, p NN50), which are the indicators of parasympathetic effect, were determined to be high. CNRT did not differ significantly between the patient groups existing with positive and negative HUTT results. However, WP was found to be higher in the patient group with positive HUTT results.
Conclusion: When the parameters of HRV are investigated in the children with VVS, effect of parasympathetic tonus has been demonstrated to increase. Impairment of the cardiac nodal functions may not be expected in the children with VVS. However, the WP may be prolonged in these patients, due to increased autonomic tonus in favour of parasympathetic activity.

P1-5
Modified Cryoablation Versus Radiofrequency Ablation of Atrioventricular Nodal Reentrant Tachycardia (AVNRT) in Children: Results of a Prospective Randomized Study

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Introduction: Both cryoablation and radiofrequency (RF) ablation are effective treatments for AVNRT. Because of the small risk of permanent atrioventricular (AV) block in RF ablation and higher recurrence rates of cryoablation there is still some uncertainty about which treatment to choose. In this report we compare RF ablation with a modified cryoablation technique.
Methods: Between January 2012 and June 2014 a total of 94 patients diagnosed with AVNRT (64 female) were treated with ablation. Patients were grouped randomly as cryoablation (n = 53) or RF (n = 41). All the procedures were performed by the same specialist. Acute success rates, complication rates and recurrence at follow up are compared between the two groups.
Results: Patients' average age and weight in cryo group were 13 ± 3.4 years and 48.3 ± 14 kg and in RF group 11.6 ± 3.1 years and 47.3 ± 17 kg respectively. While applying cryoablation, a line consisting of 5-6 lesions was put adjacent to the successful site closest to the compact AV node. There was no significant difference in acute success rates between the two groups during the process (in the cryo group, 98.1%, in the RFA group 97.5%). RF ablation group had a shorter total processing time, but according to the cryoablation group fluoroscopy time was significantly longer ($p < 0.05$). During cryomap or energy, temporary complete AV block was observed in four patients and 1st degree AV block in six patients. Mean follow up time after the procedure was 2.31 ± 0.82 years. Relapse rates were 7.6% in cryo group (4/52) and 5% in RF group (2/40). No permanent AV block or complications were observed in both groups.
Conclusions: According to our work in AVNRT in children, cryoablation therapy is as effective as RFA method for acute procedural success and recurrence rates. Being less fluoroscopy exposure in cryoablation procedures is an important advantage. Compared with the literature, these more successful results can be depend on modified ablation technique we used.

	CRYO	RF	p	ALL
N	53	41	-	94
F/M	36/17	28/13	-	64/30
Mean Age (years)	12.98 ± 3.41	11.63 ± 3.14	0.06	12.39 ± 3.34 (3-18)
Mean Weight (kg)	48.3 ± 14	47.3 ± 17	0.8	48.5
Procedure Duration (min)	116 ± 45	76 ± 28	< 0.05	98.86 ± 43.69
Fluoroscopy Time (min)	13.1 ± 7.8	19 ± 10	< 0.05	15.76 ± 9.73
Acute Success	52/53 (98.1%)	40/41 (97.5%)	0.6	92/94
Follow up Time (years)	2.40 ± 0.89 (1.25-3.75)	2.19 ± 0.70 (1.3-3.75)	0.5	2.31 ± 0.82
Recurrence	4/52 (7.6%)	2/40 (5%)	0.4	6/92
Permanent AV Block	0	0	-	0

P1-6
Characteristics and Follow Up of Children with Isolated Congenital Heart Block

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Introduction: Congenital heart block (CHB) is a rare disorder that may be associated with mortality and morbidity. It has an incidence of about 1 in 22 000 live births. Many times, no clear etiology is determined for isolated CHB. In this study we documented the characteristics of our isolated CHB patients with pacemakers (PM).

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Methods: The study was limited to patients with isolated, complete, permanent block. The medical records of children aged between 0-18 years who underwent PM implantation for CHB were systematically reviewed. Patients with any other systemic disease which may affect growth were excluded while analyzing growth parameters.
Results: Between January 2000-January 2015 a total of 63 (F/M: 31/32) patients were diagnosed as CHB. Only 5 of them (%7.9) were born from mothers with systemic lupus. The other 58 patients' mean age at diagnosis is 6.32 ± 3.78 (1-16) years. Atrioventricular block was asymptomatic in 40 (69%) patients and was diagnosed after the detection of bradycardia or murmur by chance. Five patients had chest pain, four patients had syncope and three patients had convulsions. In physical examination heart rate was 52.5 ± 5.8/min. In holter examination minimum, mean and maximum heart rates were 34.2 ± 4.7/min, 50.8 ± 8.7/min and 98.5 ± 29.3/min respectively. The mean interval between diagnosis of AV block and pacemaker implantation was 1.07 ± 0.3 years. During a mean follow-up of 8.49 ± 2.4 years (6-15 years), no patient died or developed dilated cardiomyopathy.
We also evaluated 28 patients with full information for weight and height percentiles before PM implantation and after follow up. First percentile values for weight and height were 32.5 ± 28; 46 ± 29 and the last percentile values for weight and height were 45.8 ± 33 ($p > 0.05$); 57.2 ± 30 ($p > 0.05$) respectively.
Conclusions: It is important to recognize an asymptomatic complete AV block in the pediatric population. PM implantation is safe and the only treatment for these patients. Effective cardiac output is important for growth. Although not statistically significant, we can say that PM implantation supports growth in patients with CHB.

P1-7
Endocardial VVIR Pacing in Infants and Small Children Weighing 8 ≤ Kg: A Single Center Experience

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Introduction: VVIR is the most common pacing mode in small children. The overall higher complications rate of epicardial leads has led, in some units especially in critical patients when rapid surgery is not feasible, to the implantation of endocardial systems even in very small infants.
The aim of this work is to review our experience with endocardial pacemaker implantations in infants weighing ≤8 kg.
Patients and Methods: We reviewed 43 patients (28 males (65.1%) and 15 females (34.9%)) weighing ≤8 kg who underwent endocardial VVIR pacemaker implantations from January 2008 to May 2015. Clinical evaluation before and after pacemaker implantation was performed, at 8-14 days, 3, and 6 months then every six months. Doppler evaluation of the subclavian vein was performed. Venography of the subclavian vein was done for patients after at least one year of implantation
Results: The mean age at implantation was10.6 ± 6.7 months (20 days to 30 months), and the mean weight was 6.12 ± 1.7 (3 to 8) kg. 14 patients weighed ≤5Kg. 23 patients had congenital complete heart block (CHB) while 18 patients had postoperative CHB. Two patients had sick sinus syndrome with pauses and seizures.
Two patients had a pacemaker implanted and PDA device closure at the same setting.
At the time of implantation, the mean ventricular pacing threshold was 0.66 V, mean pacing amplitude 1.82 V and mean lead impedance of 630 Ω. At follow up (27.1 ± 17.6 months), patients had a mean ventricular pacing threshold was 0.58 V, mean pacing amplitude 1.48 V and mean lead impedance of 630 Ω.
No complications occurred during implantation. One patient required lead repositioning after 24 hours due to loss of capture and excessive stimulation threshold.
None of the patients had clinical signs or symptoms of vein occlusion, and among the 32 patients who underwent Doppler examination of subclavian vein, none had an alteration of blood flow. Among the 24 patients who had venography done, only one patient showed narrowing of the subclavian vein but without flow obstruction.
Conclusions: Endocardial single chamber permanent pacemaker insertion is feasible in children weighing ≤8 kg.

P1-8
Asymptomatic Wolf-Parkinson-White Syndrome Presenting With Atrial Fibrillation/Flutter And Life Threatening Arrhythmia Or Cardiac Arrest In Fetuses And Young Children

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Introduction: Wolf-Parkinson-White syndrome (WPW) is a common cause of re-entry arrhythmia in children. The estimated incidence is 1-3:1000. There is a well-known risk of rapid conduction of atrial fibrillation through the accessory pathway resulting in ventricular fibrillation and sudden cardiac death. Such arrhythmia combination is said to be uncommon in asymptomatic and young children. Our case serious however showed that atrial fibrillation resulting in rapid ventricular conduction and sudden cardiac collapse can occur in asymptomatic young children with WPW syndrome.
Method: Retrospective review of cases presenting with WPW syndrome and pre excited atrial fibrillation leading to hemodynamic compromise and aborted cardiac arrest.
Results: 150 patients were diagnosed with WPW syndrome between 1990 and 2015 at the University Hospital of Wales. Eight patients presented with life threatening arrhythmia as the first event without any prior documented event; two had aborted sudden cardiac death and all eight required urgent DC cardioversion. One of the children, who had aborted sudden cardiac death at age 5.5 years, was known to have asymptomatic preexcitation previously. In all, four had atrial fibrillation with rapid ventricular conduction, and four had fetal and neonatal atrial flutter and rapid ventricular conduction. Four patients had successful ablation of their accessory pathways and were discharged from follow up. Three of the four patients with fetal atrial flutter developed AVRT immediately after DC cardioversion of flutter and all exhibited preexcitation on their ECG.
Conclusion: Atrial fibrillation or atrial flutter leading to rapid ventricular conduction and life threatening arrhythmias can be the first manifestation of WPW syndrome from fetal life through early childhood and adolescence. The significance of asymptomatic preexcitation needs to be re-evaluated in larger prospective multicentre studies.

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Patient	Sex	Age at presentation	Presentation	Location of AP	First presentation
Case 1	Male	15 year	AFib/VF, preexcitation	Right anterolateral	Yes
Case 2	Male	5.5 year	AFib/VF/ Aborted sudden cardiac death, preexcitation	Left coronary sinus	Yes
Case 3	Female	15 year	AFib/VF, preexcitation	Left mid-septal	Yes
Case 4	Male	15 year	AFib/VF/ Aborted sudden cardiac death, preexcitation	Left coronary sinus diverticulum	Yes
Case 5	Male	Fetal and 1 day of age	Fetal atrial flutter hydrops, postnatal atrial flutter and AVRT, preexcitation	NA	Yes
Case 6	Male	Fetal and 1 day of age	Fetal atrial flutter hydrops, postnatal atrial flutter and AVRT, preexcitation	NA	Yes
Case 7	Male	Fetal and 1 day of age	Fetal atrial flutter hydrops, postnatal atrial flutter and AVRT, preexcitation	NA	Yes
Case 8	Male	Fetal and 1 day of age	Fetal atrial flutter hydrops, postnatal atrial flutter and asymptomatic preexcitation	NA	Yes

	Shortest pre-excited RR in AFib/ AP-ERP (msecs)	Prior treatment	EP success	Outcome
Case 1	160/300	None	Success	Alive
Case 2	180/250	None	Success	Alive
Case 3	260/260	None	Success	Alive
Case 4	180/240	None	Success	Alive
Case 5	200/NA	Flecainide and digoxin in fetal life and postnatally	N/A	Alive
Case 6	200/NA	Flecainide and digoxin in fetal life and postnatally	N/A	Alive
Case 7	180/NA	Flecainide and digoxin in fetal life and postnatally	N/A	Alive
Case 8	200/NA	Flecainide and digoxin in fetal life. Has asymptomatic preexcitation after birth on no medication	Awaiting EPS	Alive

P1-9
Presenting Features And Clinical Outcomes Of Children Requiring DC Cardioversion In South Wales

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Introduction: DC cardioversion is a drastic intervention in childhood and causes great anxiety in many clinicians to use it despite being part of paediatric life support training programs. A recent survey amongst paediatric trainees reported that 56% felt they were not confident in defibrillation. There is no detailed study looking at its use and technical-clinical issues encountered in day to day practice in children. We aimed at investigating the presenting features and clinical outcomes of children requiring DC cardioversion in South Wales.

Methods: We reviewed all children who had received DC cardioversion at the University Hospital of Wales (UHW) in the past 20 years.

Results: 30 children were identified. Most children (19 = 62%) requiring cardioversion were under 1 year of age. 14 children presented to UHW, 10 transferred from other hospitals as an emergency due to reluctance to perform cardioversion locally. Six were admitted electively for DC cardioversion. 10 children (33%) were unwell for longer than 24 hours prior to presentation, mostly with nonspecific symptoms of lethargy and poor feeding. 13 (43%) required emergency intubation at presentation. Only two children of this cohort was shocked out of hospital by paramedics. Seven neonates had an antenatal diagnosis of supraventricular tachycardia (SVT) and three didn't. Only eight children (27%) had congenital heart disease with Ebstein's anomaly being the most common diagnosis and only two of these presented as an emergency. Further three children were later diagnosed with dilated cardiomyopathy. Seven children had WPW syndrome and one child had long QT syndrome. Cardiac function around the time of cardioversion was impaired in 17 children (56%). 9 (30%) children required cardioversion for ventricular tachycardia or fibrillation, 21 (70%) for SVT. In 18 children sinus rhythm was restored after 1 shock (64%), children presenting in VF required 3 or more shocks. 28 children (93%) survived and 2 died.

Conclusions: DC cardioversion in the treatment of acute SVT in children is rarely required. Majority of DC cardioversions were performed by cardiologists at a tertiary centre as local specialists felt uncomfortable with the procedure. There is a need to improve understanding of DC cardioversion procedure among paediatricians.

P1-10
Catheter ablation of supraventricular tachycardia in children using the Ensite NavX System with limited fluoroscopic approach; Two years of experience in a new center

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Aim: This study is aimed at presenting the outcomes of children with supraventricular tachycardia(SVT) treated with ablation and electrophysiological therapies(EPT) within the last 2 years.

Methods: Patients who were treated with radiofrequency ablation/ cryoablation(RFA/CRA) and EPT in the Electrophysiology Laboratory of Pediatric Cardiology Clinics, Istanbul Mehmet Akif Ersoy Chest and Cardiovascular Surgery Training and Research Hospital, Istanbul, Turkey, between November 2013 and November 2015 were included in this study.

Results: A total of 221 ablation procedures were applied on 214 patients(116 males and 98 females). The average age and weight of patients were found to be 12.9 ± 4.0years and 48.4 ± 16.8 kg, respectively. Only RFA, only CRA, and both RFA and CRA in the same session were applied in 93(42%), 112(51%), and 16(7%) of the procedures, respectively. Some patients had Ebstein abnormality(four patients), hypertrophic cardiomyopathy(one patient), and TGA+ VSD+ PS(one patient), for which Rastelli operation was performed. Two patients developed cardiomyopathy due to tachycardia. When the substrates of arrhythmia were checked(some of the patients had more than 1 substrate), 80 of them had atrioventricular nodal reciprocating tachycardia

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(AVNRT), 56 had concealed accessory pathway (AP, 14 were right sided and 42 were left sided), 79 had manifest AP, and 19 had focal atrial tachycardia (FAT) (5 were left sided and 14 were right sided). Rapid success was achieved in 202 patients (94%). The distribution of unsuccessful arrhythmia substrates was as follows: nine patients had AP (five were right and four were left sided) and three patients had FAT (two were right and one was left sided). Relapses were observed in seven patients (3%) during the follow-up of 11.0 ± 7.5 months. Among these patients, five were diagnosed with AP and two with AVNRT. Second interventions were successful in all relapse cases. The total duration of the procedures and fluoroscopy was 170.3 ± 66.7 and 3.6 ± 6.9 min, respectively. Fluoroscopy was never used in 144 procedures (67%). Transient atrioventricular (AV) block in two patients and permanent incomplete right bundle branch block in one patient were observed.
Conclusion: The RFA/CRA procedure can be used effectively and with very high rates of success in the ablation treatment of SVTs in children without any complication, and with or without any need for fluoroscopy because of the advantage provided by Ensite NavX System.

P1-11
Catheter ablation of idiopathic premature ventricular contraction/ventricular tachycardia in children using Ensite NavX System with limited fluoroscopic exposure: Two years of experience in a new center

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Aim: This study aimed at presenting the outcomes of patients with idiopathic ventricular tachycardia (IVT)/premature ventricular contraction (PVC) treated with ablation and electrophysiological therapies (EPT) within last 2 years.
Patients and Methods: Patients who were treated with radiofrequency ablation (RFA) and EPT in the Electrophysiology Laboratory of Pediatric Cardiology Clinic, Mehmet Akif Ersoy Chest and Cardiovascular Surgery Training and Research Hospital, Istanbul, Turkey, between November 2013 and November 2015 were included in this study. The procedures were applied with the accompaniment of a three-dimensional mapping system(TDMS), and by using minimal fluoroscopy.
Results: A total of 26 ablation procedures (22VT and 2PVC) were applied on 24patients (16 males and 8 females). The average age and weight of patients during the procedures were 13.6 ± 4.2years and 50.5 ± 16.4 kg, respectively. The RFA therapy (irrigated RFA for two patients and classical RFA for the remaining) was used in all procedures. The distribution of arrhythmia substrates was as follows: 6(25%) had right ventricular outflow tract (RVOT) of which 2 were posterolateral, 1 each were mid-anterior, anterolateral, posterior, and posteroseptal; 10(42%) had left VOT of which 5 were right coronary cusp, 4 were left coronary cusp, and 1 was non-coronary cusp; and 8(33%) had left posterior fascicular. Rapid success was achieved in 23 patients (96%). Ablation failed in one patient(epicardial). Relapses were observed in three patients (12%) during the follow-up of 9.7 ± 5.3months. Post-ablation of the RVOT area, relapses were observed in two patients. In second interventions, successful coronary cusp ablations were performed by focusing on VT in the same two patients. In another patient, VT arising from posterior fascicular Purkinje fibers relapsed. The total duration of the procedures and fluoroscopy was 189.3 ± 69.0 min and 3.9 ± 4.9 min, respectively. While the Ensite NavX mapping system was used in all procedures, fluoroscopy was never used in 11 procedures (42%). The duration of applied fluoroscopy was 6.8 ± 4.6 min. No complication was observed during the procedures.
Conclusion: The RFA therapy can be used effectively and with very high success rates in the ablation treatment of IVT/PVC in children without any complication and with or without any need for fluoroscopy because of the advantage provided by TDMS. If necessary, the coronary cusp area can be evaluated to reduce the relapse rate in patients with RVOT area ablation.

P1-12
Computerized interpretation of electrocardiograms poses a risk of false-negative diagnosis of long QT syndrome or WPW syndrome

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Introduction: Computerized interpretation of electrocardiograms (ECGs) is widely used in clinical setting, especially when manual reading by cardiologists is not available. However, little is known whether computerized interpretation makes a correct diagnosis of pediatric life-threatening arrhythmias like long QT syndrome or WPW syndrome.
Objectives: To clarify the rate of false negative diagnoses in a large number of ECGs using the school-based ECG screening program.
Patients and Methods: Patients were the first and seventh graders in Kagoshima City from 2005 to 2014. ECGs were interpreted using an ECG analysis program (Ver. S1 or S2, Fukuda Denshi, Inc.). Normal ECGs evaluated by computerized interpretation are re-evaluated by pediatric cardiologists, and abnormal ECGs are picked up and referred to the 2nd examination.
Results: Computerized interpretations were performed on ECGs from 52 831 first graders and 57 246 seventh graders. The ECGs of 43 279 first graders (82%) and 46 926 seventh graders (82%) were evaluated as normal. Pediatric cardiologists re-evaluated as abnormal in 24 ECGs of first graders (9 IRBBB and others) and 68 ECGs of seventh graders (21 suspected long QT syndrome, 21 abnormal ST-T findings, 6 arrhythmias, 4 WPW syndrome, and others). One first grader (1/24; 4%) and 18 seventh graders (18/64; 26%) were eventually diagnosed as having cardiac diseases after the 2nd and 3rd examinations (first graders, ASD; seventh graders, 8 with long QT syndrome, 4 with WPW syndrome, 3 with PVC, 1 with second-degree atrio-ventricular block, and 2 with others).
Discussion: Computerized interpretation was found to have inadequacies in analyzing the small or gently-sloping waves on ECGs. The false negative rate of computerized interpretation was low (0.04% in the first graders and 0.1% in the seventh graders). Nevertheless, re-evaluation is important, especially in the seventh graders, because it can identify patients who have a high risk of cardiac events.
Conclusions: Computerized interpretation of ECG poses a risk of false-negative diagnosis of long QT syndrome or WPW syndrome. In normal ECGs evaluated by computerized interpretation, it is important to re-evaluate ECGs carefully for the findings of long QT syndrome or WPW syndrome.

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P1-13

Comparison of two algorithms in the prediction of the accessory pathway localization in children with Wolff-Parkinson-White

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Introduction: Several algorithms have been developed to predict the localization of the accessory pathways (AP) in patients with Wolff-Parkinson-White (WPW).
Boersma et al. developed an algorithm specifically designed for the pediatric population.
AIM: We aim to compare the accuracy of predicting the localization of the AP using the adult algorithm according to d'Avila et al. and the pediatric algorithm by Boersma. Both algorithms are based on QRS polarity.
Method: We present our single center experience of children with WPW. The data were collected retrospectively in a database from April 2007 to November 2015. Patients aged ≤18 years with documentation of pre-excitation on a 12 lead resting ECG and with eminent prove of an accessory pathway on electrophysiological study (EPS) were included. Patients with concealed pathway were excluded. The ECG was analyzed blinded from EPS results. The primary outcome concerns an exact match between the predicted localization and site of ablation, but because both algorithms describe different categorization for AP localization, an agreement was made on which localizations could be accepted as a match in a secondary analysis. Since Boersma's algorithm points out multiple possible localizations, a match was accepted if any one of these sites corresponded with localization on EPS.
Results: Inclusion criteria were met for 36 patients. Sixty-nine percent of the patients were boys. Median age was 13 years (4-18 years). The algorithm by Boersma provided an exact match in 29 patients (80%) while the one by d'Avila only provided an exact match in 24 patients (66%). If we expand our match as previously described, the accuracy of the algorithm by d'Avila augments up to 75%.
Conclusion: The algorithm by Boersma provided a more correct, though less detailed localization of the AP in 80% of our patients. Using the more detailed adult algorithm by d'Avila this result was almost equaled on condition that we accepted some previously agreed sites as a match.

P1-14

Cardiac repolarization parameters in childhood obesity

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Studies in adults demonstrated an association of obesity and prolongation of cardiac repolarization with the potential for an increased risk of ventricular arrhythmia and sudden cardiac death. The aim of this study was to evaluate potential alterations in cardiac repolarization in obese children.
Methods: 215 consecutive healthy pediatric volunteers, who participated in the LIFE Child Study at the LIFE Leipzig Research Center for Civilization Diseases from 2011 to 2014 were prospectively enrolled and their ECG's were analyzed.
Results: There is a significant prolongation of absolute QT and corrected QT interval ($p = 0.005$), T peak-to-end ($p = < 0.001$) and QT dispersion ($p = 0.013$) in obese compared to lean children. Female patients showed overall longer QT intervals than male patients ($p = 0.002$). Despite significant differences in estradiol levels in male and female probands ($p = 0.001$) there was only an influence of estradiol on absolute QT interval ($p = 0.013$) but not on the corrected QTc interval ($p = 0.7$).
Conclusion: Childhood obesity causes a prolongation of QT and QTc interval with a more evident effect in female patients. Furthermore, an increase in cardiac electrical heterogenity could be demonstrated with rising BMI.

P1-15

ECG changes that indicate disturbances in repolarization periods in Epilepsy patients

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Abstract: Background: Both abnormal lengthening and shortening of the corrected QT interval (QTc) on the electrocardiogram indicating substantial disturbance of autonomic function in epilepsy have been reported.
Aim: We hypothesized that children with epilepsy and abnormal EEG comparative to healthy children may have ECG abnormalities in the corrected QT (QTc) interval that can indicate chanellopathy disturbances in the re-polarization period that is more pronounced during acceleration of heart rate.
Methods: Rest ECG and Holter ECG were done for 75 children that were devised into three groups: Group I: 25 children with epilepsy and normal EEG. Group II: 25 children with Epilepsy and abnormal EEG. Group III: 25 healthy children. ECG data from healthy children, children with epilepsy and normal electroencephalogram (EEG), and children with epilepsy and abnormal EEG were prospectively reviewed in rest ECG and during maximal heart rate in acceleration of rhythm in Holter ECG traces. Corrected QT interval (QTc), and QTC dispersion were assessed in 25 healthy children, 25 children with epilepsy and normal EEG, and 25 children with epilepsy and abnormal EEG.
Results: In children with epilepsy and normal EEG the mean QTc was 0.426 mesc, and the mean QTc dispersion was 0.076 mesc. In the group of children with epilepsy and abnormal EEG the QTc 0.448 was and QTc dispersion was 0.152. In the healthy children, the control group, mean QTc was 0.372 mesc, and the mean QTc dispersion was 0.003 mesc.
Conclusion: Our data suggest a significant substantial disturbance of autonomic function in patients with epilepsy. QTc and QTc dispersion were found to be significantly prolonged in children with epilepsy, especially in patients with abnormal EEG.

P1-16

Conduction property in left nodal extension to the atrioventricular node

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Introduction: It has been speculated that another slow pathway with different conduction property might be involved in the mechanism of developing some form of atrioventricular nodal reentrant tachycardia (AVNRT). Left nodal extension (LNE) is considered to work as another possible slow pathway, but there is limited information about the conduction property of LNE. A previous study (Gonzalez et al. Circulation 2002) reported that pacing at distal coronary sinus (CS) induced left atrial input to the atrioventricular node (AVN), but the study failed to examine the input in patients with dual AVN physiology. In the present study,

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we tried to reveal the characteristics of LNE conduction with the same method.
Method: Children who received radiofrequency catheter ablation for the treatment of supraventricular tachycardia were enrolled in the study. Cases after receiving ablation in the septal area were excluded. Constant (A1, 6 to 8 beats) and following extrastimulus pacing (A2) from the right atrial appendage (RAA) were conducted during sinus rhythm without ventricular preexcitation. When sudden increase of AH interval after 10 ms reduction in the A1A2 interval was obtained (AH jump), CS distal pacing was started with the same protocol. CS pacing was repeated until AH jump was obtained at the same A1A2 interval. AH and HV intervals during constant pacing (FP-AH and FP-HV) and those obtained at the time of AH jump (SP-AH and SP-HV) were compared between the two protocols.
Results: A total of 10 children with dual AVN physiology were included in the study. During constant pacing, CS pacing induced significantly shorter FP-AH than RAA pacing (74.2 ± 14.1 ms vs 89.1 ± 20.0 ms, $P = 0.03$). CS pacing also significantly shortened SP-AH (236.7 ± 84.5 ms vs 260.0 ± 82.7 ms, $P < 0.01$). Meanwhile, CS pacing did not alter FP-HV and SP-HV.
Conclusion: Shortened FP-AH and SP-AH during CS pacing indicated different fast and slow inputs to the AVN from RAA pacing. SP-AH during CS pacing might associate with LNE conduction, which is consistent with anatomical studies demonstrating that the length of LNE is shorter than that of posterior nodal extension.

P1-17
Nonfluoroscopic mapping for supraventricular arrhythmia ablation in pediatric patients. Initial results in a prospective study

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Introduction: The current standard of care for imaging during supraventricular tachycardia (SVT) ablation uses fluoroscopy, which exposes otherwise healthy children to the potential harmful effects of radiation.
Objctive: The purpose of this study was to determine whether the adjunct use of nonfluoroscopic imaging systems reduces radiation exposure during SVT ablation among children.
Methods: This was a prospective, single-center study that included consecutive patients aged with less than 18 years-old with SVT and normal cardiac anatomy referred to catheter ablation. Patients were randomized to control (fluoroscopy only) or study group (fluoroscopy + NavX electroanatomic mapping). The same operator performed all procedures. Fluoroscopic and procedure times (minutes) were registered and radiation doses (mSv) were measured by two dosimeters positioned on the anterior and posterior chest wall.
Results: Twenty six patients were enrolled (12 control group, 14 study group). Median age was 11.5 years-old (Q1: 8.1, Q3: 15.3); 21(80.7%) had accessory pathways, 4(15.4)% had AVNRT and 1 (3.8%) Atrial Tachycardia. The study group presented shorter fluoroscopic time (155 ± 94 vs 86 ± 58 seconds, $p = 0.03$). No difference was observed on x-ray exposure measured on anterior [Study: 0.4mSv (Q1:0.1, Q3: 0.85) vs Control: 0.25mSv (Q1: 0, Q3: 0.65); $P = 0.42$] and posterior [Study: 0.6mSv (Q1: 0.15 Q3 2.65) vs Control: 0.9mSv (Q1: 0.225 Q3: 2.175); $P = 0.79$] dosimeters. Procedure time was longer on the study group (61 ± 33 min vs 114 ± 52 min; $p = 0.006$). Acute success was obtained in 11 (91.7%) procedures on control group and in 12(85.7%) in the study group ($P = 1$), with no adverse events. Conclusion: Electroanatomic nonfluoroscopic imaging systems during SVT ablation in children resulted in a reduction in fluoroscopic time, without reduction in radiation, measured by dosimeters. Conversely, an increase in the procedure time was observed.

P1-18
Amiodarone-associated thyroid function abnormalities in children with arrhythmias

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It is well-known that amiodarone being one of the most effective antiarrhythmic drugs causes different extracardiac effects, particularly, changes of thyroid function that are disclosed in 15-20% of patients. The aim of the study was to assess the thyroid hormone levels during prolonged amiodarone therapy in 0–7-year-old children with arrhythmias.
Materials and Methods: A total of 42 patients with WPW syndrome ($n = 19$), atrial tachycardias ($n = 16$), and ventricular tachycardias ($n = 7$) received amiodarone therapy. The duration of amiodarone administration varied from 0.7 to 24 months (Me 6.00; IQR 1.83-9.00). The thyroid hormone levels were estimated at the following time points: before treatment, during the amiodarone treatment at least 3 weeks after the beginning of therapy, and 6 months after amiodarone discontinuation.
Results: The levels of thyroid-stimulating hormone (TSH), total thyroxine, free thyroxine, thyroglobulin increased during amiodarone therapy. The increases in the total and free thyroxine levels were statistically significant ($p = 0.043$ and $p = 0.037$, correspondingly) whereas TSH and thyroglobulin increased insignificantly during amiodarone therapy. All these indices significantly decreased 6 months after discontinuation of amiodarone therapy ($p = 0.006$ and $p = 0.036$ correspondingly). Comparison of the initial values with the tests results 6 months after amiodarone discontinuation did not show any statistically significant differences. No clinical sings of hypertyrosinemia were observed in our patients during amiodarone treatment. The most significant changes in the levels of thyroid hormones were found in infants. During the entire course of amiodarone treatment, the median values of thyroid hormones did not exceed the upper reference ranges even when statistically significant changes in hormonal status occurred.
Conclusions: Amiodarone-associated changes in thyroid status were reversible and thyroid hormone values normalized 6 months after discontinuation of therapy.
Key words: tachyarrhythmias, children, amiodarone, thyropathy.

P1-19
Results of RFA of tachyarrhythmias in small children

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The use of radiofrequency ablation (RFA) for the management of supraventricular tachycardia (SVT) in infants and small children remains controversial. Although SVT in the majority of small

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children can be managed medically, and SVT in infants can spontaneously resolve within 18 months, RFA is occasionally necessary for patients with drug-refractory tachycardias complicated by hemodynamic compromise. 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 26 patients aged 4.2 ± 3.9 months. Wolff-Parkinson-White syndrome and atrial tachycardia were detected in 15 (57.7%) and 11(42.3%) patients, respectively. Patients with structural heart pathology, including congenital heart diseases and laboratory-confirmed myocarditis, were excluded from the study.
Results: 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). 4 patients (15.4%) had tachycardia recurrence during first 2 months after RFA. Later all these patients were performed repeated RFA successfully. The total RFA success rate, considering recurrences and retreatment, was 100%. Only one patient had major complication – damage of mitral valve leaflet that accompanied by mitral regurgitation. Later it became a reason for mitral valve plasty. 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 4 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. Taking into account technical aspects of catheter interventions in small children, RFA of tachyarrhythmias should be performed in specialized clinics with relevant experience.
Key words: supraventricular tachycardia, infants, radiofrequency ablation.

P1-20

Heart rate variability in Long QT syndrome in children - beware the genotype!

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Introduction: The congenital Long QT Syndrome (LQTS) is an inherited life-threatening disease caused by mutations in genes encoding cardiac ion channels. In several LQTS genotypes, arrhythmia is associated with sudden increased sympathetic activity. The fact that an increase in sympathetic activity often triggers a cardiac event and that beta-adrenergic blocking and left cardiac denervation are effective treatments, makes it evident that the sympathetic nervous system is involved in this disease. The sympathetic-parasympathetic interaction can be studied by analysis of heart rate variability (HRV). The aim of this study was to evaluate HRV and the influence of medication in children/adolescents with long QT syndrome.
Methods: Twenty-four hour ambulatory electrocardiographic recordings from year 2000 to 2013 in 80 children with LQTS were retrospectively reviewed. HRV power spectrum analysis was performed to determine the total power (Ptot, 0.003-0.50 Hz) and the power in the low-frequency (LF: 0.04-0.15 Hz) and high-frequency (HF: 0.15-0.50 Hz) regions as well as the PLF/PHF ratio. The control group consisted of 39 healthy children. Data are presented as mean ± SE of log-transformed values.
Results: The LQTS patients off beta-blocker therapy (n = 28) presented with statistically significantly lower Ptot (3.54 ± 0.05, P = 0.02) than controls (3.66 ± 0.04). However, when analyzing subgroups of genotypes, no significant differences were found between LQT1/LQT2 patients and controls. In contrast, LQT3 patients showed a lower Ptot (3.06 ± 0.09, P < 0.001) but a higher LF/HF ratio (0.47 ± 0.08, P < 0.001) than controls (0.08 ± 0.05). Double-mutations (Jervell and Lange Nielsen syndrome, JLNS) also differed from controls: JLNS patients had a lower LF/HF ratio (-0.37 ± 0.01, P < 0.001) and a lower LF than controls (2.80 ± 0.11 vs. 3.10 ± 0.04, P = 0.01). In 18 patients that were examined both off and on beta-blocker therapy, treatment with beta-blockers resulted in a significantly reduction of the LF/HF ratio with 16% (0.07 in log-transformed units, P = 0.01).
Conclusions: This study indicates that impact on HRV in LQTS patients seems to be genotype dependent. Furthermore, beta-blockade reduces LF/HF in LQTS patients, indicating a reduced sympathetic activity acting as a protection against arrhythmias. HRV analysis in Holter recording might become a useful tool for risk assessment and treatment strategy in LQTS patients.

P1-21

A Unique case of Timothy Syndrome due to coexistence of CACN1C and Ank2 mutations

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Introduction: Timothy Syndrome is a very rare and fatal type of ion channel disease in which dysmorphic features are together with rhythm abnormalities. Mutations of CACNA1C gene are reported in the etiology. Ankyrine2 mutation is found in LongQT4 Syndrome. We present a case of Timothy Syndrome with two different mutations of CACNA1 and Ank2 genes.
Case: Six-day-old boy was consulted for bradycardia. He had respiratory distress requiring positive airway pressure support starting at the first day of life. On the 4th day intermittant bardycardia of 52/minutes was noticed which could increase up to 120/minutes. Birth weight was 2600gr. And low for gestational age. Physical examination revealed heart rate as 65/minute. blood pressure 80/40 mmHg, arterial oxygen saturation 94%. Syndactyly of fingers 3 through 5 of left hand and 4 and 5 of right hand were found. He had bald head, round face, flat nasal bridge and thin upper lip (fig. 1-2). His lower limbs were in plaster cast because of pes equinovarus, syndactyly of the toes could not be observed. Electrocardiogram showed a heart rate of 60/minutes, 2:1 atrioventricular block and prolonged QT interval (QTc = 720 msn) (figure 3). Echocardiography revealed atrial septal defect, patent ductus arteriozus and pulmonary hypertension (right ventricular pressure was estimated as 78 mmHg using the tricuspid regurgitation jet velocity). The parents were consanguineous, history of sudden cardiac death, arrhythmia or LQTS were absent in the family. Propranolol and inhaled iloprost treatments were started. Implantation of an ICD system was planned. However, within the following day the patient had sudden cardiac arrest and did not respond to resuscitation. Genetic analysis revealed compound heterozygous c.4418 C > G (p.Ala1473Gly) mutation in exon 38 of CACNA1C gene and c.11791 G > A (p.Glu3931Lys) mutation

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in exon 45 of ANK2 gene. Both parents were negative for CACNA1C mutation and mother was carrier for Ank2 mutation.
Conclusion: This is the first report with unique coexistence of CACNA1 and Ank2 mutations in Timothy phenotype causing fatal outcome.



Figure-1 Figure-2 Figure-3

P1-22

SCN5A mutations: special ventricular lead capture features in children?

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Unstable and changing lead capture threshold are a recognized difficulty when implanting cardiac devices in adult with loss-of-function SCN5A mutation.
We aim to study our pediatric cohort.
Case#1: 7y boy. PM at 3y due to a febrile cardiogenic syncope and detection of complete heart block not detected previously.First Flecainide test did not show ECG changes.VVIR implantation with oscillating capture during the implantation.Final ventricular capture:1.5 V at 0.31ms, but during follow-up capture was increased until 2.7 mV at 0.31ms. ICD implantation at 7y due to syncope and an atrial flutter.Mutation in SCN5A c.659 C > T identified and type-1 Brugada ECG under flecainide. Both final atrial and ventricular capture:1.25 V at 1.0 ms.
Case#2: 8y girl. PM at 3y due to complete heart block. According to loss-of-function SCN5A double mutation, electrophysiological test and clinical features, Lev-Lenegre syndrome was diagnosed. VVIR implanted with with oscillating ventricular capture during implantation and follow-up.
Case#3: 11y girl. Prenatal echocardiograms showed foetal tachycardia accompanied of foetal hydrops. During first years, fascicular tachycardia was diagnosed and controlled with flecainide. Despite of the ECG improvement, complete heart block was alternating with fascicular tachycardia. Pacemaker implantation Endurity ™ SR mode VVI, at 9 years old, due to bradycardia symptoms, was performed. During pacemaker implantation newly capture issue was detected, with better initial compared to final procedure poor or absent stimulation despite of high voltages within the apex; stimulation improvement was detected within right ventricular outflow tract and then we obtained better capture. Final ventricular capture: 0.5 V at 0.4ms. During 1 year follow-up, no issues were detected.
Genetic features: de novo SCN5A c.4783 G > A, missense.
Conclusions: As well as data published for adult patients, undulated capture thresholds are also an important issue in paediatric patients with loss-of-function SCN5A mutation carrying a pacemaker or implantable cardioverter defibrillator (ICD).
Genetic testing is a useful tool not just for the clinical management or familial segregation, but also it is an outstanding tool for the electrophysiologist.
Recognition of this clinical entity may help to understand the electrophysiological behaviour of SCN5A-related diseases and planning pre-implantation and follow-up strategies.

P1-23

From population ECG screening to molecular diagnosis of channelopathies: preliminary experience in pediatrics

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Introduction: There is a lack of data regarding efficacy of cardiovascular (CV) screening programs including ECG when applied in young children, for early detection of unrecognized genetic conditions associated with high risk for sudden cardiac death (SCD). We evaluated the performance of a CV screening program, including 12-lead ECG recording, when applied in selected populations of Mediterranean children.
Methods: A CV screening program for primary schools, approved by Minstry of Health, was applied in a sample of primary schools of a Mediterranean island, in geographical isolated areas with reported increased SCD incidence. Participation was voluntary following parental consent. After completion of a standarized history questionnaire by parents, children underwent clinical evaluation and 12-lead ECG recording, at local health stations. A stepwise referral pattern was established, including pediatric cardiology evaluation and molecular DNA confirmatory testing, whenever the possibility of inherited arrhythmogenic CV disease was increased.
Results: 220 primary school children, (84 male, 116 female), median age 11,4 (range 7.5-12yrs) have been evaluated during two years (2014-15). 22 children (10%) had an indication for further diagnostic or lifestyle modification for CV risk factors, including ECG abnormalities (n = 9), abnormal heart auscultation (n = 7) and adiposity/hypetension (n = 6). ECG abnormalities included WPW (n = 1), VES (n = 2), probable LVH (n = 2), and QTc prolongation (n = 2) both boys, with QTc 475 and QTc 490. Children with ECG abnormalities underwent further evaluation including ambulatory ECG monitoring and regular follow up. Family ECG screening was positive in one child with prolonged QTc (490ms, wide T wave), regarding his father (QTc = 460) and one sister (QTc = 490). Further molecular DNA testing was negative in the first child, while it revealed a novel KCNH2 heterozygous mutation (NP_000229.1:p.Ser606Tyr) in child and affected family members in the second case. In silico analysis using Polyphen-2 and SIFT suggested that the Ser606Tyr mutation might be harmful, and the family was advised to receive b-blocker prophylactic treatment.
Conclusions: A stepwise approach from ECG population screening to molecular diagnostics can detect and genetically characterize subclinical cases of inherited CV disease, associated with arrhythmogenic SCD also in pediatrics.

P1-24

Cardiovascular Features of Marfan Syndrome at Children in North-East Region of the Romania - 10 years study

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Introduction: Marfan Syndrome is a spectrum disorder caused by a heritable genetic defect of connective tissue that has an autosomal dominant mode of transmission. The defect itself has been isolated to the FBN1 gene on chromosome 15, which codes for the connective tissue protein fibrillin-1. Marfan syndrome affects most organs and tissues, especially the skeleton, lungs, eyes, heart, and large blood vessel. The prevalence of Marfan syndrome is estimated at 1/5000.

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Methods: Prospective clinical and echocardiographic evaluation of consecutively recruited children at North-Eastern Region of Romania diagnosed with Marfan syndrome.
Results: We have analysed the prevalence of cardiovascular abnormality in 58 children with Marfan syndrome recorded in the files of Medical Genetic Center Iasi, Romania in the last 10 years (January 2006 –December 2015). There were 30 girls and 28 boys diagnosed at median age of 12,4 years (range from 4 years to 18 years old).
The diagnosis was based on anamnesis, the presence of the characteristic features, EKG, thoracic X-Ray, echocardiography, established Z score, ophthalmological and orthopedic exam.
Cardiac abnormalities were present in 82.75% of cases and they are represented by: mitral valve prolaps (65.51%), aortic route dilation (46.55%), mitral regurgitation (63.8%), tricuspid valve prolaps (3.4%), dilatation of pulmonary artery (1.7%). Aortic dissection did not appear. Aortic route surgery was needed for 5.17% cases.
Conclusions: Cardiovascular manifestation of Marfan syndrome remain among the central issues in diagnosis and management. Regular monitoring of valvular function and aortic diameter, early initiation of long-term β-adrenergic blockade and elective repair of a moderately regurgitant mitral valve or of a moderately aortic root dilatation represent the standards of care.

P1-25
Clumping factor A is a membrane receptor for secreted von Willebrand factor-binding protein mediating Staphylococcus aureus adhesion to vascular endothelium

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Staphylococcus aureus (S. aureus) is a frequent pathogen causing life-threatening endovascular infections such as infective endocarditis. To establish an endovascular infection, S. aureus needs to bind to vascular von Willebrand factor (VWF) to overcome shear forces imposed by flowing blood. We previously described that secreted staphylococcal von Willebrand factor-binding protein (vWbp) interacts with VWF, enabling flow-controlled bacterial adhesion to endothelial cells (ECs) and to subendothelial matrix. However, the receptor to which vWbp anchors to the bacterial cell wall is unknown so far. Several surface proteins of S. aureus are linked to the bacterial cell wall by sortase A (SrtA). A mutation in the srtA gene leads to an anchoring defect of about 20 S. aureus surface proteins. We hypothesized that vWbp is able to interact with a staphylococcal surface protein, thus mediating the adhesion of S. aureus to ECs and to subendothelial matrix via VWF.
We measured adhesion of S. aureus Newman (WT) and mutants deficient in SrtA or in SrtA-dependent surface proteins to VWF, vWbp and ECs. Fluorescently labeled bacteria were perfused over a glass surface coated with VWF, vWbp or ECs in a micro-parallel flow chamber. We verified our findings using Lactococcus lactis (L. lactis) bacteria expressing single staphylococcal surface proteins. In vivo adhesion was evaluated in the murine mesenteric circulation using real-time intravital vascular microscopy.
First we quantified the adhesion to coated vWbp of several S. aureus strains deficient in individual surface proteins or deficient in SrtA. Compared to the WT strain, the SrtA deficient strain and the strain deficient in Clumping factor A (ClfA) showed a decreased adhesion to vWbp. The absence of ClfA also reduced the adhesion of S. aureus to VWF under flow, to activated ECs and to the activated murine vessel wall in vivo. Selective overexpression of ClfA in the membrane of L. lactis enabled the bacteria to bind to coated vWbp under flow.
We conclude that vWbp interacts both with sheared VWF and with the SrtA-dependent staphylococcal surface protein ClfA. The ternary complex formed by endothelial VWF, secreted vWbp and bacterial ClfA causally mediates adhesion of S. aureus to the vascular endothelium.

P1-26
Exome Sequencing and Linkage Analysis as Tools in solving Syndromic Cardiopathies in Small Families

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Introduction: We present three small families with syndromic cardiopathies diagnosed in two children of each family. The parents are non-consanguineous and have a normal phenotype. In one family with three children one sibling is unaffected. An autosomal recessive hypothesis is most likely, as these are very rare and unique phenotypes.
Methods: Analysis using linkage analysis and exome sequencing was performed. Genomewide parametric linkage analysis was performed, SNP typing platform was used in a recessive model. Genotyping was done in parents and both the unaffected and affected siblings. Data analysis was done using commercial and in-house developed software. Only variants in genes from the linkage regions were retained. All homozygous calls were excluded in the parents and the unaffected sibling, reference calls were excluded in the affected siblings. Only exonic and splicing variants were included, synonymous variants were excluded. Variants occurring with a frequency of $< 1\%$ in the 1000 genomes project or with an unknown frequency were included.
Results: After variant filtering, candidate genes are identified in the linkage regions with homozygous mutations in the patients, inherited from both parents, and for which the unaffected sibling is heterozygous or reference. Results of this analysis will be presented and will be confirmed by Sanger sequencing.
Conclusions: Reaching a genetic diagnosis in rare disorders remains a challenge. Sophisticated genetic tools can be combined to aid in finding causal mutations in small families.

P1-27
Novel method to quantify bacterial adhesion to endovascular surfaces in vitro

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Introduction: *Staphylococcus aureus*, *Staphylococcus epidermidis* and *Streptococcus sanguis* are frequent causes of native-valve infective endocarditis (IE). In addition, *S. aureus* is also implicated in about 50% of prosthetic-valve IE cases. In this work we present a novel *in vitro* method to quantify bacterial adhesion to heterologous bovine pericardium patch (PCP) used in congenital heart defects repair, in both static and shear stress conditions.
Methods: Tissue pieces (10 mm diameter) prepared as for clinical use were mounted in a 6-well plate, and incubated for 1 h at 37°C with 10^7 CFU/mL of *S. aureus* Cowan, *S. epidermidis* ATCC 149,900 and *S. sanguis* NCTC 7864 (labelled with carboxy-fluorescein) for static adhesion. Similar tissue pieces, bacteria and bacterial inocula were used to study bacterial adherence under laminar shear stress of 10 dyne/cm^2 in a newly developed flow chamber. After incubation, bacterial adhesion was confirmed using the fluorescence microscope IN Cell Analyzer 2000 (GE Helathcare) and the tissue pieces were sonicated in 1 mL of 0.9% NaCl for bacterial detachment and quantification by CFU count on blood agar plates (expressed as Mean Log CFU/mL ± SD).
Results: Using the fluorescence system IN Cell Analyzer 2000 it was possible to visualize bacterial attachment to PCP tissue surface. The different bacterial species showed similar adhesion capacity to the PCP tissue in static conditions, presenting in average 3.79 ± 0.10 Log CFU/mL ($P > 0.05$, one-way ANOVA). Additionally, shear stress significantly increased bacterial adhesion (average of 5.02 ± 0.47 Log CFU/mL; $P < 0.05$, one-way ANOVA) while there was no difference among bacterial species adherence ($P > 0.05$, one-way ANOVA).
Conclusions: The IN Cell Analyzer 2000 system demonstrated to be the most efficient fluorescence method to visualize bacterial adhesion to PCP surface, overcoming the PCP auto-fluorescence issue due to glutaraldehyde crosslinking using other fluorescence microscopy techniques. In this work we were able to establish a novel and reliable method to quantify in vitro bacterial adhesion to endovascular surfaces. Although the bacterial species tested present different adhesion mechanisms and propensity to colonize these surfaces, in our results they presented similar capacity to attach to PCP tissue. In addition, shear forces increased bacterial adhesion.

P1-28
FOXC1 as a potential gene responsible for rapidly progressing aortic valve dysplasia in a child with 6p25.6 microdeletion

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Introduction: Microdeletion 6p25.3 is a rare but well-known entity. The phenotype includes predominantly dysmorphic features, neuro-developmental and ocular anomalies. Cardiac anomalies are less frequently seen and poorly explained.
Case report: A 4year old girl presented to our department for rapidly evolving aortic stenosis, requiring surgery. This little girl was born at term but abnormal nuchal translucency had been observed during pregnancy. Prenatal caryotype was normal. At the age of 4 months she required surgery for a vallecular cyst. She subsequently was noted to have significant abnormalities of oculomotricity combined with dysmorphic features and mild neurological delay. Echocardiography, performed in the first year of life showed a dysplastic but functional tri-leaflet aortic valve. At the age of 14 months, array-CGH revealed a 1.4 Mb microdeletion in 6p25.6, which includes 9 genes (from EXOC2 to FOXC1). At the age of 4 years cardiac follow-up revealed worsening of the aortic stenosis. Six months later, the stenosis had significantly progressed and required surgical valve replacement. Pre-operative CTscan showed in addition a hypoplastic left internal carotid artery. The aortic arch was normal. During surgery the aortic valve was replaced by an aortic homograft. The post-operative course was complicated by respiratory failure requiring ECMO but the child eventually left the hospital in good condition after 2 months. Examination of mRNA expression revealed that among the 9 deleted genes of the 1.4 Mb microdeletion, only EXOC2 and FOXC1 are expressed in human aortic valve. Furthermore, quantitative reverse PCR experiments showed that FOXC1 expression was significantly reduced in the excised aortic valve tissue compared to control.
Discussion: FOXC1 is known to be important for the development of several organs. Mutations of FOXC1 in humans are associated with ocular and brain anomalies. Cardiac anomalies have been described but the exact role of FOXC1 in human cardiogenesis remains unclear. Analysis of Foxc1 and Foxc2 mutant mice has demonstrated that these transcription factors are both required for cardiac development. Our case report suggests a potential role of FOXC1 in aortic valve and aortic arch (carotid artery) development. Therefore, FOXC1 could play a role in normal cono-truncal development.

P1-29
Elevated C-Reactive Protein Level After Heart Transplantation in Paediatric Recipients: A Predictor of Development of Coronary Artery Disease?

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Purpose: We sought to clarify whether a persistent increase of the CRP level could be considered a marker for Coronary Arteries Vasculopathy (CAV) development in a group of pediatric heart recipients.
One hypothesis for the etiology of CAV implicates early endothelial inflammatory activation within the microvasculature.
Methods: Twenty-eight patients younger than 18 years who underwent heart transplantation between January 2010 and January 2014 were investigated with coronary angiography and intravascular ultrasound (IVUS) on a yearly basis. Triple drug immunosuppressive treatment was used. Statins were used at physician's discretion without knowledge of serum concentrations of C-reactive protein. C-reactive protein level was considered abnormal if greater than 0.05 mg/dl .Coronary artery disease was defined as any decrease in coronary vessel luminal diameter and classified as mild (I), moderate (II-III) or severe (IV)using Stanford Scale classification too. We collected data also on donor age.
Results: There were 12 females and 16 males. The median age at HTX was 69 months (range 7-176), the median graft ischemic time was 255 minutes (range 162-362). The median donor's age was 10 yrs (range 2-36). A diagnosis of congenital heart disease was made in 9 pts (32%) and of cardiomyopathy in 19(68%). Eight patients (29%) were on Statin therapy. The CRP level, as measured at the time of first catheterization study, was higher than 0,05 mg/dl in 15 pts (54%). These values remained persistently elevated during follow-up.
By the Stanford scale, CAV of degree I was found in 11 (49%) pts, II in 7 (25%), III in 4 (14%) and IV in 6 (21%). We did not observe a statistically significant correlation between CAV degree and CRP level, use of statins nor donor's age. We only observed a borderline association (p = 0.05) between older age of donors and higher postoperative CRP in recipients.

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Conclusion: In this small group of pediatric heart recipients, we have not identified an association of high CRP level and presence or severity of CAV; nevertheless, we found that recipients of grafts from older donors had a higher CRP level. This observation suggests that serial levels of CRP may be useful in defining high-risk patients for CAV development.

P1-30

Prophylactic use of human umbilical cord blood derived mesenchymal stem cells transfusion in monocrotaline-induced pulmonary artery hypertension rats

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Background: Pulmonary artery hypertension (PAH) is a life threatening disease but it shows less response to traditional therapeutic regimens. Stem cells have the ability to secrete paracrine factors, attenuate pathological vascular remodeling and promote regeneration. Mesenchymal stem cells can be a promising resource for the treatment of refractory diseases such as PAH. However, prophylactic use of human umbilical cord blood derived mesenchymal stem cells (hUCB-MSCs) in PAH models are still controversial. Therefore, we investigated the differential effects such as hemodynamics, pathologic finding and gene expressions of hUCB-MSCs according to transfusion timing.
Methods: 6-week Sprague Dawley rats were used in this research. The experimental groups were divided into the control group, the monocrotaline (M) group (60 mg/kg) and the treatment groups which were subdivided into two groups, the UA group (hUCB-MSCs were administered 1 day after MCT injection); the UB group (hUCB-MSCs are administered 1 week after MCT injection). Hemodynamics, pathological changes and protein expressions were investigated.
Results: Mean right ventricular pressure (RVP) was significantly reduced in all U groups compared with the M group at weeks 2 and 4. RV/RV+ S ratio was significantly decreased in the UA and UB groups compared with the M group at week 1. RV/RV+ S ratio was decreased in the UB group compared UA group at week 4. In aspect of pathological changes, medial wall thickness and the number of intra-acinar arteries were significantly improved in two treatment groups at week 4. The protein expressions of endothelin-1, B cell leukemia lymphoma-2 and vascular endothelial growth factor in the lung tissues were significantly decreased in the UA and UB group at week 4. Collagen I and III protein expressions in the heart tissues were significantly decreased in the UB group compared with UA group.
Conclusion: A prophylactic injection of hUCB-MSCs was not as effective as a treatment injection after the occurrence of PAH in MCT rats.

P1-31

The Effect of Sildenafil on Right Ventricular Failure in Monocrotaline-Induced Rat

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Background: Pulmonary arterial hypertension (PAH) leads to right ventricular failure (RVF) as well as an increase of pulmonary vascular resistance. Our purpose was to investigate the hemodynamic effects as well as the cellular and molecular impact of sildenafil on RVF in monocrtaline (MCT)-induced rat models.
Subjects and Methods: The rats were separated into 3 groups. The control (C) group, the monocrotaline (M) group (MCT 60 mg/kg) and the sildenafil (S) group (MCT 60 mg/kg + sildenafil 30 mg/kg/day for 28 days). Masson Trichrome staining was used to measure the collagen content in the heart tissues. Western blot analysis and immunohistochemical staining were performed.
Results: The mean right ventricular pressure (RVP) was significantly decreased in the S group at weeks 1, 2 and 4. The number of intra-acinar arteries and the medial wall thickness of the pulmonary arterioles significantly decreased in the S group at week 4. The collagen content also decreased in the S group at week 4. The expressions of Bcl-2-associated X (Bax), caspase-3, B cell lymphoma-2 (Bcl-2), interleukin-6 (IL-6), matrix metalloproteinase (MMP)-2, endothelial nitric oxide synthase (eNOS), endothelin (ET)-1 and ET receptor A (ERA) in immunohistochemical staining greatly reduced in the S group at week 4. In western blot analysis, protein expressions of troponin I and brain natriuretic peptide (BNP), caspase-3, Bcl-2, TNF-α, IL-6, MMP-2, eNOS, ET-1, ERA in the heart tissues were greatly diminished in the S group at week 4.
Conclusion: Sildenafil improved right ventricular hypertrophy, mean RVP, lung and heart pathology and gene expressions. There also was improvement in RVF.

P1-32

Novel Cardiac troponin C(TNNC1) variant causing familial dilated cardiomyopathy

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Introduction: Cardiac troponin C (cTnC), expressed by the gene TNNC1 is the calcium sensing component of the cardiac troponin complex. Disease causing mutations in TNNC1 have been described in some families with dilated cardiomyopathy(DCM). We describe a novel DCM causing mutation in TNNC1 in which aspartic acid at position 151 is replaced by valine (D151V).
Methods: We studied a large 3 generation Asian family with a strong pedigree suggestive of autosomal dominant DCM with 7 affected individuals of which 3 died between ages 13 and 35 years. Complete exon sequencing for 77 known cardiac genes was performed and all identified variants were validated by Sanger sequencing.
Results: We identified a novel missense mutation in the C-terminus calcium binding site of cTnC, resulting in a p.D151V amino acid substitution. The mutation co-segregates with all screened clinically affected family members and is absent in a control population of 300 healthy individuals. Furthermore phylogenetic analysis shows that aspartic acid-151 of cTnC to be highly conserved across all Metazoa.
Conclusion: We describe a novel disease causing mutation at a highly conserved residue in a family with dilated cardiomyopathy. Given its location, we postulate its effect relates to the modulation of calcium binding to cTnC.

P1-33

High values of NT-proBNP reflects various right-heart dysfunctions in patients with tetralogy of Fallot after repair

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Background: Values of NT-proBNP in patients with tetralogy of Fallot (TOF) after definitive repair is said to elevate reacting

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volume overload of right ventricle (RV) followed by pulmonary valve regurgitation (PR) but not reacting pressure overload of RV followed by stenosis of RV outflow tract. We predicted NT-proBNP values would rise highly because of diverse secretory stimulation as well as expanded end-diastolic volume of RV (RVEDVI). We investigated what kind of strain affected high NT-proBNP (hNT-proBNP) in repaired TOF patients. Methods. The medical records of 101 repaired-TOF patients from 1 to 53 years were reviewed. We performed cardiac catheterization to grasp hemodynamic status between 2010 and 2015. We defined hNT-proBNP as NT-proBNP 400 pg/ml or over with which patients were potentially in state of heart failure (Japan heart failure society). Cardiac performances were determined which affected hNT-proBNP. Results. In monovariate analysis hNT-proBNP was significantly associated with elevated pressure on right-heart, such as pulmonary capillary wedge (≥ 14 mmHg), end-diastolic ventricle (≥ 14 mmHg), and end-systolic ventricle (pressure of RV/LV ≥ 0.79). Similarly, hNT-proBNP was significantly associated with expanded volume on right-heart, such as end-diastolic ventricle (≥ 164 ml/m^2), and end-systolic ventricle (≥ 109 ml/m2). Additionally, hNT-proBNP was related to reduced ejection fraction of RV (≤ 31%) and increased pulmonary vascular-resistance (≥ 3.1 units/m^2). Cardiac performances on left-heart had no relation to hNT-proBNP. After multivariate analysis hNT-proBNP was independently associated with odds ratio of 11.2 for end-systolic volume of RV, 3.9 for end-systolic pressure of RV, and 3.6 for end-diastolic pressure of RV. Clinically, repaired-TOF patients with hNT-proBNP had some form of symptom more before examinations than those without hNT-proBNP had (45% vs. 17%, p = 0.0068). Discussion. Our study showed hNT-proBNP also had relation to diverse RV overload other than expansion of end-diastolic RV in repaired TOF patients. Increased pulmonary resistance and pressure of pulmonary capillary wedge were associated with hNT-proBNP. We could point out RV dysfunctions by hNT-proBNP in no symptomatic patients. We should scan repaired TOF patients with hNT-proBNP, who had no symptom or no left-heart dysfunction, to suspect right-heart dysfunction.

P1-34

Agenesis of ductus venousus in three fetuses: A rare reason of fetal cardiomegaly

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Objective: Agenesis of the venous duct(AGV) is a rare congenital anomaly resulting in abnormal drainage of the umbilical vein into the fetal venous circulation and is associated with a high incidence of fetal anomalies and adverse outcomes, including associated malformations, chromosomal aberrations and in utero heart failure. The umbilical venous flow may take various pathways. The clinical presentation and prognosis is variable, and may depend on the specific drainage pathways of the umbilical vein.
Methods: We described 3 fetuses with agenesis of ductus venousus. Indications for fetal echocardiography were cardiomegaly, extra-cardiac anomaly and suspicion of cardiac anomaly, respectively. First fetal echocardiography was performed 29-33th weeks of gestation. Echocardiograms showed that all of them have marked global cardiomegaly and dilated umblical vein. In two fetuses the umbilical vein bypassed the portal sinus and the liver and connected to the right atrium directly. One of them had unilateral renal agenesis and the other had left renal hypoplasia. In the third fetus, absence of the ductus venosus with connection between the umbilical vein (UV) and a dilated inferior vena cava was detected. Early delivery was planned in all of them between 34-36th weeks because of hemodynamic compromise.All patients survived without any intervention. One patient had signs of congestive heart failure and required medication at early postnatal period.
Conclusions: An ADV should be ruled out in a third trimestr fetus with unexplained cardiomegaly or dilatation of the umbilical vein, systemic veins, or portal sinus. The fetus with an ADV must be followed for signs of congestive heart failure due to volume overload, including cardiomegaly, tricuspid valve regurgitation, effusions, and hydrops. Worsening of fetal heart failure may be an indication for early delivery. Beside this, associated malformations and chromosomal aberrations must be investigated.
Key words: absent ductus venosus; fetal echocardiography; prenatal diagnosis.



Figure
a. Absence of the ductus venosus with a direct connection between the umbilical vein and the right atrium. b. Absence of the ductus venosus with a direct connection between the umbilical vein and the right atrium.

P1-35

Evolving Management Of Fetal Tachycardia: Comparison Of Monotherapy, Stepwise Escalation And Maximum Dose Combination Treatment At A Single Institution

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Objectives: To compare the efficacy of initial monotherapy, stepwise escalation, and high dose combination treatment in fetal supraventricular tachycardia (SVT) and to evaluate the relationship between tachycardia response-time and treatment protocol.
Methods: Retrospective review of 50 cases managed with flecainide and digoxin for fetal SVT at the University Hospital of Wales between 2001 and 2015. Patients were divided into three groups:

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Group 1 (n = 9) had initial digoxin or flecainide monotherapy for up to two weeks before combining the two; Group 2 (n = 10) had digoxin plus flecainide stepwise escalation treatment; and Group 3 (n = 31) had maximum dose of flecainide and digoxin combination treatment from the start.
Results: In all, tachycardia termination or fetal heart rate reduction below 180 beats per minute was achieved in 49 (98%) fetuses with a median response-time of two days (range 1-27 days). Median response time was one day (mean 1.8 ± 1.1 days, range 1-7 days) in group-3 fetuses compared to nine days (range 5-18 days) and 19 days (range 9-27 days) in group-2 and group-1 fetuses respectively ($p < 0.001$ group-3 vs. group-1 and group-2).
Conclusions: High dose flecainide and digoxin combination treatment is safe and offers more effective control of fetal SVT than monotherapy or stepwise escalation protocols resulting in the shortest arrhythmia response time.

P1-36
Prenatal diagnosis and outcome of fetuses with absent pulmonary valve syndrome
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Objectives: to describe prenatal echocardiographic evaluation and outcome of absent pulmonary valve syndrome (APVS) at a single tertiary care center.
Background: APVS is a rare congenital malformation defined by a rudimentary pulmonary valve with significant regurgitant flow. It is usually associated with tetralogy of Fallot (APV/TOF), microdeletion 22q11 and arterial duct agenesis. APVS in the setting of an intact ventricular septum (APV/IVS) and a patent ductus arteriosus is less common. Reported mortality rates for patients affected is still high.
Method: we included all patients with a prenatal diagnosis of APVs between January 1995 and December 2015.
Results: among 12 fetuses with diagnosis of APVs, 8/12 (66%) had APV/TOF and no arterial duct and 4/12 (33%) had APV/IVS. None presented tricuspid atresia. Median gestational age was 35 and 38.5 weeks, respectively at last echo and at birth. None resulted affected by 22q11 deletion. In the APV/TOF group, all had concurrent severe pulmonary stenosis and significant PAs dilatation. 2/8 (25%) elected for termination of pregnancy and 2/8 (25%) died in the neonatal period for cardiopulmonary arrest. The other 4/8 patients with APV/TOF presented respiratory distress and cyanosis immediately after birth requiring respiratory support but they were all discharged and underwent surgical repair at a median age of 6 months. In the APV/IVS group all presented dilated and hypokinetic right ventricle, significant pulmonary regurgitation but only mild stenosis and PAs of normal size. One of them presented unilateral hydronephrosis. At birth 2/4 neonates with APV/IVS were asymptomatic whereas the other 2 /4 required respiratory support for severe cardiorespiratory distress and showed a wide ductus arteriosus with an unusual course and morphology ('aortopulmonary window' like). They both underwent surgical ductal closure and aortic arch reconstruction through a median sternotomy. Overall, among patients with APVS who survived the neonatal period, 7/8 are alive and well and one was lost at a median follow up of 40 months.
Conclusions: outcome after fetal diagnosis of APVS is improving for actively managed patients. In patients with APV/IVS early surgical closure of wide patent ductus arteriosus allows to improve the hemodynamic condition and to delay the homograft valve implantation.

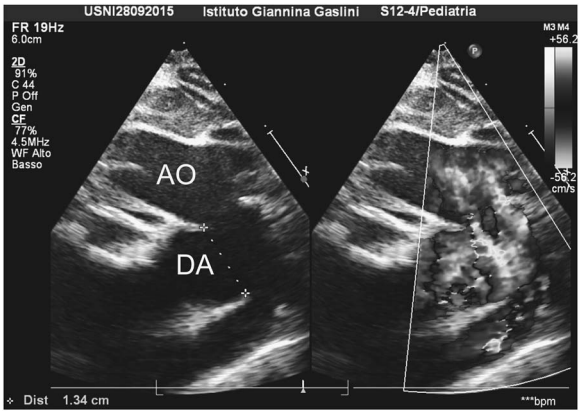


Figure 1
APV/IVS with wide patent ductus arteriosus (aortopulmonary window-like). AO: Aorta; DA: Ductus Arteriosus.

P1-37
Iindication Specific DiagnosticYield of Fetal Echocardiography
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Objectives: To document the indication specific diagnostic yield of fetal echocardiography, when performed by established referral indications.
Methods: Retrospective study based on referral indications and documented findings of fetal echocardiograms performed over 6 years (1997-2013) in an academic referral center for fetal cardiology. Referral indications have been classified as either supported by literature (Ind+) or not (Ind-). Indication specific diagnostic yields and Odds Ratios (O.R) for any abnormality, fetal congenital heart disease (fCHD) and critical fCHD when fetal echocardiography was applied in Ind+ and Ind- pregnancies have been estimated.
Results: 1782 out of 1847 fetuses (1804 pregnancies) were included, having complete referral data. 915 (51.3%) corresponded to Ind+ cases (50.4% fetal, 36.9% familial, 31% maternal origin of referral indication). Most of Ind- cases (52%) had no referral indication (family or physician wish), the remaining not established indications (including previous abortion, reduced fetal heart imaging, echogenic foci, borderline NT < 3.5mm, etc).
Overall, the incidence of any abnormality, fCHD and critical fCHD was 35.7%, 27.6% and 1.9%, respectively. The majority of fCHD were ventricular septal defects (38.6%) followed by (5-10% each) aortic coarctation, atrial septal defects and (< 5% each) valvular stenosis, arch abnormalities and more complex forms of CHD.
Ind+ compared to Ind- referrals were associated with significantly increased risk (Chi-square $p < 0.001$) for any abnormality (46.1% vs 24.5%, O.R: 2.6, 2.1-3.2), for fCHD (35.8% vs 18.9%, OR: 2.3, 1.9-2.9) and for critical fCHD (3.2% vs 0.6%, O.R:5.6, 2.6-14.5).
Diagnostic yield (%) for fCHD/critical CHD were highest (>50%) in Ind+ referrals for polyhydramnion (71/14) abnormal heart configuration in anomaly scan (56/8), chromosomal abnormalities (62/1), followed (25-50%) by referrals for fetal malformations (39/3) increased NT (36/1.8), history of maternal CHD (45/4.5). Lowest yields (< 25%), were documented in referrals for IVF (21/1.3), maternal diabetes (18/1.9), teratogen drug exposure (18/ 0), monochorionic twins (16/0).

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Conclusions: Fetal echocardiography performed for established indications is associated with significantly increased diagnostic yield for fetal CHD, which is referral indication specific. However, critical fetal CHD cases might still escape antenatal diagnosis in the absence of indication for fetal echocardiogram.

P1-38
Fetal diagnosis of pulmonary atresia with VSD vs. common arterial trunk using antenatal MRI

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Introduction: Differentiating common arterial trunk from pulmonary atresia with ventricular septal defect can be challenging antenatally. We present a case of pulmonary atresia with aorto-pulmonary collateral arteries diagnosed using fetal cardiovascular MRI as an adjunct to ultrasound.
Methods: A 20-week fetus was referred following routine antenatal screening. Serial echocardiography demonstrated a large VSD with a single outlet vessel, however the pulmonary blood supply could not be conclusively determined using ultrasound alone. The patient was therefore referred for prenatal MRI at 31 weeks. Using multiple overlapping multi-slice 2D single-shot fast spin echo sequences (Philips, 1.5 T, TR = 15,000 ms, TE = 100 ms, flip angle = 90 degrees, voxel size = 1.25 × 1.25 mm, slice thickness = 2.5 mm, SENSE factor = 2, partial Fourier-factor 5/8, slice duration 468 ms) combined with a GPU accelerated super-resolution algorithm for slice-volume registration to compensate for fetal movement, a 'black-blood'-like 3D dataset was produced with an isotropic voxel size of 0.5 mm.
Results: The resulting 2D and 3D images are demonstrated in figure 1. At least two large collateral vessels are seen originating from the anterior descending aorta. No native branch pulmonary arteries were identified. In particular, no pulmonary vessels were seen emanating from the ascending aorta, ruling out the major variants of common arterial trunk. Parental counselling was adapted accordingly.
Conclusion: Antenatal MRI offers the potential to visualise vascular anatomy more comprehensively than ultrasound alone. Clarifying the extracardiac vascular pattern can have an important impact on antenatal counselling in many forms of congenital heat disease.

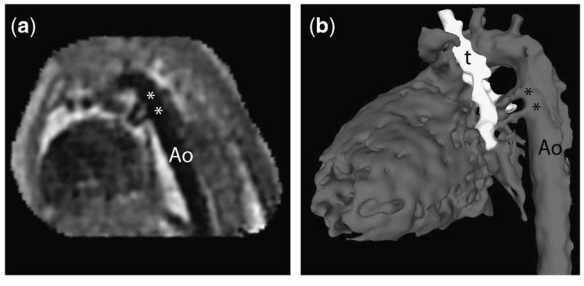


Figure 1
a: Reconstructed image from a 31 week fetus 2D image showing two large aortopulmonary collateral arteries (*) originating from the anterior descending aorta. b: 3D reconstruction of the same data. The trachea (t) is shown in white. Ao = aorta.

Funding: This work was supported by the iFind Project (Wellcome Trust IEH Award 102,431), and by the Department of Health via the National Institute for Health Research (NIHR) comprehensive Biomedical Research Centre award to Guy's & St Thomas' NHS Foundation Trust in partnership with King's College London and King's College Hospital NHS Foundation Trust.

P1-39
Long term outcome in 32 fetuses with cardiac rhabdomyomas

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Background: Rhabdomyomas (R) are more common cardiac tumors detectable in fetuses. Prenatal counselling regards a possibility of association to tuberous sclerosis (TS). The aim of this study was to analyse retrospectively the long term outcome of our cases.
Material and methods: Between 1987 and Dec. 2015 32 fetuses (0.63% of 5010 studied by echocardiography) showed cardiac masses, suggestive of R, at 21-36 w.g.; they were followed-up in utero and postnatally for a median period of 7 years (6 m-27 yrs).
Results: The diagnosis (dg) was made before 24 w.g. in 9 fetuses, 6 of them opted for the termination of pregnancy (TP) having a postmortem histological dg. of R. Two fetuses diagnosed at 27 and 28 w. with multiple large masses opted for TP (in another country), after positive MRI findings for TS.
The remaining fetuses were diagnosed at 29-36 w.g., had 2 or more larger masses with partial inflow and/or outflow obstruction in half of them. Two fetuses had mothers with TS - one had a large intrapericardial mass. One fetus had polycystic kidneys of adult type.
Growth in utero: Cardiac masses grew usually till 35-36 w.g. Diagnosis of TS was made in 18 cases by positive CNS MRI findings of supendymal/cortical tuberi after birth- in 16 of 24 (67%) continuing pregnancy and in 2 after the late TP; total percentage of 69%. Genetic analysis was performed up to now in a third of cases.
Outcome: none of the live born cases needed cardiac surgery and the cardiac masses progressively regressed over time, 8 cases have extrasystolic arrhythmia and one had a short episode of supraventricular tachycardia. All TS cases have neurological signs-seizures, 3 were operated for astrogliomas. One case is on dialysis, others have minor renal, dermatological and ocular signs. Conclusions: Cardiac R show variable characteristics, postnatal regression of cardiac masses but a relevant association with TS that conditions the clinical state.

P1-40
Fetal Cardiac Time Intervals in Healthy Pregnancies- An Observational Study by Fetal ECG (MONICA Healthcare System)

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Background: Fetal electrocardiogram (fECG) can detect QRS signals in fetuses from 17 weeks' gestation onwards; however,

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the technique is limited by the minute size of the fetal signal relative to noise ratio. The aim of this study was to evaluate precise fetal cardiac time intervals with the help of a newly developed fetal ECG device.
Methods: The small and wearable Monica AN24 monitoring system uses standard ECG electrodes placed on the maternal abdomen to monitor fetal ECG, maternal ECG and uterine EMG. 149 fECGs were performed on healthy fetuses (> 32 weeks gestational age). Fetal cardiac time intervals were estimated on 1000 averaged fetal beats. Detection was deemed successful if there was a global signal loss of less than 30% and an analysis loss of the Monica AN24 signal separation analysis of less than 50%.
Results: After applying the requirements 117 fetal ECGs remained for CTI analysis. CTI measurements (in ms) were comparable to other available methods, such as e.g. fetal magnetocardiography.
Conclusions: Although limited and preclinical in its use, fetal ECG (MONICA Healthcare System) could be an additional and less expensive useful tool to detect precise fetal cardiac time intervals from 32 weeks gestational age onwards. This helps to better define fetal arrhythmias.

P1-41
Standard values and influencing factors for fetal acceleration and deceleration capacity: providing insights into the maturation of the fetal autonomic nervous system
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Background: Fetal heart rate variability is an indirect measurement of the fetal autonomic nervous system. Phase-rectified signal averaging is a new method of complex biological signal analysis. Its acceleration and deceleration capacity provides insights into the maturation of the fetal autonomic nervous system. The aims of this study were to determinate standard values for average acceleration capacity (AAC) and deceleration capacity (ADC) in the third trimester of pregnancy and to investigate influencing factors on AAC and ADC.
Methods: Measurements of fetal heart activity by cardiotocography from 32nd to 40th week of gestation in uncomplicated pregnancies (n = 149). Studied influencing factors were maternal age, gravidity, artificial insemination, BMI, ethnicity, medication intake (Mg, Fe, others), nicotine abuse, socioeconomic status. Outcome parameters were birth weight, gender, pH, Apgar-Score. AAC and ADC were determinated by the means of Phase-rectified Signal Averaging (PRSA).
Results: No significant alterations in the sympathovagal balance were found from 32nd weeks of gestation until delivery. But significant decrease of AAC was found beyond the presumed date of birth indicating an impairment of the fetal autonomic nervous system (ANS). In addition, low birth weight was associated with an activation of the fetal autonomic nervous system and high maternal age was related to a reduced fetal heart rate variability.
Conclusion: In the presence of high maternal age, low birth weight or pregnancy beyond the presumed date of birth, influence on the fetal autonomic nervous system can be found.These mechanisms can have an influence on cardiovascular events.

P1-42
Prenatal diagnosis of 51 fetuses with Aortic Arch Anomalies :Prevalence, associated findings and perinatal outcome in a single institution
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Objective: To evaluate the incidence and associations of AAA in a population of fetuses referred for fetal echocardiography in our third level center (AORN dei colli – AO Monaldi, Second university of Naples). To report the pre and postnatal outcome of affected cases.
Methods and results: Since January 1995 to March 2015 we practiced 9048 fetal heart scans. Among these, we diagnosed 1281 (14%) consecutive fetuses with CHD. An AAA was diagnosed in 51/9048 (0.6%) fetuses (51/1281 CHD; 4%). Mean gestational age at diagnosis was 24.4+/- 2.4 weeks. The detection rate of a AAA increased over the study period: 41/51 (80%) cases were diagnosed from 2007 to 2015; this finding is in accord with literature.
There were 51 (42%) AAA, 47 (92%) of which were RAA (right aortic arc), 2 (4%) of which were ARSA (right subclavian artery aberrant) and 2 (4%) were DAA (Double aortic arch).
There were 18 (38%) isolated RAA. A RAA was found in association with ct-CHD in 29 cases (62%): 20 Tetralogy of Fallot, 5 Pulmonary atresia with ventricular septal defect, 3 double outlet right ventricle with pulmonary stenosis and 1 ventricular septal defect .
Karyotype was performed during fetal life in 28/51 AAA (55%), 10/28 (55%) were pathological.
Of the chromosomal abnormalities: 7 /10 (70%) were 22q11 deletions, in 2 of the 7 cases (29%) RAA was isolated and 5/7(71%) RAA was associated with CHD .2/10 (20%) were trisomy 21, of which one was isolated ARSA and one TOF with RAA, 1/10 (10%) was trisomy 18.
There were 9 (18%) pregnancy interruptions, 1 (2%) intrauterine death, 33 (65%) live births, 4 (8%) neonatal deaths. In the remaining 4 (8%) pregnancies is still continuing.
Conclusions: AAA are rare, but during the last years, we observed an increasing number of AAA because of the increased skill of fetal echocardiographers.
An AAA can be associated to ct-CHD and /or anomalies of the Karyotype in a significant proportion.
This is the reason why, when diagnosed an AAA, the couple must be offered a genetic study.

P1-43
Prenatal Prediction of aortic coarctation: 15 years of experience
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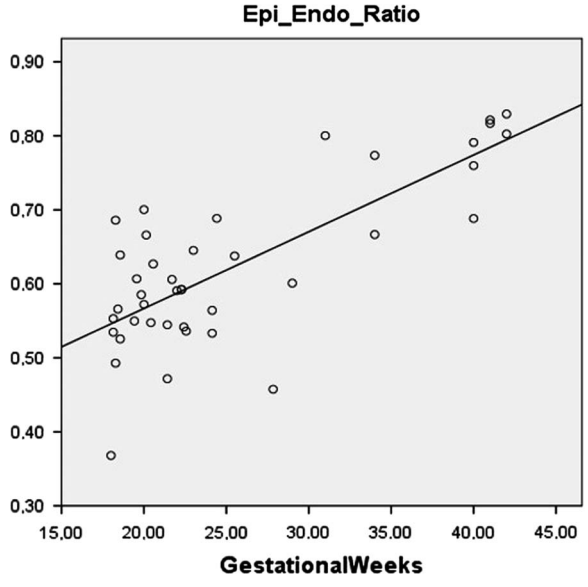
Introduction (or Basis or Objectives): To assess which parameters provides the best prenatal prediction of coarctation of aorta (CoAo) and how our approach change during the years.
Methods: We selected all cases of right prevalence prenatally diagnosed in 2000-2015; then we reviewed the prenatal echocardiograms and postnatal outcome of the infants with and without

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coarctation of the aorta established postnatally, to identify how our percentage of reliability has changed over the years and which echocardiographic findings proved to be the most useful.
Results: Between 2000 and 2015, 143 fetuses referred for great vessel disproportion and cardiac asymmetry were studied. We divided it into two groups:
Group 1: From 2000-2007 we suspected CoAo in 70 fetuses (EG 29+ /- 4) and the diagnosis was confirmed in 38 cases (54%). The benchmarks were basically tricuspid valve/mitral valve ratio with the main pulmonary artery/AAo ratio (0.7) GROUP 2: From 2008-2015 we suspected CoAo in 73 fetuses (EG 28+ /- 4) and the diagnosis was confirmed in 55 cases (75%). The false positive rate was lower in the second group, the improvement was due to a combination of parameters used: aortic annulus size compared gestational age (z-score of the ring valve < -2), the z-score of ascending aorta (AAo) (< - 1.5) and aortic isthmus (three vessels and trachea view)(< -2) related to EG, tricuspid valve/mitral valve ratio with the main pulmonary artery/AAo ratio. The earlier in the EG much more meaning we give it (< 28 W).
Conclusions: Prenatal diagnosis still remains burdened with a high rate of false positives, but the use of the z score related to gestational age and the construction of a multi-parametric scoring system allowed us to lower the margin of error in this diagnosis.

P1-44
Assessment of left ventricular myocardium development during the fetal life by speckle tracking echocardiography
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Background: Human left ventricular myocardium is an anisotropic tissue in which the constituent cardiomyocytes are arranged in a complex three-dimensional network. However, the timing of the development myocardial layers during the fetal life is still unknown.
The aim of the study is to study the development of left ventricular myocardial layers by speckle tracking at different gestational ages.



Methods: We enrolled 30 normal fetuses aged between 18 and 34 weeks of gestation and 11 normal infants. Longitudinal endocardial and epicardial strain by speckle tracking echocardiography was obtained by apical four chambers view.
Results: During the fetal life, endocardial and epicardial longitudinal strain values progressively increased (Endo: $r = -0.49$; $p = 0.006$; Epi: $r = -0.39$; $p = 0.04$). The endocardial layer developed earlier than the epicardial layer, however the ratio between epicardial and endocardial strain progressively increase during the fetal life (Figure) ($r = 0.73$; $p < 0.0001$).
Conclusion: Left ventricular myocardium maturation is a process that begins relatively late during fetal life and is ongoing also after birth. The subendocardial layer appears earlier than the epicardial layer.

P1-45
Cord blood cardiac biomarkers profile and echocardiographic ventricular function parameters in newborns with prenatal diagnosis of congenital heart defects
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Objective: The aim of this study was to evaluate the usefulness of cardiac biomarkers in the cord blood newborns with prenatal diagnosis of congenital heart defects (CHD).
Method: From August 2012 to March 2015, newborns with prenatal diagnosis of CHD were admitted consecutively at a Neonatal Intensive Care Unit. Healthy newborns delivered in the same hospital were recruited as controls. Plasma levels of cardiac biomarkers were measured in the cord blood and echocardiogram performed in both groups. Biomarkers cut-off value was generated and correlations with echocardiographic parameters were performed.
Results: No significant differences were observed between BNP, CK-MB and myoglobin between CHD and control groups. CK-MB levels were significantly higher in cyanotic CHD patients than in acyanotic CHD. The cut-off offering optimal accuracy was 3.45 ng/mL for CK-MB (sensitivity 75%, specificity 67%). CK-MB cord blood levels were positively correlated with the tricuspid valve E/E' ($p = 0.002$; $rho = 0.772$). Troponin I levels were negatively correlated with the mitral valve lateral annulus slope ($p = 0.04$; $rho = -0.412$).
Conclusion: BNP, CK-MB and myoglobin cord blood levels were similar between CHD patients and healthy newborns. CK-MB cord blood levels were higher in newborns with cyanotic than in acyanotic CHD and were correlated with right ventricular diastolic function parameters.

P1-46
Perinatal care of fetuses with heterotaxy syndromes
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Objectives: To evaluate the spectrum of prenatal findings and obstetric perinatal care of fetuses with heterotaxy syndromes.
Methods: A retrospective review of fetuses with heterotaxy syndrome. Fetal echocardiography findings, perinatal care and obstetric outcomes were evaluated.

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Results: 37 fetuses with heterotaxy syndrome, 21(57%) with left isomerism(LAI) and 16(43%) with right isomerism(RAI) were diagnosed and delivered in our institution between January 2006 and December 2014. The main cardiac pathologies were AVSD 49%(18/37), pulmonary stenosis 32%(12/37) and pulmonary atresia 22% (8/37). Dextrocardia was in 22%(8/37). Other abnormalities in RAI: total or partially anomalous pulmonary venous drainage 44%(7/16), TGA 31% (5/16) and SV 31% (5/16). In LAI: common atrium 24% (5/21), DORV 19%(4/21), hypoplastic left ventricle 19%(4/21), interrupted inferior vena cava with azygos continuation in 52% (11/21). Rhythm disturbances: bradyarrhythmias were only observed in fetuses with LAI 57% (12/21). Situs inversus was the most common visceral anomaly in 41%(15/37) of all fetuses. Asplenia was in 22% (8/37) of fetuses, mainly in RAI 37%(6/16). Polysplenia was only seen in 14%LAI patients (3/21). The median gestational age at delivery was 38.6 weeks. 73% (27/37) were born by vaginal delivery, 16%(6/37) by elective cesarean section and 11%(4/37) by urgent CC. Excluding the parameter of bradycardia, abnormal intrapartum fetal heart rate patterns were observed only in two cases: reduced variability lasting more than 80 minutes (5.4%) and high variability lasting more than 10 minutes (5.4%). There were only 3 cases of Apgar score less than 7 at 5 minutes, and no cases of umbilical cord blood pH lower than 7.0. Out of 37 fetuses 20(54%) children, 13 with LAI (35%) and 7 with RAI (19%) alive at the time of publication.
Conclusions: The spectrum of cardiac and extracardiac abnormalities of HS is wide. Vaginal delivery is safe for this fetuses so CC should be reserved only for traditional obstetric indications. Electronic fetal monitoring is not always possible due to rhythm disturbances, especially in LAI.

P1-47
Cardiovascular Risk Assessment and Ventricular Strain in Children with Psoriasis

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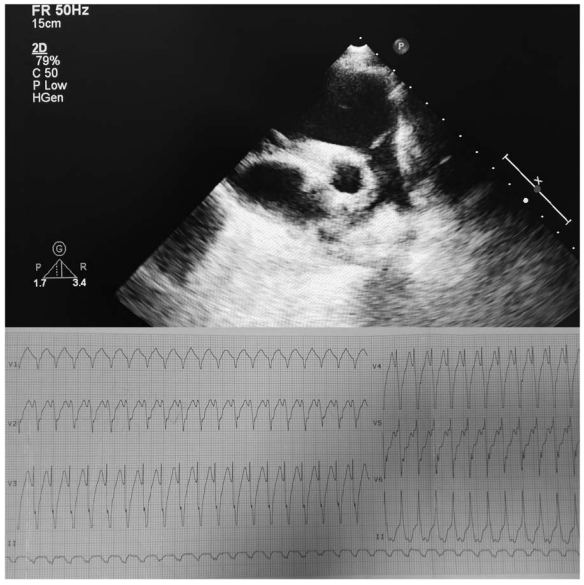
Introduction: Psoriasis is a systemic inflammatory disease affecting both adults and children. It is increasingly recognized that psoriasis may cause cardiovascular events in adults. This increased risk wasn't studied in children up to date. We investigated cardiovascular risk in childhood psoriasis by using carotisintima-media thickness(CIMT), arterial functions and ventricular strain.
Methods: The study consisted of 20 patients with psoriasis and 20 controls. Psoriasis activity and severity index (PASI) was calculated. Echocardiographic examinations were performed by using Philips IE33 echocardiography machine equipped with a L-11MHz lineer probe for carotid and arterial studies and 5-MHz transducer for ventricular strain. CIMT, arterial stiffness, distensibility, flow mediated dilatation of brachial artery (FMD) were calculated. M-mode and Doppler studies were performed in standart fashion. Longitudinal and global strain were studied using apical four-chamber (4 C), long axis (LAX),and two-chamber (2 C) views.
Results: The mean age of patients was 14.2 ± 0.89 (12-16 years) (12 girls, 8 boys) and the control group 14.05 ± 0.88 (12-16 years) (12 girls, 8 boys). Mean follow-up period was 8.7 months (5-14 months), PASI was 7.0 ± 1.84 (4-10.7). None of the patients had arthritis, overt cardiovascular disease or under medical treatment.
CIMT was higher in patients than control group, difference was not statistically significant. Aortic stiffness was significantly higher in psoriasis group (0.32 ± 0.14; 0.18 ± 0.089, $p = 0.02$). FMD (at 1st and 3rd minutes) didn't differ significantly between the groups.
There were significant differences in terms of interventricular septum diastolic (IVSD), left ventricle posterior wall diastolic diameter (LVPWD), Mitral E, Mitral A and E/A values between the patients and the control group (0.71 ± 0.065, 0.68 ± 0.04; 0.73 ± 0.041, 0.75 ± 0.038; 0.68 ± 0.07, 0.76 ± 0.056; 0.43 ± 0.07, 0.35 ± 0.06; 1.64 ± 0.22, 2.17 ± 0.35, $p < 0.05$ respectively). Tissue Doppler imaging (TDI) revealed significant changes in terms of mitral lateral annulus E', A', E'/A, isovolumetric contraction time (IVCT), and ejection time(ET) (0.62 ± 0.07, 0.70 ± 0.02; 0.39 ± 0.036, 0.50 ± 0.031, 1.58 ± 1.88, 1.34 ± 1.14, 74.7 ± 6.65, 75 ± 5.77, 154.85 ± 3.57, 137.95 ± 10.84; $p < 0.05$ respectively).
Global circumferential and longitudinal strain were significantly lower in patients (-21.3 ± 3.75, -23.6 ± 3.78; -19.8 ± 1.89, -23.4 ± 3.92; $p < 0.05$ respectively).
Conclusion: Our study demonstrated impaired ventricular and arterial functions and increased risk for atherosclerotic heart disease in psoriatic patients and the early changes are detectable even during childhood. Eliminating other preventable risk factors and close monitorization may be helpful in decreasing deletorious cardiovascular events.

P1-48
Arrhythmogenic dysplasia, Cutaneous Features and Desmoplakin Mutation; Carvajal Syndrome

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Introduction: Carvajal syndrome is rare autosomal recessive form of ARVD with distinct dermatologic features. We present a case with alopecia, palmoplantar keratosis, sustained VT and biventricular dysfunction.
Case: Four-year-old boy admitted to emergency room with palpitation and vomiting. Previous history of arrhythmia was absent. Parents were non-consanguineous. Mother's aunt has died in sleep in fifties. He was conscious, heart rate was 214/ min, blood pressure was 90/57 mm Hg. Alopecia and palmoplantar keratoderma were present. ECG showed sustained VT with left bundle branch block morphology and superior QRS axis. Tachycardia was unresponsive to adenosine and amiodarone. Lidocain was effective in termination. Resting ECG revealed diminished QRS voltages, epsilon waves in right precordial leads (V1-3), slurring of S waves (>50 msn). Echocardiography revealed dilatation of both ventricles, left ventricular EF was 45%, RVOT was dilated. MRI revealed global hypokinesia of both ventricules, transmural contrast enhancement on apico-basal segments of infero-lateral walls of left ventricle, subendocardial contrast enhancement at right ventricular septum and RVOT. Alopecia and palmoplantar keratosis was present since infancy. Heterozygot c.3564 T > A and c.4395 T > A mutations were found in exon 23 of desmoplakin coding gene. Mother was carrier for c.3564 T > A mutation. Metoprolol, Kaptopril, Furocemide was started; extracardiac defibrillator was implanted for prevention of VT attacks. Pathogenic mutations, sustained VT with left bundle branch block morphology, epsilon waves were compatible with major criteria of ARVD and its syndromic type Carvajal Syndrome.
Conclusion: Cardiocutaneous syndromes with cardiomyopathy and serious rhythm abnormalities should be considered in patients with alopecia and palmoplantar keratosis.

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P1-49
Echocardiographic Assessment of Mitral Regurgitation in Children and Relation to Pro-BNP Levels

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Introduction: We studied children with chronic mitral regurgitation (MR) due to rheumatic fever (RMR) or mitral valve prolapse (MVP) using echocardiographic parameters and their relation to serum NT-ProBNP levels and QTc dispersion as markers of volume overload and impaired repolarization.
Methods: The study included 39 patients (mean age = 11.8 ± 3.3; range: 4-19) with chronic mitral regurgitation due to either mitral valve prolapsus (MVP)(27) or rheumatic valve disease (RMR) (12) and 23 healthy children (mean age = 10.9 ± 2.81; range: 7-16) as control group. Serum NT-ProBNP levels were obtained, QTc dispersion was calculated using 12-lead ECG; 2D, M-mode, Doppler, Tissue Doppler and Strain echocardiography were performed using Philips IE33 Ecocardiography machine equipped with 5 MHz transducer. LV sytolic and diastolic diameters, volumes, ejection fraction, vena contracta, regurgitant volume, effective regurgitant orifis area; global circumferential and longitudinal strain of left ventricle were calculated. The patients were divided into two subgroup as mild or moderate/severe subgroups according to the criteria of European Association of Echocardiography and divided two another subgroups as MVP or rheumatic mitral regurgitation (RMR).
Results: Plasma NT-ProBNP levels have increased in severe MR comparing to mild MR subgroup (170 ± 225 pg/ml in severe MR, 53 ± 30 pg/ml in mild MR p = 0.019) significantly. Plasma NT-ProBNP levels of MR group correlated to mGLS (mean global longitudinal strain) (p = 0.017 r: -0.414). mGCS (mean global circumpherential strain) correlated to QTc dispersion (p = 0.007 r:0.44) and vena contracta (p = 0.018 r: 0.39) in MR group. mGLS correlated to QTc dispersion in RMR subgroup (p = 0.032 r: 0.040). In MVP subgroup QTc dispersion correlated to LVESV/m^2 (p = 0.01 r: 0.48). MPI (myocardial performance index) correlated to Regurgitant volume (p = 0.011, r: -0.41) in MR group.
Conclusion: NT-ProBNP levels increased with severity of MR. We found a relation between NT-ProBNP and mGLS which may provide more powerful assesment of global ventricular function when used together. MPI is another echocardiographic parameter indicating global ventricular function and its relation to mGLS is in accordance with this knowledge. Increased QTc dispersion also accompanies left ventricular dysfunction and reflects impaired repolarisation. This study emphasises NT-ProBNP levels, mGLS, mGCS and MPI together provides a powerful assesment of global LV function ın chronic MR in children.

P1-50
Prolonged Tp-e interval and Tp-e QT ratio in children with Mitral Valve Prolapse

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Introduction: Although it is considered as a benign condition, previously reported studies have showed that a subset of patients with mitral valve prolapse (MVP) may be at risk of ventricular arrhythmia and sudden cardiac death (SCD). Previously reported studies have suggested that the interval between the peak to the end of the T-wave (Tp-e) can be used as a marker of transmural dispersion of repolarization (TDR) and increased Tp-e interval and Tp-e/QT ratio Also, Tp-e/QT are associated with ventricular arrhythmias and SCD. The aim of this study was to assess alterations in ventricular repolarization by using Tp-e interval and Tp-e/QT ratio in children with MVP and to investigate its relationships with degree of valvular regurgitation.
Methods: This study prospectively investigated 110 children with MVP and 107age-and sex-matched healthy control subjects. Tp-e interval, Tp-e/QT ratio, QT and QTc dispersions were measured form a 12-lead electrocardiogram were compared between groups.
Results: QT, QTc dispersions, Tp-e interval, Tp-e/QTc ratios were found significantly higher in patients with MVP (Table 1) .A positive correlation was found between Tp-e/QTc ratio and the degree of MR ($p < 0.05$; $r = 0.2$). However, degree of MR was not associated with QT, QTc and Tp-e intervals, QT, QTc, Tp-e dispersions and Tp-e/QT ratio (all p values > 0.05).
Conclusion: Individuals with MVP may be more prone to ventricular arrhythmias due to prolonged QTd, QTcd, Tp-e interval, increased Tp-e/QT and Tp-e/QTc ratios. Therefore, due to their longer life expectancy, children with MVP should be followed up in terms of life-threatening arrhythmias.

Table 1. Electrocardiographic parameters of patients and controls

Parameters	Patient group (n:110)	Control group (n:107)	p
QT (ms)[a]	340 (40)	320 (40)	0.46
QTc (ms) (mean ± SD)	388 ± 25.8	390 ± 25.1	0.85
QT dispersion (ms)[a]	40 (20)	20 (20)	<0.001
QTc dispersion (ms)[a]	20 (30)	10 (10)	<0.001
Tp-e interval (ms)[a]	90 (20)	80 (20)	0.02
Tp-e interval dispersion (ms)[a]	40 (20)	40 (10)	<0.001
Tp-e /QT[a]	0.26 (0.04)	0.25 (0.05)	0.001
Tp-e/QTc (mean ± SD)	0.23 ± 0.03	0.21 ± 0.03	<0.001

[a]Data are expressed as median with range in parentheses;
ms: milliseconds; n = number of the subjects.

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P1-51
Evaluation Tp-e interval and Tp-e/QT ratio in children with aortic stenosis

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Objective: Patients with aortic stenosis (AS) may experience some life-threatining cardiac events due to the ventricular arrhythmias. Recently, new ECG-derived indexes such as Tp-e which is the measurement of the interval between the peak and the end of the T-wave has emerged as a marker of transmural dispersion of repolarization (TDR). As Tp-e interval, Tp-e/QT ratio is also used as an index of ventricular repolarization. Prolongation of Tp-e interval and increased Tp-e/QT ratio have been found associated with malignant ventricular arrhythmias. Although, these parameters have been studied in adult patients.The novel repolarization indexes Tp-e and Tp-e/QT have not been studied among children with AS previously. The aim of this study was to evaluate Tp-e interval and Tp-e/QT ratio in children with AS.
Methods: The standard 12-lead electrocardiograms of 66 children with aortic stenosis and 58 age-and-sex matched healthy children were assessed by a blinded specialist.
Results: Tp-e, QTc and QT dispersions were found significantly higher in AS group compared to healthy subjects (Table). However we did not find any difference in Tp-e interval, Tp-e/QT and Tp-e/QTc ratios between AS patients and the controls. When AS patients were compared according to degree of aortic stenosis (mild, moderate and severe). We did not find any difference Tp-e interval,Tp-e dispersion, QT dispersion, Tp-e/QT and Tp-e/QTc ratios between groups. And we did not find any correlation between these parameters and severity of aortic stenosis.
Conclusions: Our study showed that Tp-e, QT and QTc dispersions were increased in children with AS irrespective degree of Aortic stenosis. Further prospective studies are needed for demonstrating the clinical importance of these parameters.

Table. Electrocardiographic characteristics of patients and controls

Variables	*Patient group (n:66)*	*Control group (n:58)*	*p*
Age	6.7 ± 5.9	7.2 ± 3	0.58
Gender (M/F)	49/17	29/29	0.36
Tp-e interval (ms)	79 ± 21	82 ± 14	0.44
Tp-e interval dispersion (ms)	24 ± 11	41 ± 20	< 0.001
QT interval (ms)	310 ± 51	321 ± 32	0.17
QT dispersion (ms)	29.2 ± 17	21 ± 14	0.02
OTc interval (ms)	393 ± 21	398 ± 26	0.23
QTc dispersion (ms)	25 ± 18	15 ± 11	0.001
Tp-e /QT	0.25 ± 0.05	0.25 ± 0.04	0.85
Tp-e/QTc	0.19 ± 0.05	0.20 ± 0.03	0.5

P1-52
Effects of Balloon Pulmonary Valvuloplasty on ventricular arrhythmogenesis

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Objectives: Recently, increasing number of studies have showed that the peak and the end of the electrocardiographic T wave (Tp-e) corresponds to the transmural dispersion of ventricular repolarization and the prolongation of the Tp-e interval has been reported to be associated with ventricular arrhythmias. Tpeak-end interval, Tp-e/QT and Tp-e/QTc ratios are used as an index of ventricular arrhythmogenesis. As an independent variable from heart rate that is suggested a strong predictor of sudden death in cardiac patient population. The aim of this study was to assess ventricular repolarization in patients with severe pulmonary valve stenosis who underwent balloon valvuloplasty and also was to assess the relation between the ventricular repolarization and residual pulmonary valve gradient.
Methods: Thirty patients who underwent balloon valvuloplasty between January and August 2014 were enrolled into study and 50 age and sex matched healthy subjects were also recruited as control group. ECG recordings of patients vs control group measurements revealed Tp-e interval of 85 ± 17.3 vs 82.5 ± 14.9msec (p: 049), QTc 426.9 ± 33.9 vs 393.7 ± 25.6msec p = 0.03, QT dispersion 33.6 ± 20.4 vs 29.4 ± 18.5msec p = 0.01, Tp-e/QTc ratios 0.19 ± 0.03 vs 0.21 ± 0.04 p = 0.23. The ECG measurement of these parameters showed a strong correlation between pulmonary valve residual gradient, Tp-e interval (r = 0.88, p < 0.01), QT dispersion (r = 0.86, p < 0.01) and Tp-e/QTc (r = 0.77, p < 0.01).
Conclusion: Balloon valvuloplasty is the procedure of choice for the valvular stenosis at all ages however, patients with residual gradient may have the risk of ventricular repolarization heterogeneity. Therefore, patients who have significant pulmonary residuel gradient after pulmonary balloon valvuloplasty may more prone to ventricular arrhythmias and these patients should be closely follow up for the risk of ventricular arrhythmias and sudden cardiac death.

P1-53
The effects of L-Thyroxine treatment on the cardiac functions of the infants with congenital hypothyroidism

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Objective: This study aims to determine how the cardiac functions are altered in infants with congenital hypothyroidism before and after L-thyroxine treatment.
Methods: The patient group consisted of 25 infants who are aged between 0 to 90 days and who are diagnosed with congenital hypothyroidism. The infants with congenital hypothyroidism were treated with L-thyroxine until thyroid function tests were normalized. The control group included 20 healthy infants who were matched with respect to age and body mass index.
Results: Conventional echocardiography showed that the patient group had significantly shorter deceleration time than the control group (p = 0.001). After L-thyroxine treatment, systolic volume, end-diastolic volume, diastolic left ventricular posterior wall diameter, end-systole and end-diastole left ventricular internal diameter increased significantly in the infants with congenital hypothyroidism (respectively p = 0.009, p = 0.02 and p = 0.015 and p = 0.001). Pulse Doppler echocardiography demonstrated that the patient group had significantly lower mitral A, tricuspid E, right ventricle E' and E'/A' values but significantly higher right ventricle A' values than the control group (respectively p = 0.011, p = 0.016, p = 0.033, p = 0.03 and p = 0.024). The E', E'/A', S', TAPSE and TE/TA values of the right ventricle and mitral E and S' values increased significantly after L-thyroxine treatment in the patient group (respectively p = 0.014, p = 0.017, p = 0.004, p = 0.006, p = 0.004, p = 0.03 and p = 0.003). Tissue Doppler echocardiography indicated that right ventricle myocardial performance index was significantly higher in the patient group (p = 0001). When compared with the control group, the aort diameter was significantly larger in the patient group (p = 0.037). The S' value for the interventricular septum and the aort diameter

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increased significantly after L-thyroxine treatment ($p = 0.038$ and $p = 0.015$ respectively).
Conclusion: Hypothyrodism exerts negative effects on the systolic and diastolic functions of the heart and these effects are more prominent in the right ventricle. L-thyroxine treatment provides improvement up to a point and this improvement is more pronounced in the right ventricle. Conventional and pulse Doppler echocardiography may not detect the systolic and diastolic dysfunction in infants with congenital hypothyroidism. Tissue Doppler echocardiography can help to specify the subtle but significant alterations in cardiac functions which are related with hypothyroidism and/or L-thyroxine treatment.

P1-54
Can Electrocardiography Findings Indicate Cardiac Autonomic Dysfunction in Children Presenting with Vasovagal Syncope?

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Objective: This study aims to determine the usefulness of electrocardiography findings as a sign of cardiac autonomic dysfunction in children presenting with vasovagal syncope.
Methods: This is a prospective review of 92 children presenting with vasovagal syncope and 50 healthy children who were matched with respect to age and sex. Fifty children presenting with syncope had negative head up tilt (HUT) test while 42 children presenting with syncope had positive HUT test. Comparisons were made according to the electrocardiography findings related with p-wave, QT interval, T-wave and heart rate variability.
Results: The healthy controls, HUT test negative children and HUT test positive children were statistically similar in aspect of age, sex, height, weight and body mass index. When compared with the healthy controls, the HUT positive children had significantly slower heart rate ($p < 0.05$), longer p minimum ($p < 0.01$), longer p maximum ($p < 0.01$), greater p-wave dispersion ($p < 0.01$), longer QT minimum ($p < 0.01$), longer QT maximum ($p < 0.01$), greater QT dispersion ($p < 0.05$) and longer T-peak-to-T-end intervals in V2, V3, V4, V5 and V6 leads ($p < 0.01$, $p < 0.01$, $p < 0.05$, $p < 0.05$ and $p < 0.01$ respectively). The HUT positive children had significantly longer T peaks in V2, V5 and V6 leads than the T-peak-to-T-end intervals in V2, V5 and V6 leads of the HUT negative children ($p < 0.01$, $p < 0.05$ and $p < 0.05$ respectively). When compared with the HUT negative children, the HUT positive children had significantly higher heart rate variability.
Conclusion: Autonomic cardiac dysfunction may be predicted by ECG findings such as significantly greater p-wave dispersion, greater QT dispersion, prolonged T-peak-to-T-end intervals and increased heart rate variability in children with vasovagal syncope.
Keywords: children; electrocardiography; head up tilt test; vasovagal syncope

P1-55
Serum Vitamin B12 Levels in Children Presenting with Vasovagal Syncope

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Objectives: Vasovagal syncope is usually defined as sympathetic-parasympathetic imbalance but its precise pathophysiology remains obscure. Vitamin B12 deficiency may cause neurologic deficits and affect the autonomic nervous system. This study aims to determine the serum concentrations of vitamin B12 in children presenting with vasovagal syncope.
Methods: This is a prospective review of 160 children presenting with vasovagal syncope. Patients with cardiac, neurologic, and psychiatric illness, chronic disease and arrhythmia, and any drug use that can alter cardiac conduction velocity heart rate and blood pressure were excluded. Subgroup analysis was done based on the results of head up tilt test.
Results: Head up tilt test gave positive results in 80 children and this test yielded negative results in the remaining 80 children. The tilt test positive children had significantly lower TSH concentrations ($p = 0.06$), total iron binding capacity ($p = 0.04$) and vitamin B12 levels ($p = 0.01$). The prevalence of vitamin B12 deficiency was significantly higher in the tilt positive group (80% vs 52.5%, $p = 0.001$). Out of 80 children with positive tilt test, 8 children (10%) showed cardioinhibitory response, 22 children (27.5%) demonstrated vasodepressor response, 24 children (30%) displayed mixed response and 26 children (32.5%) had postural orthostatic tachycardia syndrome (POTS). Erythrocyte sedimentation rate was significantly lower in the mixed response group than in the vasodepressor group (6.2 ± 0.8 mm/h vs 14.3 ± 2.5 mm/h, $p = 0.001$). Serum vitamin B12 concentrations were significantly lower in the POTS group than in the vasodepressor group (240.8 ± 38.2 pg/ml vs 392.7 ± 27.1 pg/ml, $p = 0.001$). The prevalence of serum vitamin B12 deficiency was significantly higher in the POTS group than in the vasodepressor group (92.3% vs 45.5%, $p = 0.001$).
Conclusion: Vitamin B12 acts as a co-factor for three enzymes: (1) phentolamine N-methyltransferase which is needed for the conversion of noradrenaline to adrenaline, (2) catecholamine-O-methyltransferase which is required for the degradation of catecholamines, (3) methylmalonyl coenzyme A (CoA) mutase which catalyzes the conversion of methylmalonyl-CoA to succinyl-CoA in myelin synthesis. Vitamin 12 deficiency causes reduction in myelinization, deceleration in nerve conduction and elevation in serum concentrations of noradrenaline. These factors may contribute to the impairment of autonomic functions which are involved in the pathogenesis of vasovagal syncope.

P1-56
Decreased cardiovascular function late after ventricular septal defect repair

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Introduction: Survival after ventricular septal defect (VSD) repair is excellent but in adulthood late residua including conduction disease, arrhythmia and heart failure are not uncommon. This study aims to evaluate cardiac function, heart rate variability, exercise capacity and physical activity in children late after VSD repair.
Methods: 28 patients after VSD repair and a healthy agematched control group underwent an echocardiogram, maximal exercise test and 24 h holter monitoring. Physical activity including physical education at school, regular biking/walking behavior and sports participation was mapped using a questionnaire.
Results: Age of the patients and controls ranged from 8-18y. Patients were operated at a mean of 1.7y and time after intervention was on average 10.1y. Right ventricular function, measured by Tissue Doppler Imaging was significantly less in patients compared to controls (S':0.11 ± 0.02 vs 0.12 ± 0.02 E':0.12 ± 0.04

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vs 0.15 ± 0.03 A':0.06 ± 0.02 vs 0.09 ± 0.02, all $p < 0.001$) and lower systolic function (S') was associated with physical activity ($R2 = -0.166$; $p = 0.039$). Average strain in the apical long axis view was also lower in patients (-17.5 ± 3.4 vs -19.2 ± 3.7, $p = 0.023$). Ejection fraction and stroke volume from 3D echocardiography were both lower in patients (47.1 ± 5.1 vs 51.3 ± 6.3, $p = 0.002$ and 40.2 ± 20.2 vs 50.7 ± 14.3, $p = 0.008$). Patients achieved a lower maximal power (130 ± 52 vs 152 ± 57, $p = 0.023$) and maximal oxygen uptake (39.6 ± 6.7 vs 44.6 ± 7.3, $p = 0.002$) in the exercise test and had a lower level of weekly physical activity compared to controls (37.8 vs 50.7 MET/week; $p = 0.028$). Heart rate was significantly higher during sleep in patients (70.5 ± 8.2 vs 65.9 ± 9.1, $p = 0.023$). During the 24 h recording, patients had more premature ventricular and atrial contractions (145 ± 464 vs 8 ± 33, $p = 0.023$ and 2 ± 7 vs 1 ± 9, $p = 0.016$). Heart rate variability analysis revealed a lower low frequency power (624 ± 447 vs 880 ± 615 $p = 0.049$) and lower standard deviation of interbeatintervals (61.3 ± 17.3 vs 78.1 ± 22.5, $p = 0.001$) during physically active periods in patients.
Conclusions: 10 years after surgical correction of a VSD, cardiovascular function is decreased compared to healthy controls. This study shows that exercise capacity, cardiac function and heartrate variability are already decreased in childhood. Therefore, it is important to continue followup in patients after VSD repair into adulthood.

P1-57
Ischemia Modified Albumin, Cardiac Troponin T And N-Terminal Pro-B Type Natriuretic Peptide Levels in Infants Of Diabetic Mothers

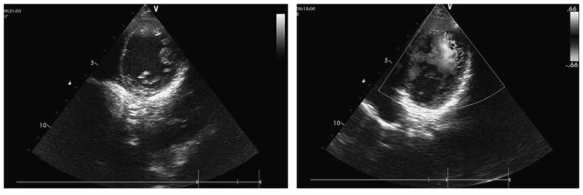
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Introduction: Maternal diabetes mellitus (MDM) is a risk for the health of both the pregnant women and her infant. It is reported that MDM increases oxidative stress and decreases antioxidant enzyme activities. Cardiomyopathy is noted in up to 40% of infants of diabetic mothers, and the exact mechanisms responsible for it are unknown. The aim of this study was to compare between infants of diabetic mothers (IDM) and healthy control newborns as regards of ischemia-modified-albumin (IMA), cardiac troponin-T (cTnT) and N-terminal brain natriuretic peptide (NT-proBNP) levels as markers of cardiac dysfunction.
Method: A prospective, comparative study included 40 infants of diabetic mother (IDM) as patient group and 40 healthy full term neonates as a control group. Umbilical cord blood IMA, Troponin T and NT-pro BNP levels were studied. Echocardiographic and electrocardiographic parameters were recorded in the first day of life.
Results: Interventricular septal thickness were higher in IDM group ($p < 0.05$). Myocardial performance index were significantly decreased in the IDM group ($p < 0.05$). The cord blood IMA, cTnT and NT-proBNP levels were higher in the IDM group compared with healthy control newborns, and the difference was statistically significant for IMA and NT-proBNP. IMA levels in IDM and control group were 225.52 ± 92.39ng/ml and 147.05 ± 87.41 ng/ml respectively ($p < 0.05$). Cardiac troponin T levels were 119.80 ± 45.59 pg/ml and 102.55 ± 42.65 pg/ml without significant difference ($p > 0.05$). Cord blood NT-proBNP levels were 2886.05 ± 4280.66 pg/ml and 956.30 ± 450.05 pg/ml in IDM and control group respectively.
Conclusions: This study demonstrated elevated IMA and cardiac markers in asymptomatic infants of well controlled diabetic mothers, representing the subclinical cardiac findings.

P1-58
Barth Syndrome and Left-Ventricular Non-Compaction: Case Report

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Introduction: Barth syndrome (BTHS) is an X-linked recessive disorder characterized by cardiomyopathy, skeletal myopathy and cyclic neutropenia in male patients. It is caused by mutations in the TAZ gene coding for the tafazzin, a protein involved in the remodeling of cardiolipin. We report a case of BTHS confirmed by TAZ gene analysis.
Methods: Three years old boy presented recurrent episodes of respiratory distress, progressive muscle weakness, hypotonia and growth failure. Family history was remarkable for 2 male siblings who died because of undetermined etiology.
Results: Hematologic and metabolic studies revealed that the patient had neutropenia, lactic acidemia, and 3-methylglutaconic aciduria; the diagnosis of BTHS was suspected. Echocardiography revealed mild dilated cardiomyopathy, hypertrophied and spongiform trabeculated pattern of the left ventricular posterior wall (ejection fraction = 58%, shortening fraction = 28% (Fig. 1). Electrocardiogram showed sinus rhythm with QTc duration of 460 ms, so propranolol 2 mg/kg was started and implantable cardioverter defibrillator fitted. Arrhythmia and pause not observed in the 24-hours rhythm Holter monitoring. The patient remained well at follow up 6 months later.
Conclusion: This case highlights that the diagnosis of BTHS should also be suspected in patients with left ventricular non compaction cardiomyopathy.



P1-59
The preparticipation screening in young trainers: What do we need exactly?

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Objectives: Studies have indicated that preparticipation screening of trainers may reduce the incidence of sudden cardiac death. The purpose of this study was to assess the utility and feasibility of a comprehensive cardiac screening protocol in young trainers before participation to sports.
Methods: A total of 380 young trainers refering pediatric cardiology clinic before participation to sports, between April 2014 and April 2015 were included in this study. The mean age was 12.4 years.

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A standart screening protocol has been applied to all, including personel and family history, physical examination, 12-lead electrocardiography (ECG), transthorasic echocardiography (TTE), 24-hour rhythm Holter analysis and exercise testing.
Results: The most frequent complaints were chest pain in 19 (5%) and dyspnea in 13 (3.4%) on exercise, dizziness and fainting in 5 subjects (1.3%). There was sudden death and arrhythmia in 41 subjects (10.7%) at family history. A heart murmur was present in 20 (5.6%) and hypertension in 10 subjects (2.6%) on physical examination. The 12-lead ECG was abnormal (right bundle branch block, ventricular hypertrophy, long QT, extra-systole) in 9 subjects (2.4%). The TTE was normal in 328 (86.3%) and mildly abnormal (rheumatic or structural valve disease, valve regurgitation, septal defect, valve stenosis) in 47 subjects (12.3%).
In 5 subjects (1.3%) a structural heart disease (septal defect, ventricular hypertrophy) was detected by TTE. 24-hour rhythm Holter analysis was abnormal (non-sustained VT, sinus pause >2.5 s, frequent extra-systole) in 6 (1.5%) and mildly abnormal (extra-systole in mid-frequency) in 4 subjects (1%). There was significant ST changes in 2 subjects (0.5%) on exercise testing with normal findings on myocardial perfusion scans. Adding 12-lead ECG, TTE, 24-hour rhythm Holter analysis and exercise testing to medical history, family history and physical examination identified a potentially serious condition in 70 subjects (18.4%).
Conclusions: This study demonstrated no relation between findings of screening protocol and 24-hour rhythm Holter analysis or exercise testing results ($p>0.05$). Therefore, preparticipation screening in young trainers should consist of a targeted personal history, family history and physical examination. The other tests should be applied only if the screening indicates the presence of a cardiovascular disease.

P1-60
An unusual presentation of carcinoid syndrome

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Case report: Thirteen years old male patient has admitted to oncology department for tumor that made extensive liver metastases with an unknown primary origin. As a result of investigations it was diagnosed as a neuroendocrine tumor.
Repeated echocardiographic studies have showed progressive right ventricular failure, loss of coaptation and thickening of the tricuspid valve, moderate pulmonary stenosis and hypoplasia. Primary or metastatic cardiac involvement was not observed in whole body scintigraphy scan. Therefore, these symptoms were thought to be due to circulating neurotransmitters rather than primary tumoral invasion. Tricuspit biological valve replacement and pulmonary valvotomy were performed. Aggressive anti-tumor treatment was started simultaneously.
Discussion: Carcinoid tumors are neuroendocrine tumors which usually originate from appendix and terminal ileum. Carcinoid syndrome is a clinical entity characterized by episodic flushing, secretory diarrhea, hypotension and bronchospasm linked to neurotransmitters. Cardiac involvement is observed in 50-60% of patients with carcinoid syndrome. Carcinoid heart disease is the most important cause of morbidity in patients with carcinoid syndrome. It is speculated that serotonin is the main neurotransmitter responsible for the carcinoid heart disease. Particularly activation of 5-HT2A give rise to myofibroblast deposition and accumulation of extracellular matrix on valves surface. Thereby possibility of valvular proliferation and fibrosis may increase. Due to high level of neurotransmitters are metabolized in the lung, the primary target of the carcinoid syndrome are right heart valves. However, 10% of those can be found with left heart involvement. Although carcinoid tumors have a slow progression; presence of cardiac involvement significantly decreases the life expectancy and quality. The diagnosis of this disease might be difficult and requires high index suspicion. Medical therapy, is performed to reduce the tumor burden, neurotransmitter activity and heart failure symptoms. In resistant cases valve surgery is preffered.
Conclusion: Early diagnosis and treatment is important in carcinoid heart syndrome.

P1-61
Blood Pressure Monitoring during Follow-Up in Coartated Pts after Percutaneous Therapy

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Background: Aortic isthmic coarctation is only one part of an arteriopathy with lifelong implications that persist after initial correction of the aortic obstruction. Long-term follow-up after successful repair show that life expectancy remain reduced, mostly due to ARTERIAL HYPERTENSION. Hypertension may be associated with anatomical and functional changes in the arterial tree. Early detection and treatment of AoCo is associated with the best outcomes, however, some patients will develop hypertension despite repair.
Methods: from 2000-2015, 178 pts underwent to angiography procedure and transcatheter treatment. Population study was divided in: 58 native (28.1%), and 128 re Coa(71.9%), 64% males and 36% female. Median age at first procedure is ± 6.9 years. Mean FU is ± 5.9 years. All patients underwent to Echocardigraphic exam, studied in cath lab, and blood pressure was evaluated with office monitoring, 24 h-Holter and exercise testing.
Results: all patients showed a decreased gradient peak to peak both in cath lab and in echo lab. At FU ± 5.9 years : 77% of patients were hypertensive after transcatheter therapy . So of this 38% were hypertensive at AMPB monitoring. 39% of patients had masked hypertension: 14% showed a BP no dipper at 24 hours Holter and 25% had an hypertensive answer at ergometric exercise. Data also have highlighted that the N of antihypertensive drugs increased from discharge to last assestment.
Conclusion: Hypertension is endemic in patients with stented coarctation, irrespective of the absenceof residual obstruction. Due to abnormal BP homeostasis, hypertension should be aggressively pursued by ABPM assessment and exercise stress testing in this population. Relative hypoplasia of the stented arch after somatic growth may contribute to this tendency and should provoke considerationof elective serial redilation of coarctation stents. Arterial hypertension is the most common complication in the COA correction in cath lab. Our data are totally in agreement with literature.

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P1-62

Outcomes of late presentation of Coarctation of the Aorta at a tertiary paediatric cardiology centre

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Introduction: Aortic coarctation accounts for 7% of congenital heart disease, and generally presents in neonates with collapse/ heart failure or in older children as hypertension/ incidental finding. We aim to investigate outcomes of late presentation of aortic coarctation.
Methods: A retrospective, observational study of children >3 years with isolated aortic coarctation (± - bicuspid aortic valve) between 2000-2015 from a tertiary paediatric cardiology centre.
Results: 170 children (0-16 years) diagnosed with isolated aortic coarctation; 47 (28%) of these patients were >3 years. Median age of delayed presentation was 8 years; 77% were male. Twenty-three patients (49%) had bicuspid aortic valve. Hypertension & murmur were the commonest presentations accounting for 70% of referrals. On review, 86% had a murmur, 89% had reduced/ absent femoral pulses. All patients with hypertension were on medication, majority beta-blockers (79%).
Of those with documented echocardiograms (n = 46), 76% had visible coarctation, with 11% having features suggestive of coarctation. The remaining 13% couldn't be adequately diagnosed with echocardiography. 55% had cross-sectional imaging, the majority having MRI. Of those that had chest radiographs, 52% had evidence of rib-notching. 22% had abnormal renal function, possibly representing abnormal renal perfusion.
Thirty-eight patients (81%) underwent cardiac catheterisation with balloon/ stent dilation of coarctation, of which 4 needed cardiac surgical intervention later. Seven patients had a primary surgical repair, of which 4 had prior diagnostic cardiac catheters. Two patients needed no intervention. Of the patients undergoing interventional cardiac catheterisation, 11 needed repeat interventions with balloon/ stent dilation, on average twice. No patients needed repeat surgical intervention.
Only one patient (catheter) had procedural complication, with balloon rupture. No patients had evidence of spinal ischaemia or permanent renal pathology, and there was no cardiac-related mortality.
Conclusions: This series shows a proportion of aortic coarctation present late, with hypertension/ murmur being the main finding. Weak femoral pulses can be a useful adjunct to diagnosis. Cross-sectional imaging is necessary as echocardiography does not always sufficiently image the arch and descending aorta in older children. Catheter intervention is a viable alternative to surgery; there is a need for further catheter intervention and this should be considered when counselling families.

P1-63

RSV prophylaxis in heart disease – indication and limits of worldwide administration

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Background: Respiratory syncytial virus is a common pathogen affecting almost all children by the age of 2. The morbidity from such lower respiratory tract infection results in 20% of those with haemodynamically significant congenital heart disease (hsCHD) being admitted to hospital during the winter season. Palivizumab (PVZ) is the standard immune prophylaxis, in the absence of a vaccination, and we wished to understand global variations in its use.
Methods: A steering committee, with 1 clinician from 5 countries (England, Spain, USA, Colombia, Japan), devised a set of questions, using Delphi methodology, concerning PVZ usage. We determined the importance of these questions with an extended faculty with one clinician (including paediatric cardiologists, intensivists and cardiac surgeons) from each of 7 countries (Germany, Spain, Korea, Taiwan, UAE, USA, Mexico). Evidence based answers were obtained with regional variations for use of PVZ prophylaxis in each type of cardiac disease and additional indications and determined the resistance to use.
Results: There was agreement on many items, such as the main indication that hsCHD in the first 12 months of life included those on medication for left to right shunt, those with cyanosis (oxygen saturation < 85%), those with cardiomyopathy or pulmonary hypertension on treatment and those on transplant waiting lists. There was divided opinion on the evidence for use after 1 year old, in nosocomial outbreaks, immune deficiencies and those being admitted for cardiac intervention. The paediatric cardiologist usually determined patient selection, there was no evidence of benefit after 2 years of age, in those with arrhythmia or those with co-morbidities conferring no independent risk. In some countries use of PVZ was restricted by access to funding, in others there was lack of clinician knowledge concerning indications. In subtropics, it was both access to the prophylaxis and the lack of a clearly defined RSV season, with cases appearing all year round.
Conclusion: There is general consensus that PVZ prophylaxis should be used in hsCHD under 1 year and that it should be available more freely in resource limited countries. There is still insufficient evidence to guide its use in subtropical countries and in certain co-morbidities.

P1-64

Ethnicity and seasonality in a national 2 year study of Kawasaki Disease

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Background: Kawasaki disease (KD) is the commonest cause of paediatric acquired heart disease in the Western world. The incidence is doubling every 15 years. The risk in ethnic groups is unclear and each country has seasons with varying incidence. We wished to understand risk factors in our population and to determine our current incidence and rate of complications.
Methods: We undertook a national survey of cases of Kawasaki disease from January 2013 to February 2015 via the British Paediatric Surveillance Unit. The survey was sent to all paediatricians and paediatric cardiologists in the UK and Ireland. It included questions on demographics, ethnicity, and seasonal of incidence in addition to treatment and complications of Kawasaki disease especially cardiac disease.
Results: 600 notifications of KD were received. Of these 552 were cases of complete (n = 388), atypical (n = 46) and incomplete (n = 116) with 2 deaths. There were 38 duplicates and 10 were

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excluded with alternative diagnoses. There were 215 girls, 322 boys (1:1.5), 15 with gender unreported with no difference in the sex ratio between complete, atypical and incomplete forms. The peak incidence was in January of all 3 years, with lowest incidence September to November. Age at diagnosis ranged from 2.5 months to 15 years (30% after 5 years). Whilst the majority of cases were Caucasian (67.6%), Chinese and Japanese Asians were over represented compared to the normal incidence in the population (3.8% v 0.8%). In addition we found that there were more Black African children with KD than expected from the healthy population (7.2% v 2.3%). 94.5% received intravenous immunoglobulin but despite this, 28% had coronary artery dilation or pericardial/myocardial involvement. Subgroup analysis showed that certain ethnic groups are at increased risk of specific complications.
Conclusions: In this large population based study; we showed a high incidence in children of black African origin and of coronary artery involvement despite treatment. Any under-recognition of this disease and education of parents and clinicians should follow in order to minimise the future health burden of Kawasaki disease.

P1-65
Complications after pediatric heart transplantation – 9 years single centre experience
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Background: Complications are the major determinants of long term survival after pediatric heart transplantation (HTX). In 2007 a pediatric heart transplant program has been established in Budapest, Hungary. Our aim was to summarize our experience concerning complications after pediatric HTX.
Patients and methods: Since 2007 29 children were transplanted at the Pediatric Cardiac Center, Budapest. They were 7.1 (5.7) years old at the time of (HTX). Seven patients required ventricular assist device prior HTX. After induction, initial immunosuppressive therapy was tacrolimus and mycophenolate mofetil. Regular controls were timed every 2-6 weeks, mean follow up was 3.4 (2.1) years.
Results: Major complications were infections, gastrointestinal and hematological diseases. Posttransplant infections included Clostridium difficile-related enteritis in 4 patients, CMV infections (1 lethal pneumonitis, 2 pts with treated hepatitis) in 3 patients, mild, but recurrent upper respiratory tract infections in 4 patients, severe fungal infection (immediately after HTX) in 1 patient, lethal myocarditis in 1 patient. Gastrointestinal complication: 10 of 29 patients had abdominal pain and enteritis. It was due to infective diseases in 4 cases (in 2 patients enteral symptoms persisted even after eradication of C. difficile). One patient has de novo inflammatory bowel disease. In 6 patients GI problems were mycophenolate related, they were converted to everolimus, however in one patient everolimus had to be withdrawn. Haematological complications consisted of drug related leukopenia in 2 cases and autoimmune haemolytic anaemia (AIHA) in 2 patients. One patient with thrombocytopenia associated AIHA was steroid sensitive. The other AIHA patient had severe, steroid resistant haemolysis (AIHA persisted even after rituximab and plasma exchange). One patient was lost due to treatment resistant post transplant lymphoproliferative disease (PTLD). Controlled hypertension was present 11 patients, 6 patients needed treatment for dyslipidaemia.
Conclusion: Our observations underline the importance of infection control in the long term survival after HTX as 50% of posttransplant mortality was due to infective diseases. Drug related diseases are of mild-to-moderate intensity, but treatment may be difficult. Autoimmune diseases are present at around 10%, but due to posttransplant immune dysregulation, effective treatment could be problematic.

P1-66
The Prevalence of Congenital Heart Defects in Infants with Cholestatic Disorders of infancy: A single center study
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Introduction: There is deficiency of data about congenital heart defects (CHD) in cholestatic disorders of infancy other than Alagille syndrome. There are many conditions that cause neonatal cholestasis, the most commonly identifiable are biliary atresia (BA), genetic disorders, metabolic diseases, and α1-antitrypsin deficiency. BA could be associated with other congenital anomalies.
Aim of work: To define the prevalence and types of CHD in infants with various causes of cholestatic disorders of infancy.
Methods: This cross sectional study was conducted on 139 infants presenting with cholestasis whether surgical or non-surgical. The study was carried out at the Pediatric Hepatology Unit, Cairo University Children's Hospital, Egypt. Full examination and investigations were done in an attempt to reach an etiologic diagnosis for cholestasis; in addition to a comprehensive echocardiographic study.
Results: The age at the onset of cholestasis ranged from 1 day to 7 months. Males constituted 61.2%. BA was diagnosed in 39 patients (28%), AGS in 16 patients (11.5%), 27 patients had miscellaneous diagnoses and 57 cases had indeterminate etiology. CHD were detected in 55 patients (39.5%). Shunt lesions were detected in 24 patients; 43.6%, pulmonary stenosis in 18 patients (32.7%) and combined lesions in 9 cases (16.4%). Three patients (5.5%) had abnormal cardiac situs. Only seven patients had clinical presentation suggestive of CHD. CHD were detected in 14 patients with BA (35.9%), 15 patients with AGS (93.7%) and 26 patients in the remaining group (30.9%). Among the 39 patients with BA, 14 (35.7%) had CHD: 9 patients (23%) had shunt lesions, the most common cardiac anomaly was PDA in 5 patients (12.8%) and the second most common was ASD secondum in 3 patients (7.7%). Only 3 patients had major congenital cardiac defects (7.7%) with two of whom having situs inversus associated with cardiac defects (5.1%).
Conclusion: CHD are not uncommon among cholestatic infants other than AGS. Echocardiography is recommended as part of the preoperative assessment infants with BA before undergoing hepatic portoenterostomy to exclude presence of CHD which may impact the anesthetic planning, timing of surgery and the outcome of hepatobiliary surgery.

P1-67
Incomplete Kawasaki disease, is it time for modified criteria?
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Introduction: The diagnosis of Kawasaki disease (KD) in the absence of the classical criteria is a big challenge especially that features are non-specific. The terms 'atypical' and 'incomplete' have been used

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to describe such cases and guides the administration of intravenous immunoglobulins (IVIG).
Patients and Methods: We analyzed the data of 4 patients who were diagnosed with KD not fulfilling criteria for incomplete KD from 02/2014 to 05/2015.
Results: The first was a 4 months-old boy presenting with generalized mucopapular rash, purulent conjunctivitis, lip fissuring and 1 day of fever. His left coronary system was ectatic initially, that progressed to aneurysmal dilatation of the circumflex artery (CX). Symptoms responded to IVIG after an initial episode of severe anaphylaxis. He continues to have CA aneurysm and is on anticoagulation.
Second was a 5 year-old boy with a 20-day history of fever without any other manifestations, his left anterior descending artery (LAD) had a fusiform dilatation at its midportion. Fever resolved following IVIG administration and CA dilatation regressed in 2 months.
Third was a 4 year-old boy presenting with a 7-day history of fever associated with nonpurulant conjunctivitis, follicular tonsillitis, with no other criteria of KD. Both coronary systems were dilated. Responded to IVIG and his coronary dilatation regressed within 1 month.
Fourth is a 2 year-old boy presented with 5 days of fever and generalized lymphadenopathy, hepatomegaly, and marked elevation of transaminases. Pericardial effusion was discovered on work-up, and on follow-up the LAD became ectatic and finger-pealing appeared. Fever responded to IVIG, effusion resolved in 3 days and coronary dilatation regressed in 2 weeks.
Conclusion: We report 4 cases diagnosed with KD despite insufficient criteria: typical fever being absent in one, fever was the only manifestation in another, a third with only 2 criteria in addition to fever and a fourth with generalized lymphadenopathy as the predominant feature besides fever. All of which had CA dilatation and responded clinically to IVIG, with the persistence of large coronary aneurysm in one case. We suggest that the criteria for KD be modified to a scoring system including major and minor criteria.

P1-68
Cerebral function changes in children with congenital heart disease

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Introduction: Infants with congenital heart disease (CHD) as well as Survivors of cardiac surgeries are at risk for brain injury and neurodevelopmental defecits. It is thus necessary to evaluate and follow up cerebral functions by the use of electroencephalogram (EEG) in patients with CHD.
The aim of this work is to evaluate the cerebral function in children with congenital heart disease using aEEG to detect abnormalities in the cerebral function and to correlate this to the oxygen saturation.
Patients and Methods: We performed digital EEG and brain mapping in 60 children (28 (46.7%) males and 32 (53.3%) females) with CHD (40 (66.7%) with cyanotic CHD (%) and 20 (33.3%) with acyanotic CHD (%)) as well as 40 age and sex matched healthy controls.
Results: The patients' mean age was 39.8 ± 15.9 months, and the mean weight was 9 ± 5.7 kg. There was no significant difference between patients and controls regarding the age but patients had significantly lower anthropometric measures (weight, height and BMI) with $p < 0.0001$, $p < 0.0001$ & $p = 0.001$ respectively.
14 (23.3%) patients had prior cardiac surgery, of them 8(57%) had open heart surgeries & 6(43%) had closed heart surgeries. 38(63.3%) of our studied patients had pulmonary hypertension.
EEG findings were present in 30 (50%) patients (28 patients with cyanotic and 2 with acyanotic CHD) while all controls had normal EEGs. Detected EEG changes were: Epileptic activity in 2(3.3%) patients (both with cyanotic CHD), Background activity slowing in14 (23.3%) patients (all with cyanotic CHD), while both epileptic changes &back ground activity slowing were detected in 14 (23.3%) patients (12 with cyanotic and 2 with acyanotic CHD). The cut off value of O2 saturation below which epileptic changes are liable to occur was 73.5%, while that for back ground activity slowing was 77.5%.
Conclusion: Children with cyanotic CHD had significantly higher EEG changes than those with acyanotic CHD. The aEEG should be performed routinely in patients with CHD (particularly cyanotic) to detect abnormalities in the cerebral function, allowing for early treatment.

P1-69
Age, Sex and Residence Features of Children with CHD and Different Toxic Metals Content in Cardiovascular Tissue

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Intoduction: There are about 30 metals and metalloids that are potentially toxic to humans. Some of them, according to the experimental data, able to penetrate the placental barrier and violent cardiogenesis.
Methods: We had determined content of toxic metals (TM) in 107 biosubstrates of heart and great vessels of 55 patients with CHD: 34 boys and 21 girls (males in 1.6 times more often than females); 2 fetuses, 6 newborns, 24 children of 1-12 months, 15 pts. – 1-5 years, 8 pts. – older 5-17 years; 39 patients were resident of industrial regions and 16 – agricultural. All patients were examined by the spectral analysis of TM content in heart and great vessels tissue by methods of the atomic emission spectrometry in the inductively coupled plasma and atomic absorption spectrometry with electrothermal atomization.
Results: the presence of 13 toxic metals and metalloids (Al, Ba, Li, Ni, Sb, As, Sr, Pb, Zr, Sn, Ti, Ag, W) was documented in the tissues of the heart and great vessels of patients with CHD, while in each age group their number was different (8 to 11). The greatest quantity of toxic metals in pathological concentrations (Ba, Ni, Li, Sb) was found in fetuses and newborns in comparation to children older than one year of age. Was surveyed that 97.1% of boys had toxic substances pathological concentrations in cardiovascular tissue that significally higher than girls – 71.0%, $p < 0.05$. The most often in the tissues of the heart and great vessels was documented excess of barium permissible concentration - in 75.8% of boys, that is significant more often, than girls (47.6%), $p = 0.035$. Residents from industrial regions had more toxic metals and higher average concentration of strontium, zirconium, tungsten, silver and tin in cardiovascular tissue in comparison

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with children from agricultural, $p \leq 0.05$. Only patients that lived in industrial regions had pathological concentration of arsenic and antimony.
Conclusions: Our results confirmed pathological possible impact of toxic substances on cardiogenesis violation and a higher risk of cardiovascular malformation developing in males and residents of the industrial region.

P1-70

A novel simulation-based approach to train study teams for clinical trials in neonates, infants and children with heart failure

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Introduction: Based on the need of an orally administered age-appropriate enalapril formulation for use in children, the EU's Seventh Framework Programme (FP7)-funded research project LENA (Labeling Enalapril from Neonates up to Adolescents; grant agreement n°602295) was initiated. Within the project, paediatric clinical trials assessing pharmacokinetic (PK), pharmacodynamics (PD) and safety data have to be performed in neonates, infants and children. In addition to challenges involved in every paediatric drug development, the strict framework of FP7 adds further time pressure and budget limitations. To meet these particular challenges, a novel approach for the training of study teams was chosen that goes beyond current standards. A simulation training was used to improve the study teams' skills in PK/PD investigations and patient recruitment. This intense, focused training allows study teams to practice critical situations under realistic circumstances. Thus it is expected to reduce preventable sampling and recruitment failure. This hypothesis is subject to a systematic evaluation study.
Methods: Small-volume sampling of time-critical and sensitive parameters as well as communication to potential participants, parents and colleagues were identified as most critical hurdles in the trials. Thus, simulation scenarios for the training of these skills have been implemented using simulation manikins and original medical devices. All study teams attended a two-days-training. Video-based debriefing of the scenarios enriched the learning experience. Participants' performance and preparedness for the study as well as the usefulness of the training were assessed using surveys based on five-point Likert scales.
Results: 23 participants from five different European countries were trained at the Salzburg simulation centre. The performance in sampling of time-critical humoral parameters was optimised to meet the predefined time limits, and to enable maximum reliable data extraction by reducing invalid samples. Communication scenarios allowed improving of critical communication skills. Video-based debriefing facilitated self-reflection and joint discussion. Participants' abilities to communicate core elements of the studies and to successfully perform PK/PD sampling increased significantly ($p = 0.0003$).
Conclusions: Simulation training significantly improved the participants' performance. This tailored training was assessed as a helpful teaching tool in trial preparation. Further follow-up surveys will assess the actual impact of this training on the study success.

P1-71

Effect of cycle exersice on regional oxygen saturation in Failing-Fontan patients

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Introduction: Failing-Fontan patients suffer from increased morbidity and mortality, while pathophysiologic changes leading to a failing still remain unknown. We investigated differences in regional oxygen saturation (rSO2) by near infrared spectroscopy (NIRS) in Failing-, Non-Failing Fontan and biventricular (TGA) patients during ramp incremental cycle exercise, following the hypothesis that rSO2 is lower in Failing-Fontan patients, which could contribute to the development of a failing.
Methods: 38 patients underwent ($n = 7$ Failing Fontan, $n = 22$ Non-Failing Fontan and $n = 9$ TGA patients) incremental ramp cycle exercise under continuous blood pressure, heart rate and ECG-surveillance. The average age was 16.5y ($\pm$ 5.57). NIRS was measured with 4 electrodes (kidney, frontal cortex, quadriceps femoris and triceps brachii) during the exercise, a 5-minute recovery period and during one hour physiologic regeneration by a portable NIRS monitor. Absolute and relative (from baseline) were calculated.
Results: Failing-Fontan had significantly shorter duration of exercise than Non-Failing or TGA patients ($p < 0.01$), less watts per kilogram ($p < 0.01$), a higher restring heart rate ($p = 0.01$), a lower maximum heart rate ($p < 0.01$) and lower resting and maximum blood pressure ($p < 0.01$).
Failing and Non-Failing Fontan showed significantly lower baseline in cerebral oxygen saturation than biventricular patients (66.2 and 69.9 vs. 76.7 $p = 0.03$/$p = 0.04$) and Failing patients showed a trend of lower renal oxygen saturation than TGA patients (64.3 vs. 82.8; $p = 0.09$).In musculature rSO2 was significantly lower in triceps brachii for Failing patients compared to TGA (61.2 vs. 72.1; $p = 0.02$), in quadriceps femoris absolute values of rSO2 were lower in Failing ($p = 0.09$) and Non-Failing but didn't pass the significant threshold. Failing patients showed a significantly ($p < 0.01$) faster decrease of rSO2 according to exercise time than Non-Failing or TGA patients. On the other side TGA patients showed a significantly faster regeneration regarding rSO2.
Conclusions: This is the first study demonstrating regional oxygen measurements in Failing-Fontan patients under cycle ergometry, showing that Failing-patients present with significant decreased rSO2 in rest and under exercise compared to Non-Failing and especially biventricular patients. Decreased rSO2 univentricular patients compared to biventricular controls demonstrates low output of the single ventricle. The effects of chronic desaturation in Failing-Fontan patients need to be discussed and further evaluated.

P1-72

How much IVIG dose is optimum for low risk cases of Kawasaki disease?

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Background: In the guideline from the American Heart Association of 2004, patients of Kawasaki disease (KD) should be treated with 2 g/kg IVIG in a single infusion for initial treatment. In Japan, more than 80% KD patients receive 2 g/kg IVIG in a single

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infusion. In the Japanese Society of Pediatric Cardiology and Cardiac Surgery guideline of 2012, administration of 1 g/kg IVIG for 1 or 2 days is also approved as a modified procedure. In fact we often have experience of success in only 1 g/kg IVIG. Objective: To evaluate efficacy and safety of starting initial treatment with 1 g/kg IVIG for low risk cases. Subjects and Methods: We defined those who have low scores in the three major Japanese prediction scoring systems as a low risk patient. From January 2012 to October 2015, a total of 108 hospitalized low risk patients excluding those patients who did not receive IVIG (n = 6) were enrolled. The patients were separated into two groups. Group 1 (n = 78): who started initial treatment with 1 g/kg IVIG and Group 2 (n = 30): who were administered 2 g/kg IVIG in a single infusion. Clinical data, clinical course, and circumstances of additional therapies were compared. Results: There were 115 low risk patients out of the total 213 KD patients (54%). There were 47 patients who finished treatment with only 1 g/kg IVIG in Group 1 (60%). There were 8 patients who required another 1 g/kg IVIG more than 2 days after (10%), and 23 patients who required 1 g/kg IVIG for 2days in a row (29%) in Group 1. There were 8 patients in Group 1 who required additional treatment (10%) and 4 patients in Group 2 (13%). The day of discharge of those who required a total 2 g/kg IVIG for initial treatment in Group1 was day 13 which was almost the same as that in Group 2. No patient had coronary region. Conclusions: More than half of KD patients are low risk cases. More than half of patients do not need 2 g/kg IVIG in low risk cases. It may be effective and safe to start initial treatment with 1 g/kg IVIG for low risk patients.

P1-73

Outcomes and Risk Stratification of Tricuspid Atresia in Scotland: A 15 year Retrospective Review

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Introduction: Tricuspid atresia can occur in isolation or in association with other congenital cardiac abnormalities, for example: aortic arch abnormalities; ventriculo-arterial (VA) discordance; varying degrees of right ventricular hypoplasia; pulmonary stenosis and pulmonary atresia. The presence or absence of associated abnormalities influences initial and subsequent surgical options.
Objectives: To report the associated cardiac lesions in our population with tricuspid atresia and assess whether the presence or absence of such lesions alters the long term prognosis.
Methods: A retrospective analysis of 40 consecutive cases of tricuspid atresia diagnosed in the national congenital cardiac centre from January 1st 2000 – December 31st 2014.
Results: A diagnosis of tricuspid atresia was confirmed in 40 children who were live born during the study period, 21 females and 19 males.
Morphology in association with tricuspid atresia included: 22 (55%) with concordant VA connections; five (12.5%) with discordant VA connections; two (5%) with concordant VA connections and an associated aortic arch abnormality; four (10%) with discordant VA connections and an aortic arch abnormality; three (7.5%) with pulmonary atresia and four (10%) with pulmonary stenosis.
Three patients died within the first month of life without surgical intervention; three died post shunt procedure and one died post pulmonary artery band, secondary to associated ventricular failure. 17 patients required a shunt procedure, of which three (17.6%) died post procedure and prior to Glenn palliation. The presence of pulmonary atresia in association with tricuspid atresia resulted in 100% mortality in the 1st year of life. 33 (82.5%) patients survived to Glenn palliation.
There was only one (2.5%) reported case of plastic bronchitis and no reported cases of protein losing enteropathy (PLE) or heart transplantation. There were no deaths after 9 months of age and 33 children are alive to date. Survival to 5 and 10 years was 81%.
Conclusions: Survival in the Scottish population is similar to international published rates of survival following palliation of tricuspid atresia. The presence of associated cardiac lesions allows paediatric cardiologists to risk stratify tricuspid atresia at diagnosis, predicting the initial surgical pathways and ultimately provide parents with a more accurate long term outlook.

P1-74

Post-transplant lymphoproliferative disease (PTLD) among heart transplanted children in Gothenburg 1989-2014 (25 years)

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Introduction: Heart transplantation has been an option for children in Sweden since 1989. The immunosuppressive treatment, a prerequisite for transplantation, carries an increased risk for malignancies, the most common type being lymphoma, post-transplant lymphoproliferative disease (PTLD). As our institution faced an increased rate of PTLD among heart transplanted children, the objective of this study was to analyze the rate of PTLD over time and to identify possible risk factors.
Methods: This is a retrospective study of all heart transplanted children (0-18 years of age) in Gothenburg from 1989 to 31 December 2014.
Result: A total of 71 children underwent heart transplantation. The overall incidence of PTLD was15% (11/71), however 20% (9/44) of those being heart transplanted after 2001 developed lymphoma, compared to 7% (2/27) transplanted before 2001. Median age at transplantation was 10 years (0-17), equally for subjects developing PTLD, post-transplant follow-up time was 10 years (0-25) for those who did not develop PTLD, compared to 6 years (0-21) in the PTLD-group. In the group that developed PTLD, listing diagnosis was exclusively surgically palliated congenital heart defects with an overweight for hypoplastic left ventricle. Induction therapy with ATG-Fresenius was twice as common among those who developed PTLD and the majority (7/9) was seronegative for EBV at heart transplantation.
Conclusion: The incidence of PTLD was 15% (11/71), with a tendency to increase during later years. All subjects developing PTLD had congenital heart defects, the majority hypoplastic left ventricle, and all had undergone sternotomy before transplantation.

P1-75

Possible superiority of angiotensin II type 1 receptor blockers in patients with Marfan syndrome. Do we have a dose-related effect?

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Introduction: Dilatation of sinus of valsalvae (SV) is the most common pathology in Marfan syndrome (MFS). To prevent aortic complications, initiation of medical therapy is often necessary. Due to pathogenesis of MFS medical therapy with angiotensin II type 1

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receptor blockers (ATII) was expected to be highly effective. Now first published large randomized trials demonstrate that ATII is not superior to prophylaxis with betablockers (BB).
Nevertheless dosing of ATII in childhood is variable and has not been investigated yet. To ensure best impact on connective tissue, different doses may be necessary and could potentially prove that ATII is superior to BB.
Methods: Since 2008 we investigated 343 patients (11.12 ± 5.71 y) with suspected MFS whereas 132 patients were diagnosed. Thereby 82 showed indication for prophylactic treatment (n(ATII) = 56, n(BB) = 26)). To evaluate dose related impact we retrospectively divided patients in ATII groups (A:0.5-1.0, B: > 1.0-1.5, C: > 1.5 mg/kg/d) to measure progression of SV dilatation (Z-score) via echocardiography. The ATII groups showed no difference regarding age, gender and indication for therapy.
Results: Mean follow up period in group A(n = 31), B(n = 38) and C(n = 19) after initiation of ATII was 29.88 ± 21.24 months (BB 48.10 ± 43.35). Z-score decreased in all groups after follow-up period (Table1). Using linear regression rate of change between the three groups with different ATII dose did not show linear correlation.
Conclusions: This selective and non-randomized collective indicates a tendency towards the superiority of ATII prophylaxis in children with MFS. In contrast to recently published data therapy with ATII seems to be more effective, but there is no proof for a dose related effect. And it seems that non-responders to ATII exist as enhancement of dose does not correlate with decrease of SV dilatation. To estimate effective dosing and reasons for possible non-responding further, larger pharmacokinetic and pharmacodynamic studies are indispensable.

Table 1. Z-score of SV before (SV1) and after (SV2) treatment with BB, ATII, different ATII dose.

Group	Follow-up [months]	Z-Score SV1	Z-Score SV2	N	P (Z-score)
BB	48.10 ± 43.35	2.54 ± 1,70	2.14 ± 1.36	26	p = 0.067
ATII (overall)	29.88 ± 21.26	1.74 ± 1.03	1.31 ± 1.31	56	P = 0.001
ATII 0.5-1 mg/kg/d (A)	14.26 ± 12.01	1.71 ± 1.07	1.12 ± 1.45	31	P = 0.003
ATII 1-1.5 mg/kg/d (B)	08.32 ± 08.38	1.70 ± 1.03	1.48 ± 1.31	38	P = 0.060
ATII > 1.5 mg/kg/d (C)	14.68 ± 12.68	1.89 ± 1.04	1.82 ± 1.03	19	P = 0.567

P1-76
Neurological disorders in children with tetralogy of Fallot and pulmonary atresia with ventricular septal defect

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Background: Tetralogy of Fallot (TOF) and pulmonary atresia with ventricular septal defect (PA-VSD) are the most common cyanotic congenital heart disease (with right to left shunt). There is growing recognition and concern that patients with these heart defects are at increased risk of adverse neurodevelopmental outcomes. In fact, they can be frequently associated with ischemic stroke resulting from thromboembolism and others infectious or anoxic neurological complications.
Purpose: The aim of our study is to determine the frequency and nature of different neurological damage that can be associated with tetralogy of Fallot and PA-VSD.
Methods and Materials: We performed retrospective epidemiological study of neurological damage in infants with TOF and PA-VSD in the Pediatric department of Sahloul hospital (Sousse, TUNISIA) between January 1993 and June 2015.
Results: A total of 32 patients were enrolled in this study. 27 children were diagnosed with TOF and 5 children with PA-VSD. The mean age of diagnosis of the cardiac disease was 9.3 months. Only two children had a complete cure of their heart disease. Fifteen children (47%) had neurological damage, diagnosed at a mean age of 24 months; They were: 2 cases of brain abscess, 2 stroke cases, 2 cases of hydrocephalus, 1 case of cerebral thrombophlebitis, 1 case of association brain abscess and stroke, 2 cases of epilepsy and 5 children had only cognitive impairment. In 4 cases, the neurological involvement revealed the congenital heart defect. The neurological disease was confirmed by cerebral MRI in twelve cases and by brain CT in three cases. All patients had specific treatment of their neurological lesions. During follow-up 8 children had stabilization of their neurological diseases, 4 children died and 3 children were lost to seen.
Conclusion: Our study showed that these heart diseases are frequently associated with neurological complications if they are not early primary repaired. Neurological complications include essentially embolic stroke and brain abscess. Apart from these complications, neurodevelopmental impairment occurs in some children as sequelae to chronic hypoxemia. This neurological damage with different degrees of severity proves the vital importance of early surgical repair of patients with tetralogy of Fallot and PA-VSD.

P1-77
Cardiovascular Complications of Obesity in the Pediatric Population - Ten Years Case Study in the North East Region of Romania

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Introduction: Obesity in children has a strong impact on all systems and organs, causing metabolic and cardiovascular complications, both short and long term, significantly increase morbidity and mortality rate. The prevalence of obesity and overweight in children has increased dramatically in Europe, the number of cases tripling in the last 25 years. In Romania, the prevalence of obesity in children aged 3-17 years has increased considerably in the last 10 years, ranking it third in Europe.
The purpose of the study is to determine the relationship between overweight and cardiovascular complications in children and adolescents, and at the same time, to establish therapeutic management for both excess weight and to prevent or treat them.
Methods: In the study, we analyzed 464 children, aged 1-18 years, hospitalized for a period of ten years (January 2006 – December 2015) in the Pediatric Cardiology Department, 'Sfanta Maria' Children's Emergency Hospital of Iasi, Romania.
We observed age, sex, body mass index, blood pressure, biological VSR, serum fibrinogen values, total cholesterol and the results of the echocardiography and ophthalmological examination. All patients in the study group have performed a neuropsychiatric and psychologically exam.
Prospective echocardiographic measurements were performed in 464 obese children. Two-dimensional, M-mode and color M-mode ultrasound, conventional pulse wave Doppler imaging were used to assess cardiac function.
Results: We found increased blood pressure values in 21.3% of cases. Measurements of LV mass, LV wall thickness and

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LV end-diastolic diameter and volume were significantly elevated in 73 obese children (15.7% of cases).
VSR, fibrinogen and total cholesterol levels were found to be higher among female subjects (hypercholesterolemia in 15.9% of cases, and 40% of cases with increased plasma fibrinogen).
Conclusion: In this study, the occurrence of cardiovascular complications of obesity in the pediatric population was recorded. The echocardiographic exam confirms the elevated LV mass in obese children.
It is necessary to follow the periodic height and weight, as well as to evaluate blood pressure, total cholesterol, fibrinogen among children with obesity, in order to prevent cardiovascular complications and to include these patients in a cardiovascular risk group.

P1-78
Cardiovascular sequelae of Bronchopulmonary Dysplasia in Prematurely Born School Children with new echocardiographic methods
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Introduction: Bronchopulmonary dysplasia (BPD) is an important complication of prematurity and has long-term pulmonary consequences. Cardiovascular sequelae related to BPD have also been reported in severe BPD patients of the presurfactant era.
The aim of this study is to investigate cardiovascular consequences of BPD at school ages and to find out possible risk factors related to cardiovascular sequelae especially with new echocardiographic methods.
Methods: In our study totally 43 children (21 F/22 M: 23 aged 104 ± 7.4 months born preterm with BPD, 20 aged 112 ± 8.2 months born preterm without BPD and 21 healthy children aged 100 ± 16 months born at term (control group) were evaluated with conventional and myocardial tissue Doppler echocardiography. And also Left (LV) and Right ventricular (RV) strain and strain rate were assessed by speckle-tracking echocardiography.
Echocardiographic examination was performed using a Vivid 9 ultrasound machine, and LV and RV myocardial strain were analyzed offline using Echo PAC software.
Results: BPD patients had a decreased pulmonary artery acceleration time and higher left and right ventricular myocardial performance indexes in school ages. Longitudinal and circumferential strains were abnormal in premature with BPD group even in the presence of normal RV and LVEF.
Conclusions: Negative effects of BPD on global cardiac performances of both ventricles and pulmonary arterial pressure persist up to school ages.

P1-79
Does ketogenic diet have any negative effect on cardiac systolic and diastolic functions in children with intractable epilepsy?: One-year follow-up results
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Purpose: The ketogenic diet (KD) has been referred to as an 'effective therapy with side effects' for children with intractable epilepsy. Among the most recognized adverse effects, there are cardiac conduction abnormalities, vascular and myocardial dysfunction. However, very limited and controversial data are available regarding the effects of the KD on cardiac functions. We sought to analyze the mid-term effect of ketogenic diet on cardiac functions in patients with intractable epilepsy who received a ketogenic diet for at least 12 months using conventional and relatively new imaging techniques.
Methods: This prospective study included 61 patients with intractable epilepsy who received ketogenic diet for at least 12 months. Clinical examinations, serum carnitine and selenium levels as well as electrocardiographic and echocardiographic examinations were scheduled prior to the procedure and at 1, 3, 6 and 12 months. We utilized two–dimensional, M-mode, colored Doppler, spectral Doppler and pulsed wave tissue Doppler imaging techniques to investigate ventricular systolic and diastolic functions of this subgroup of patients.
Results: In our study, there was no significant difference after one year of KD therapy compared to baseline values—except a significantly decreased A wave velocity—in terms of pulse wave Doppler echocardiographic measurements of the diastolic function. The tissue Doppler measurements obtained from the lateral wall of tricuspide and mitral annuli were not different at baseline and at month 12 of the treatment, as well.
Conclusion: The ketogenic diet appears to have no disturbing effect on ventricular functions in epileptic children in the midterm.

P1-80
Knowledge base of adolescents with congenital heart disease
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Introduction: Most congenital heart disease [CHD] is diagnosed and treated in early childhood with parents acting as surrogates for decision-making and recipients of information. The adolescent assumes this role in preparation for transition to an adult congenital heart program. We performed an observational study to evaluate adolescent and parental recognition of their CHD.
Methods: Established clinic patients, ≥ 11 y.o. and their parents were independently surveyed prior to being seen. Participation was voluntary. Electrophysiology, cardiomyopathy and cardiac transplant patients were excluded. Results were scored as full, partial or incomplete recognition.
Results: Most (98% parents; 83% adolescents) reported that their cardiologist had provided education. Summary results are noted in Table I. Older adolescents performed better. Parents performed better than all patients, including those ≥ 18 y.o. History of prior surgery/intervention had no significance. Patients undergoing last surgery ≥ 11 y.o. scored better than those with surgery at younger age or without surgery ($p < 0.05$). Patients with complex lesions (single ventricle, tetralogy of Fallot, transposition of the great arteries) were better at providing the medical name of their defect ($p < 0.05$) but similar in ability to describe the anatomy when compared to those with repaired shunt lesions and mild unrepaired lesions. Those with more complex disease were better at providing the full name of the defect (81/134; 60%) than in fully describing it (43/134; 32%; $p < 0.005$).
Conclusions: Unlike their parents, only 1/2 of adolescents are able to name their cardiac defect and 1/3 are able to adequately describe their cardiac anatomies. Older adolescents and those with more complex defects are more knowledgeable. A stronger emphasis should be made on educating this population as they transition to adulthood.

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Table 1.

	Name defect (n = 522)		Describe defect (n = 521)	
	Full	Partial	Full	Partial
11-14 y.o	40% (n = 102)*	13% (n = 34)	23% (n = 58)*	35% (n = 89)
15-17 y.o.	51% (n = 87)*	17% (n = 29)	35% (n = 56)*	37% (n = 63)
> 18 y.o	68% (n = 65)*	16% (n = 15)	54% (n = 51)*	31% (n = 29)
TOTAL	49% (n = 254)*	15% (n = 78)	32% (n = 165)*	35% (n = 181)
Parent	78% (402/512)*	11% (n = 56)	61% (312/512)*	29% (n = 151)

*$p < 0.005$.

P1-81

A Rare Endocrine Causes Of Tachycardia: Refetoff Syndrome

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A Rare Endocrine Causes Of Tachycardia: Refetoff Syndrome
Introduction: Thyroid hormone resistance (Refetoff syndrome) is a syndrome that caused by the decrease in susceptibility to thyroid hormone in end-organs. The incidence was reported as one in 40,000 live births. Mutations in thyroid hormone receptors play role in the patogenesis. Typical laboratory findings are elevated T3, T4 levels and normal or mildly elevated TSH levels. Same mutations may lead to different symptoms in different patients because of different end organ expressions for a mutation.
There are different therapy strategies like thyroid hormone replacement for patients with hypothyroidism or antithyroid treatment for patients with hyperthyroidism. However only propranolol therapy may be sufficient for patients who has only tachycardia compliant. We aimed to report a case with Refetoff Syndrome, tachycardia caused by thyroid hormone resistance ameliorated with propranolol treatment.
Case Report: Nine years old boy admitted to hospital with palpitation. In physical examination, he had tachycardia with 126/minute heart rate. Other physical examination findings were normal. His electrocardiogram revealed sinusal tachycardia. No other rhythm problem was seen in 24 hour electrocardiogram monitorisation. In laboratory examination, free T4 level was 4.27 ng/dl (0.96-1.77) and TSH was 2.87 μIU/ml(0.7-5.97). Thyroid auto antibodies were negative and thyroid ultrasonography was normal. Laboratory findings indicated thyroid hormone resistance. Genetic analysis was performed. c.926 > G mutation which is responsible for Y321C amino acid was detected as heterozygous positive. He was given propranolol therapy: 2 mg/kg/day and after one month, in the second control he didn't have any tachycardia compliant.
Discussion: The sensitivity of peripheral tissues to thyroid hormone is different. While some patients have hyperthyroidism or hypothyroidism signs, some of them are asymptomatic. So that, there is no certain consensus about treatment. Propranolol takes control adrenergic symptoms and decrease T4 to T3 conversion by inhibiting 5-deiyodination path. Propranolol treatment only by itself may be sufficient for the treatment of a this rare disease called Refetoff syndrome.

P1-82

Pulmonary Arterial Hypertension Eisenmenger Syndrome Dıagnosıs Of Patıents Serum Levels Kallistatin

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Introduction: Kallistatin, serin proteinase inhibitor, is first discovered and identified as tissue kallikrein binding protein and a unique serine proteinase inhibitor, and has emerged as a novel inhibitor of angiogenesis and inflammation.
Pulmonary arterial hypertension/Eisenmenger Syndrome, from left to right shunt congenital heart disease is a important ıssue for children. Eisenmenger syndrome is progressive obliterative vasculopathy; the pathogenesis endothelial dysfunction and function of ion channels, calcium homeostasis, changes in platelet and endothelial function, intravascular thrombosis proliferation reactivity increased vascular inflammation and remodeling.
In this study, pulmonary arterial hypertension Eisenmenger syndrome diagnosis of patient's serum levels kallistatin was evaluated; by this way. Pulmonary arterial hypertension is considered a possible relationship between serum levels of kallistatin.
Methods: We enrolled the patients with pulmonary arterial hypertension diagnosis that confirmed by the angiocardiography in our center and their serum kallistatin levels were studied.
Study population:Pulmonary arterial hypertension and Eisenmenger Syndrome caused bycongenital heart diseases having left to right shunt (GroupI), Pulmonary arterial hypertensive patients who didnot develop Eisenmenger Syndrome (GroupII), primary pulmonary hypertension (GroupIII), Congenital heart defects having left to right shunts who did not have pulmonary arterial hypertension (GroupIV), patients with Innocent murmur (GroupV).
Pulmonary arterial hypertension is defined as mean pulmonary arterial pressure greater than 25 mmHg.
Results: The study included total 78 patients(44 girls, 34 boys). 16 with Eisenmenger syndrome, 15 with mild to moderate, 5patients with severe pulmonary arterial hypertension (without EisenmengerSyndrome). Seven patients had primary pulmonary arterial hypertension (without left-to-rightshunt),19patients had left to right shunt without pulmonary hypertension(13ASD,6VSD). Control group consisted 16 patients with innocent murmur.
Serum levels of kallistatin were significantly lower ($p < 0.05$) in patients with Eisenmenger Syndrome. Negative correlation was detected between mean pulmonary arterial pressure and serum kallistatin levels.
Conclusion: Kallistatin as a novel inhibitor of angiogenesis may be an important non invasive marker in the follow-up of pulmonary hypertensive patients.

P1-83

Evaluation of the effect of severe anemia by speckle tracking echocardiography on cardiac function pre and after treatment

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Objectives: Anemia is an independent risk factor for poor outcome in chronic heart failure, but its role in heart failure with normal EF (HFNEF) remains unclear. We aimed to examine the effect of correction of iron and vitamin b-12 deficiency on cardiac mechanics in children with anemia.
Methods: We studied 38 children with severe anemia (hemoglobin levels < % 8 gr/dl) with normal ejection fraction (median age 5.2 years; 6 months - 17 years). The control group comprised age- and sex- matched 30 healthy children (median age 5.7 years; 5 months - 17 years). Children with anemia evaluated using echocardiography basally and after correction of the anemia with oral iron and vitamin b-12 supplements. Cardiac functions were assessed by using conventional echocardiography, tissue Doppler

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imaging (TDI) and two dimentional speckle tracking echocardiography (2DSTE).
Results: The mean hemoglobin concentration of patients was 6.3 ± 1.2 gr/dl. Of 38 children with anemia, 30 patients had iron deficiency anemia, 4 patients had vitamin b-12 deficiency anemia and 4 patients had iron and vitamin b-12 deficiency. After iron and vitamin b-12 supplementation, the mean hemoglobin concentration was 11.8 ± 1.3 gr/dl. Initial the left ventricle global and regional longitudinal strain and strain rate values were significantly lower in patients compared with after treatment of patients and healthy children, while circumferantial and radial strain and strain rate did not differ among groups. Supplementation with iron and vitamin b-12 longitudinal strain and strain rate values improved significantly in patients.
Conclusion: Our results suggest that left ventricle longitudinal myocardial deformation is decreased in children with severe anemia. Supplementation with iron and vitamin b-12 results in a significant increase in longitudinal srain comparable with before treatment in children with anemia.

P1-84
The effect of body mass index and insulin sensitivity on vasovagal syncope in adolescents
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Background: We aimed to determine the effect of body mass index (BMI) and insulin sensitivity on adolescent with vasovagal syncope and to compare positive head-up tilt-table test (HUT+) results with negative (HUT-) results.
Methods: The study included 360 adolescents who were referred for HUT as a part of the diagnostic investigation of syncope. Patients were divided into four groups according to their BMI percentiles: < 5p (underweight), 5- < 85p (normal), 85-95p (overweight), and ≥ 95p (obese). Different indices of insulin sensitivity that are obtained by fasting serum glucose and insulin level were assessed. Fasting glucose and insulin levels (G0, I0), G0/I0 ratio, G0xI0, insulin resistance of homeostatic model assessment (HOMA-IR), quantitative insulin sensitivity check index (QUICKI) were calculated for each patient.
Results: The mean age of the patients was 13.7 years and 62% were female. The ratio of HUT+ was 57.2%. The prevalence of an HUT+ was not statistically different between BMI groups. However, the percent of HUT+ was higher in underweight patients. In contrast to boys, BMI, BMI p, and BMI SDS values were found statistically lower in girls with HUT+ than HUT-. However, none of the insulin sensitivity indices showed significant differences between patients with HUT+ and HUT-. The number of syncope episodes was not different between BMI groups. Moreover, it was not associated with insulin sensitivity indices.
Conclusions: In our sample, BMI are associated with HUT+ especially in young girls. However, insulin sensitivity has no effect on response to HUT. Low BMI could be one of the predisposing factors for vasovagal episodes. Further studies are required to better understand these results.

P1-85
Combined treatment including resection and postoperative systemic chemotherapy for intrapericardial yolk sac tumor presenting with pericardial tamponade in a 15-month-old boy
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Mediastinal germ cell tumors account for only 2-5% of all germinal tumors, but they constitute 50-70% of all extragonadal tumors in which yolk sac tumor (YST) is a malignant variant. Although their peak incidence is in the third decade, several cases have been reported in infantile age group. Herein, we report management of an infant with an unusual location of YST who presented with signs of pericardial tamponade.
Case: A previously healthy, 15-month-old boy was admitted to a state hospital with complaints of malaise 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, he had tachypnea, tachycardia, and poor peripheral perfusion. Immediate echocardiography revealed a massive pericardial effusion with signs of cardiac tamponade and a large intrapericardial mass was noted anterior to the right ventricle (Figure). His serum AFP was increased to 8167 ng/ml (reference range: 8.5 ± 3.5 ng/ml) and serum human chorionic gonadotropin level was normal. To better define the anatomical relationship of the mass, a computerized tomography was performed. A heterogenous 4 × 3.5 × 5 cm intrapericardial tumor inseparable from the right ventricle was detected and there was compression of the superior vena cavae and atelectasis of the left lower lobe of the lung. After pericardiocentesis of 150 cc of hemorrhagic fluid his clinical condition improved. Due to the suitable location of the mass, a needle biopsy under fluoroscopy was also performed and diagnosis of YST was confirmed. The next day, he underwent surgical resection of the tumor via median sternotomy. After an uneventful postoperative period, he was started on BEP (Bleomycin, Etoposide, Cisplatin) regimen and is doing well.
Conclusion: Yolk sac tumor should be added to the differential diagnosis of intrapericardial masses. Early recognition of intrapericardial YST is important because of its rapid course and fatal outcome.

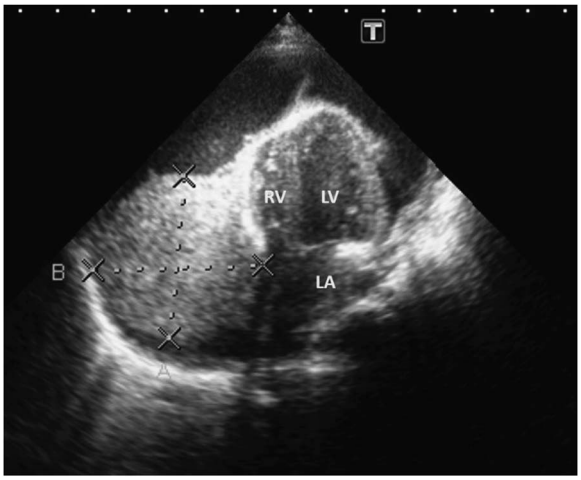


Figure
Echocardiographic view showing a massive pericardial effusion and a large intrapericardial mass.

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P1-86

Idiopathic recurrent pericarditis in an adolescent: management with interleukin-1 receptor antagonist

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Recurrent pericarditis is defined as the recurrence of pericardial symptoms after acute pericarditis and associated with various conditions. However in some cases the cause of recurrences remains unknown and called as Idiopathic Recurrent Pericarditis (IRP). The most commonly accepted theory for pathogenesis is that IRP represents an autoinflammatory phenomenon. Herein we describe an adolescent with IRP who is treated with interleukin-1 receptor antagonist (anakinra) and showed dramatic therapeutic efficacy.
Case: A-17-year old boy was admitted to Pediatric Cardiology Department with complaints of chest pain and dyspnea in August 2015. His medical history was unremarkable except for the last 18 months during which he had 8 similar episodes of chest pain that were responsive to NSAIDs and each one lasting for 3-7 days. He had diagnosis of pericarditis and was given colchicine for the last episode for 3 months. However, after cessation of colchicine, his symptoms recurred and admitted to our hospital. Physical examination showed shortness of breath, an increased heart rate of 118 beats/min with deep heart sounds. Laboratory tests showed an increase in acute-phase reactants. Echocardiography revealed moderate pericardial effusion. Viral serology, C3, C4, ANA studies showed no abnormality. Genetic studies testing for familial Mediterranean fever and tumor necrosis factor receptor-associated periodic syndrome were negative. He responded well to an appropriate dosage of ibuprofen, colchicine and discharged without no pericardial effusion. However, after 6 weeks of wellness, his symptoms recurred while receiving colchicine. He needed pericardiocentesis due to a large amount of effusion and prednisolone was started. In November 2015, two weeks after discontinuation of prednisolone, the patient experienced a new episode of pericarditis, characterized by precordial pain, increased levels of acute-phase reactants and pericardial effusion seen on echocardiography. Administration of anakinra was followed by dramatic clinical response and normalization of the laboratory findings within 24-48 hours.
Conclusion: This case has been assessed worth to be reported as the patient was treated with only anakinra. It is concluded that anakinra is to be considered as an important treatment option of IRP.

P1-87

Long term follow-up of children after treatment of isolated aortic coarctation

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Introduction: The treatment of aortic coarctation (CoAo) depends on many factors including coexisting heart defects, the age and body weight, as well as the experience of cardiosurgery centre. In Cracow during the last 15 years we changed the strategy for treatment of CoAo. We introduced interventional procedures (balloon aortic angioplasty BAA and stent implantation) and the end to end anastomosis became the method of choice in cardiosurgery. The aim of the study is to evaluate the rate of recoarctation and the occurance of systemic hypertension with necessity for interventional procedures or reoperations depending on the age and type of treatment.
The methods: the material consisted of 216 patients (pts) (143 males and 73 females with ratio 2:1) with CoAo treated with interventional procedures or surgery for the last 15 years in Cracow. The patients were divided into 3 groups: Gr I: n = 135 infants, Gr II: n = 52 children operated on at the age >1 year, Gr III: n = 29 pts treated with BAA (n = 7) or stent implantation (n = 22).
Results: The age of the treatment was: x = 2.0 ± 2.5months in GrI, x = 6.7 ± 5.2 years in Gr II, x = 10.1 ± 4.7years in Gr III. Follow-up was x = 7.8 ± 4.3 years in GrI, x = 10.7 ± 4.1years in Gr II and x = 4.7 ± 2.4 years in Gr III. All patients survived. Altogether 46 pts required BAA mostly in Gr I-41pts (30%) vs 2 pts (4%) in GrII and 3pts (9%) in Gr III. Stents were implanted in 3pts (6%) in Gr II and 2pts (7%) in Gr III (Covered-stent Grafts due to aortic aneurysm). Systemic hypertension was found predominantly in Gr III - 13 pts (44.8%) vs 6pts (4.4%) in Gr I and 11pts (21%) in Gr II. Reoperation was necessary in 9 pts due to re-coarctation with hypoplastic aortic arch (6pts-4.4% in GrI and 3pts-6% in GrII).
Conclusions: Late diagnosis and treatment of CoAo is a risk factor for persistent systemic hypertension. The necessity for reinterventions is significantly higher in children operated on in neonatal period.

P1-88

Prenatal diagnosis predicts respiratory failure in patients following absent pulmonary valve repair

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Background: Absent pulmonary valve syndrome (AVPS) is a rare congenital heart disease. Mortality and morbidity after repair is mainly related to acute or chronic respiratory failure due to the dilated pulmonary arteries.
Objective: To describe outcome and risk factors for mortality and respiratory morbidity for AVPS.
Methods: Retrospective analysis of all consecutive patients undergoing surgical repair from 1995-2015 in a single center. Patients' and procedure-related variables were analyzed by means of cox analysis and logistic regression.
Results: 31 patients underwent surgery at a median weight of 4.5 kg (range: 2-63) and a median age of 4.5 months (range: 0.4-184). 58% (n = 18/31) had a prenatal diagnosis of APVS and 26% (n = 8/31) had Di George syndrome. 23% (n = 7/31) had to be operated in the neonatal period. 39% (n = 12/31) required preoperative mechanical ventilation. Mortality before hospital discharge was 13% (n = 4/31). All deaths were due to respiratory failure. Lower weight at surgery, younger age, and larger left pulmonary artery were univariate risk factors for death. Prenatal diagnosis of APVS and type of right ventricular outflow tract (RVOT) repair, e.g. valved conduit, transannular patch or monocusp patch, did not impact mortality. 23% (n = 7/31) needed more than 15 days of mechanical ventilation after surgery. Multivariate risk factors for mechanical ventilation > 15 days were prenatal diagnosis (p = 0.0001), younger age (p = 0.0001) and Di George syndrome (p = 0.021). 1, and 10-year survival rate after repair was 87% (CI95%: ± 12%), and 83% (CI95%:+ /-14%), respectively. 5-year survival without surgical RVOT reintervention was 60% (CI95%: ± 20%). Mean follow-up was 6.3 years (range: 0.3-16.8). At last visit, all survivors were in good clinical condition without need for chronic cardiac medication in 92%. Four had undergone catheter interventions for RVPA stenosis. 19% (n = 5/26) had a recent history of respiratory symptoms or repetitive infections.

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Conclusion: Hospital mortality from respiratory failure is significant. Prenatal diagnosis identifies patients at risk for postoperative respiratory failure. Hospital survivors have a good midterm outcome with absence of symptoms in the vast majority of the patients.

P1-89

Infective endocarditis in children – Retrospective observations between 2000 and 2015

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Background: Infective endocarditis (IE) remains a diagnostic and therapeutic challenge associated with high morbidity and mortality. We aimed to evaluated the profile of microbial organisms and clinical manifestations during the past decade as guidelines shifted.
Methods: We conducted a retrospective study examining pediatric IE cases (< 16y) treated between January 1, 2000 and September 1, 2015. Clinical presentation, treatment, complications and outcome of IE as well as underlying microorganisms and congenital heart defects were reviewed.
Results: A total of 45 patients were diagnosed with 'definite IE' using the Modified Duke Criteria. Prevalence of IE was consistent over time with 24 episodes between 2000 and 2007 and 21 episodes between 2008 and 2015. Overall, 16 patients (36%) required cardiac surgery due to IE. 7 patients (16%) died with 6 children after hospital acquired (HA) IE < 1year of age.
87% patients had an underlying congenital cardiac defect (septal defect in 7, none with ASD or muscular VSD; open ductus arteriosus in 1). Out of 17 included patients operated for TOF or truncus arteriosus, 12 presented with endocarditis of a prosthetic graft (homograft, Melody valve or Venpro graft).
A causative organism was found in 33 cases (73%): viridans Streptococci in 11 (24%), coagulase negative Staphylococci in 9 (20%) and Staphylococcus aureus in 8 (18%).
We found 8 (33%) cases of community acquired (CA) IE between 2000 and 2007 against 16 cases (76%) between 2008 and 2015. With viridans Streptococci being more prevalent in the CA group, we observe a relative increase of IE cases caused by viridans Streptococci between 2008 and 2015 (not statistically significant).
14 patients (31%) presented with HA IE during the first year of life with 79% after heart surgery and a prevalence of coagulase-negative Staphylococci (43%).
Conclusion: Pediatric IE incidence remains similar over the investigated time period with a shift towards CA IE. Streptococci accounted for the majority of cases.
In our population, patients with muscular VSD, ASD en open duct are at very low risk to develop IE. Awareness of IE prevention is crucial for all physicians and patients, especially after implantation of prosthetic grafts.

P1-90

Plastic bronchitis as a late complication in children after Fontan operation

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The etiology of plastic bronchitis (PB) as a late complication after Fontan operation is still unclear.
Patients: In our center out of 280 Fontan patients (median follow-up 4.2 years (2 mo - 14.5 years) 7 children (5 girls, 2 boys) developed PB (2.5%). Six had HLHS and 1 a single ventricle with hypoplastic arch. All underwent 3-stage palliation with Norwood- (Sano: 5, BT-Shunt: 2), Glenn- (median: 3.6 months (2.5-5.3) and extra cardiac Fontan OP (median: 3.3 years (2.3-7.2)), 5 of them fenestrated.
Results: Onset of PB happened at a median age of 7.3 years (3-12.4) or 1.1 years (0.2-10.1) after Fontan OP. Transient post-operative chylothorax was present in 5 patients. Therapy of PB consisted of bronchoscopy (n = 7) ECMO (n = 1) and cardiac interventions to improve hemodynamics (n = 5) as well as inhaled rt-PA, steroids and medication to improve cardiac function, to reduce pulmonary vascular resistance. After a follow-up of median 3.1 years (1.4-8.5) all patients are alive. One child with heart failure and tricuspid regurgitation was transplanted and has no casts 4 months after HTX. 2 patients are cast free without rt-PA inhalations for 1.5 years following successful interventional treatment of hemodynamic problems with increased CVP (APCAs and AV fistula), 1 patient is now cast free for 2.5 years without therapy after re-opening of the fenestration and pacemaker implantation, 3 patients are still producing casts for 3, 3.1 and 10 years respectively. One 15 year old girl without lymphatic anomalies responding to high dose cortisone, one 7 year old girl with an obvious lymphatic fistula from the thoracic duct to the right lung (awaiting selective lymphatic intervention) and one 13 year old girl after 11 interventions including re-fenestration, repeated APCA coiling and isthmus-stenting now awaiting lymphatic imaging.
Conclusion: PB after Fontan remains a diagnostic and therapeutic challenge as some etiologies are still unknown and multifactorial and each patient has specific characteristics needing tailored treatment.

P1-91

Quality of life in children after surgery for congenital heart disease - Univentricular heart and Tetralogy of Fallot

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Introduction: In the last decades there has been an outstanding improvement in survival of children with congenital heart disease, including children with complex forms, such as univentricular heart (UH). Nevertheless, this group of diseases is associated with a significant long-term morbidity. The aim of this work was to evaluate quality of life (QL) of children who underwent surgery for UH and a simpler cyanotic lesion, tetralogy of Fallot (ToF) and compared the two groups with normal values obtained previously in children with the same age.
Methods: We performed a cross sectional, descriptive study. We assessed QL of children with UH and ToF aged 8-12, using the *Peds QL™ 4th version* for parents. This is a QL questionnaire validated for this age group in our population. Additional data such as number of surgeries, cardiac catheterizations and hospital admissions were also assessed.
Results: 37 children with congenital heart disease participated in the study, including 21 with UH and 16 with ToF (60 children were randomly selected to participate but 23 were excluded due to unavailability to participate). Mean number of surgeries

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(ToF: 1.5, UH:2.76), catheterizations (ToF: 1.31, UH:2.45) and total admissions (ToF: 3.25, UH:6.04) in the 2 groups were significantly different ($p < 0.01$). The results of total indices of QL and of subscales are presented in Table 1.

Conclusions: Children with UH had lower QL indices in all categories, however there was no significant difference between this group and the ToF group. Compared to healthy children, ToF group had lower scores in all categories except emotional performance, however the only significantly lower score was school performance. Children with UH had lower scores than healthy children in all categories, particularly in physical, school and total QL scores. Mean number of cardiac surgeries, cardiac catheterizations and total number of hospitalizations were higher in the group of children with UH, contributing to a lower QL score. These results are in accordance with the known higher long term morbidity of UH.

Table 1	ToF (n = 16)	UH (n = 21)	p	ToF (n = 16)	Healthy Children
Physical performance Mean ± SD	71 ± 27.7	68.2 ± 24.9	0.65	71 ± 27.7	83.5 ± 14.7
Emotional performance Mean ± SD	74.6 ± 16.4	71.6 ± 17.1	0.71	74.6 ± 16.4	73.3 ± 16.7
Social performance Mean ± SD	83.8 ± 18	76.4 ± 24.9	0.53	83.8 ± 18	84.6 ± 15.1
School performance Mean ± SD	61.2 ± 25.5	50.9 ± 20	0.22	61.2 ± 25.5	78.2 ± 15.8
Total Mean ± SD	72.8 ± 17	66.9 ± 17.3	0.27	72.8 ± 17	79.8 ± 12
	p	**UH (n = 21)**	**Healthy Children**	**p**	
Physical performance Mean ± SD	0.37	68.2 ± 24.9	83.5 ± 14.7	**0.035**	
Emotional performance Mean ± SD	0.83	71.6 ± 17.1	73.3 ± 16.7	0.794	
Social performance Mean ± SD	0.53	76.4 ± 24.9	84.6 ± 15.1	0.271	
School performance Mean ± SD	**0.02**	50.9 ± 20	78.2 ± 15.8	**0.001**	
Total Mean ± SD	0.31	66.9 ± 17.3	79.8 ± 12	**0.007**	

Significance level was 0.05

P1-92

Exercise-based cardiac rehabilitation programme in children with Congenital Heart Disease increased LV - ejection fraction

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Introduction: Role of pediatric cardiac rehabilitation has been shown to lead to sustained improvements in exercise response and clinical correlation to lesser heart failure manifestations. However, in Europe these programmes are still only beggining to be implemented.

Aim: To demonstrate feasability, safety and to evaluate the cardiovascular effects of a formal exercise programme in a population of patients with Congenital Heart Disease.

Methods: Selected patients had a complete clinical, ECG and echocardiogram, spirometry and CPET with Bruce protocol before and after the programme. Exercise training sessions in hospital, with continuous ECG and saturation telemetry, consisted of respiratory kinesyotherapy and muscular entrainement of inspiratory muscles and diaphragma and aerobic training of 10-15 minutes with intervals, during 3 months, twice/weekly, individual heart rate goal was the heart rate at anaerobic treshold on CPET. Perceived intensity in Borg scale was moderate to rather strong. Results are presented as mean ± SD and pre and post RHB values were compared (2-sided T-test, $p < 0.05$).

Results: Twenty one patients were included in the programme, during the last five years. Mean age was 10.8 years-old (6-22), height 110-166 cm, weight 24-51 Kg. The population was: two heart transplant patiens, two Fontan circulation and the remaining were TOF submitted to complete correction, with clinical heterogenous features: variable degrees of pulmonary valve regurgitation (mild to severe), one patient had a percutaneous pulmonary valve implanted 2 years before and another patient had VEs, couplets, bigeminy and non-sustained TV on 24h-Holter previous to the programme. However, during CPET the VEs disappeared and no arrythmic events during the programme were observed. Of the results obtained we point out that the there was a significative increase of the ejection fraction from 61.3% to 68% ($p = 0.04$). No complications or adverse events were observed during CPETs nor exercise sessions in this population.

Conclusions: In a selected population, this exercise- based cardiac rehabilitation programme led to an increase in LV ejection fraction, that is meaningfull in these patients.

P1-93

Targeted pulmonary vasodilator therapy in children. Experience of a single reference center

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Introduction: The management of pulmonary arterial hypertension (PAH) in children is complex because most evidence-based practices derive from adult PAH studies.

Aims/Methods: We retrospectively analyzed all consecutive patients (pts) followed in a PAH consultation that began targeted therapy between 2003 and 2015.

Results: We analyzed 41 pts, of which 54% were female. PAH was idiopathic on 12.2%, associated with congenital heart diseases on 46% and with other diseases on 4.9%. Separately we formed a group with pts that underwent Glenn/Fontan surgery (36.6%), all with high pulmonary vascular resistance (PVR). All pts began targeted therapy (mean age 6.1 ± 5.8 years): 61% with bosentan (B); 17% with sildenafil (S), 19,5% double therapy with B+ S or a prostacyclin (P) and 2.4% triple therapy with B+ S+ P.

Evaluation before PAH therapy implementation showed: 1) Functional class III or IV on 78%, right heart failure (RHF) on 51.2% and syncope on 8.1%; 2) Mean of 6-minute walk test (6MWT): 408.6 ± 105.2 meters, performed in 22% of pts; 3) Mean BNP: 748.3 ± 1704.5 pg/mL; 4) On echocardiogram: mean PASP: 80 ± 16.4 mmHg; right ventricle (RV) dilatation on 51.2% and pericardial effusion on 2.4%; 4) CATH: mean right atrium pressure (RAP): 7.1 ± 4.6 mmHg, mPAP: 44.6 ± 22.8 mmHg, cardiac index: 3.2 ± 1.5 L/min/m^2, PVR: 16 ± 1.5 U Wood and SvO2: 63.9 ± 10.8%; all pts have negative AVT; 5) 46.7% of pts had high pressure on Glenn/Fontan.

The mean follow-up time after PAH diagnosis was 4.8 ± 5.5 years. Two pts were submitted to percutaneous atrial septostomy and one to an attempting of Potts shunt. After at least 3 months of targeted therapy, we found, although without statistical significance, improvement in most of the pts, on the functional class, 6MWT, mean BNP value, mPAP and on the pressure of Glenn/

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Fontan circuit. Overall the global mortality was 24.4%. At 5 years, the Kaplan-Meier estimates a survival of 65.5%.
Conclusions: Paediatric PAH is a progressive disease whose mortality rate remains high even with early targeted therapy implementation. Pts with Fontan circulation may benefit of early targeted therapy as form of prevention of circuit failure.

P1-94
Experience with total cavopulmonary connection in children

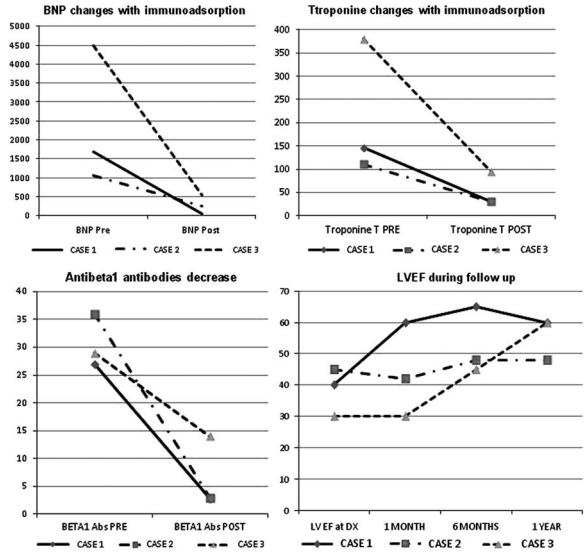
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The objective of this study was to analyze a cohort of children with single ventricle heart after total cavopulmonary connection (TCPC) and assess the factors predictive of early and late outcome.
Methods: Patients who underwent TCPC at age < 18 years from 2000 to 2015 were retrospectively reviewed. Demographic data, cardiac disease, previous medical and surgical history, echocardiographic measurements, all events and outcomes were collected. Predictive factors for survival and events were assessed.
Results: 98 patients were included in the study, Age at TCPC was 7.9 ± 3.9 years (range 20 months to 17 years, media, 7.2). Underlying cardiac disease was: 26 tricuspid atresia, 13 double inlet ventricle, 11 pulmonary atresia, 11 unbalanced atrioventricular defect. 9 single ventricle with right outflow tract obstruction, 13 single ventricle with left outflow tract obstruction, 9 complex CHD, 5 hypoplastic left heart syndrome,and 1 Ebstein. Preoperative ventricular function was normal in 50%, middy impaired in 50%. Age at TCPC was lower in the 2010-2015 period than before (mean 6.6 versus 8.9 years, p = 0.004). Postoperative duration of drainage ranged 8 days to 9 months (median 10 days). Fenestration was performed in 7 cases (7.2%). One or more complications occurred in 27 cases (27.5%): arrhythmia in 6, heart failure in 2, PLE in 7 (7.2%). Two deaths occurred (2.2%) and 4 underwent HTX (4.2%), at a median time of 1.6 and 7 years after TCPC respectively. The incidence of PLE was 10.7% in cases operated before 2010 and 2.4% TCPC performed in 2010-2015 period. All deceased and transplanted cases had underwent TCPC before 2010. Age at TCPC did not differ between failed cases (i.e. death, HTX, PLE) and others. PLE was significantly correlated with death or transplantation (28.5% versus 4.4%, p = 0.01). Overall survival was 97.9% at follow-up pf 14 ± 7.3 years after TCPC (median 13.5 years).
Conclusion: TCPC survival rates in children is favourable and improved since 2010 while age at surgery was decreased. PLE is predictive of death or transplantation.

P1-95
Antibeta1 adrenergic antibodies in acute miocarditis in children: Immunoadsorption treatment

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Introduction: Remarkable advances in understanding the pathophysiology of acute myocarditis have been gained, but no standard immunological treatment has as yet been defined.
Antibeta1-adrenergic antibodies (Antiß1) and their pathogenic effect have been described in DCM. Their elimination by immunoadsorption has been associated with better prognosis.
Our aim was to detect Antiß1 antibodies in children with acute myocarditis and check their hemodynamic evolution after immunoadsorption treatment.
Methods: We reviewed 3 cases of acute myocarditis with positive Antiß1. Antibodies were determined by Celltrend laboratory (positive titers > 15 U/ml). Immunoadsorption was performed daily 4 times with IgG columns (Therasorb®). We analyzed clinical evolution and changes in BNP, Ttroponine, LVEF and Antiß1.
Results: CASE 1: 3 year old boy with Kawasaki disease who developed cardiogenic shock with LV severe dysfunction. Mechanical ventilation and inotropic support was required. He received one cycle of immunoadsortion. After treatment LV function improved and mechanical ventilation and inotropic support were withdrawn. Currently the patient is asymptomatic.
CASE 2: 4 year old girl with 3 episodes of myocarditis treated with milrinone and diuretics. PCR Parvovirus B19 (in blood and endomiocardial biopsy) and Antiß1 were positive so treatment with immunoadsortion and inmunoglobuline perfusion was decided. During the last 4 years she presented several episodes of recurrent myocarditis partially controlled with periodic inmunoglobulin and immunoadsorption (3 cycles). She continues in a good functional class (NYHA II), receives IECAS and beta-bloquers oral treatment and has a moderate LV dilatation with mild disfunction.
CASE 3: Infant(16 months) with cardiogenic shock. Requiring mechanical ventilation and inotropic support. In etiologic study CMV, VHH6 and Parvovirus PCR in blood were positive. Treatment was: ganciclovir, 2 cycles of immunoadsorption and gammaglobuline. The patient improved allowing inotropic and oxygen suppression. Currently, one year after, receiving IECAS treatment and with good functional class.
In our patients immunoadsorption was well tolerated. This technique decreases BNP, Ttroponine, antibodies and improves clinical status. The changes in echo appear later in time.



Conclusions: Antibeta1-adrenergic antibodies could be present in acute myocarditis as a marker of immune response. We believe that immunoadsorption, in these patients is a secure technique and its use is associated with good hemodynamic evolution.

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P1-96
Infective Endocarditis in young population in a 15-year period in a tertiary hospital

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Objectives: To determine the epidemiological, clinical, microbiological characteristics and outcome of infective endocarditis (IE), in patients less than 25 years during a 15-year period in our Spanish tertiary hospital and its relationship to congenital heart disease (CHD).
Methods: Descriptive retrospective study by chart review of patients from 0-25 years who met modified Duke criteria for IE and were admitted to our hospital from January 2000-September 2015. Risk factors (underlying diseases, presence of prosthetic heart materials, central catheters, previous dental procedures, teeth infections, cutaneous wounds, cardiac interventions within 8 weeks), clinical features, diagnostic imaging, microbiological and laboratory findings, management and outcome were reviewed.
Results: 31 patients with a total of 34 episodes of IE were identified (76% males). The mean age was 13.8 years SD 6.7 (range: 0.3-25). 30 patients (88.2%) had underlying heart disease of which 23 (76.6%) had prosthetic heart materials. Only 3 (8.8%) were previously healthy and 1 (3%) was a sever burn. The main presenting symptom was fever with a median duration before admission of 13 days (IQR 4-60). Two presented with septic shock. Viridans streptococci was the most common isolated organism (12), followed by S. aureus (3) accounting both for 43% of total. Three were fungal IE (2 Candida, 1 Aspergillus). In all cases, vegetations were documented via echocardiography (in 23.52% transesophageal echo was required). In half the cases (50%) lesions were localized on prostethic material (70.5% conduits; 17.7% pacemaker leads, 11.8% valves) whereas tricuspid was the most affected valve among native IE (35.3%). The median duration of antibiotherapy was 42 days (IQR 28-126). Acute heart surgical procedures were necessary in 16 cases (47%) all due to hemodynamic impairment secondary to valvular dysfunction or conduit obstruction. In 7 cases, prosthetic valves were placed (including the 3 previously healthy). Systemic complications occurred in 13 patients (septic emboli and shock). Mortality related to endocarditis occurred only in one case.
Conclusions: IE in paediatric population occurs mostly in patients with CHD specially if prosthetic materials present . Mortality seems to have decreased. Multidisciplinary teams are the basis for success in its management.

CONGENITAL HEART DISEASE	Number of episodes	CONGENITAL HEART DISEASE	Number of episodes
Aortic /subaortic disease	2	Mitral valve dysplasia	1
Atrial septal defect	1	Pulmonary atresia with ventricular septal defect	4
Coarctation of Aorta	1	Pulmonary valve stenosis	1
Complete TGA	2	Tetralogy of Fallot	9
Congenitally corrected TGA	1	Ventricular septal defect	4
Double outlet right ventricle	3	Other	1

* 3 patients with IE associated to pacemaker leads (1 Congenital complete heart block 2 Complete heart block secondary to post heart surgery: 1 Fallot, 1 Ventricular septal defect)

P1-97
Left ventricular non-compaction in children

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Introduction: Non-compaction left venticle is a rare, geneticaly determined diseases. It is characterized by trabecular character of the left ventriclular myocardium containing deep recesses, which are perfused form left ventricular cavity. It may occur as an isolated form of NCLV or it may be a part of another congenital heart diseases.
Study goals: Creation of a diagnostic algorythm by using magnetic resonance imaging in children with NCLV. Comparison of MRI findings with echocardiogaphic examination as a reference method.
Patients and methods: The patient group consisted of 16 children (12 boys and 4 girls) with a diagnosis of NCLV. Diagnostic echocardiographic criteria were compared with modified MRI criteria for the diagnostics of NCLV. Echocardiography was used to exclude congenital heart diseases, to confirm increased trabeculization and presence of deep intra-trabecular recesses perfused form left ventricular cavity and to confirm endsystolic non-comacted to compacted myocardium ratio above 1,4. MRI examinaton was done by using Magenton AVANTO SQ 1,5 T with specific cardiologic software and hardware. Non-compacted to compacted myocardium ratio (NC/C) was evaluated in enddiastolic phase, in contrast to echocardiography.
Results: MRI was confirmed to be more accurate in the assessment of non-comacted myocardium layer than echocardiography. Both imaging methods are accurate enough in the measurement of left ventricular size, with minimal diferencies in obtained values. Increased trabeculization has been found mainly on the lateral wall and the apex of left ventricle, sparing the interventricular septum. Post-contrast dye induced myocardial changes of the LV have not been confirmed in children with NCLV.
Conclusions: NCLV is present also in childhood. The gold standard in diagnostics of NCLV remains echocardiography. MRI is appropriate additional method in case of non-conclusive echocardiographic finding.

P1-98
Assessing the miRNA Values of the Children with Rheumatic Carditis: A Possible Biomarker for Future?

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Introduction: MiRNAs are fine regulators of gene expression which participate in the regulation of almost every phase of cell physiology, including the development of immune cells and the

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adjustment of the immune response. In the studies with in-vitro and in-vivo model systems, that specific miRNAs have various roles in cardiovascular development and physiological functions are revealed. Furthermore, some studies have been performed to understand the role of the miRNAs about the myocarditis, heart failure and coronary artery diseases previously. However, their crucial role in the pathogenesis of other rheumatic diseases are investigated, miRNAs have not been studied in children with rheumatic carditis yet. Thus, the purpose of this study is to assess the miRNAs values of the patients with rheumatic carditis.
Methods: Rheumatic carditis patients of 36 children with mean age of 12.1 ± 2.1 and 35 healthy controls were included in this study. Using High Throughput Real-Time PCR device (Fluidigm, Biomark, USA) with 96.96 Dynamic Array IFCs we analyzed the expression of some miRNAs (hsa-miR-16-5p, hsa-miR-221-3p, hsa-miR-223-3p, hsa-miR-10a-5p, hsa-miR-24-3p, hsa-miR--92a-3p, hsa-miR-320a, hsa-miR-21-5p, hsa-miR-155-5p, hsa-miR-132-3p, hsa-miR-146a-5p, hsa-miR-499a-5p, hsa-miR-1, hsa-miR-125, hsa-miR-196a-5p, hsa-miR-130b-3p, hsa-miR-133b, hsa-miR150-5p,hsa-miR-204-5p ve hsa-miR-203a).
Results: There were two-valve involvement in 20 patients (55.6%) and one-valve involvement in 16 patients (44.4%). Hsa-miR-16-5p (-1.46 fold, $p < 0.01$), hsa-miR-223-3p (-1.46 fold, $p < 0.01$), and hsa-miR-92a-3p (-1.27 fold, $p < 0.05$) of the children with rheumatic carditis were lower than those of controls. Whereas, concentrations of other examined miRNAs resulted not differently expressed between the groups.
Conclusion: Our results demonstrated that significant down regulation of hsa-miR-16-5p, hsa-miR-223-3p and hsa-miR-92a-3p in children with rheumatic carditis. Thus, due to this is the first study in this patient group, in order to find out whether these miRNAs might be helpful as biomarkers, further studies are required.

P1-99
Increased Microvolt T-wave Alternans in Children and Adolescents with Pulmonary Hypertension

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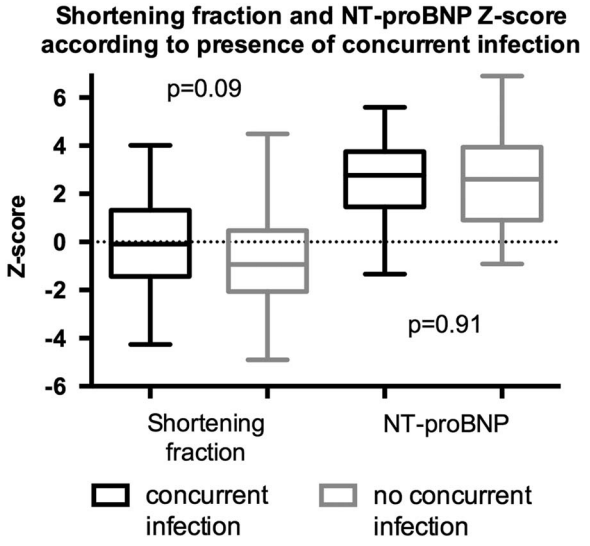
Background: Pulmonary arterial hypertension (PAH) is a devastating disease with poor prognosis of which the major factor is developing of right heart failure. Structural, mechanical and electrical remodeling on the right ventricle led to increase the risk of arrhythmia and even sudden death. There are limited studies about non-invasive tests, such as T-wave alternans (TWA), used to determine the risk of ventricular arrhythmias and sudden cardiac death in adults with PAH. This is the first study which aims to compare the microvolt TWA values in children with PAH and controls.
Methods: Fourteen patients with PAH: 9 with ventricular septal defect (VSD), 4 with atrioventricular septal defect, 1 with primer PAH and 18 healthy controls were included in the study. The TWA values through 24-hour ECG recordings of the patients were evaluated. Analysis of microvolt TWA was considered on the basis of three leads (V5, V1 and aVF). In all patients right heart catheterization, 6-minute walk test (6-MWT), BNP levels, and echocardiographic parameters were obtained.
Results: TWA values; lead V5 was 83.15 ± 10.4 in the patient group (63.5 ± 18 in the control group), lead V1 was 73.6 ± 17 (73 ± 16.8 control group), and lead aVF was 72.1 ± 18.2 (72.6 ± 16.2 in the control group) respectively. When TWA values compared between patients and controls, only lead V5 values statistically increased in PAH group ($p = 0.003$). 6-MWT, pulmonary artery acceleration time and RV free wall thickness significantly correlated lead V5 respectively ($r = -582$, $p = 0.029$, $r = -553$, $p = 0.017$ and $r = 448$, $p = 0.040$). RA volume and inferior vena cava diameter significantly correlated with lead V1 ($r = -589$, $p = 0.01$ and $r = -498$, $p = 0.035$). Tei index ve inferior vena cava diameter significantly correlated with lead aVF ($r = -593$, $p = 0.01$ and $r = -546$, $p = 0.019$).
Conclusion: As a result, microvolt TWA lead V5 values were increased in children with PAH patients and these values were correlated with 6-MWT, pulmonary artery accelaration time and RV free wall thickness. In order to determine the cut-off levels of microvolt TWA as well as their possible predictable values for arrhythmia or cardiovascular mortality in these patients, further studies are required.

P1-100
Coronary outcome in Kawasaki disease is independent from the presence or absence of concurrent infection

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Introduction: Missing the diagnosis of Kawasaki disease (KD) with incomplete criteria is a major problem, which increases the risk of coronary artery (CA) complications. Furthermore, similarities between common pediatric infections and KD may further defer clinicians from diagnosing KD, causing rejection or delay of intravenous immunoglobulins (IVIG) therapy. This study evaluated the prevalence and impact of concurrent infection in KD patients.
Methods: A retrospective analysis of consecutive patients suspected of KD between 2008 and 2014 in a tertiary pediatric hospital.
Results: From 128 patients (3.4 ± 2.7 yo), 42 (33%) had concomitant infection, similar for patients with complete and incomplete KD (27% vs 37%, $p = 0.25$). During clinical course, 69 (54%) patients received antibiotics, with treatment failure in 43 (64%). Patients who received antibiotics, as well as those with infections, were more likely to be resistant to IVIG (38% vs 9%, $p < 0.001$ and 36% vs 18%, $p = 0.03$). Coronary aneurysms and dilatations were more common in IVIG resistant patients (20/29 vs 42/93, $p = 0.03$), suggesting refractory KD as opposed to non-KD infectious disease. Ventricular shortening fraction and NT pro-BNP, a marker of myocardial inflammation in KD, were similar between patients with and without infection (figure). Acute coronary dilatation was present in 11/42 (26%) of patients with infection, and 26/86 (30%) of patients without infection ($p = 0.63$), which persisted in 4/42 (10%) and 9/86 (10%) respectively ($p = 0.87$). Similarly, coronary aneurysms, the ultimate KD complication, occurred in a similar proportion (6/42 (14%) and 12/86 (14%); $p = 0.96$), and were related to delayed diagnosis and treatment ($p = 0.001$).
Conclusions: KD and infection are not mutually exclusive. Recognizing that both can coexist, including incomplete KD, will ensure timely IVIG treatment and appropriate containment of coronary artery complications.

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P1-101

Pulmonary hypertension and cor pulmonale in infants with bronchopulmonary dysplasia: predisposing factors, therapy and immunoprophylaxis of RSV infection

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Objectives: To optimize the tactics of management of children with bronchopulmonary dysplasia (BPD) complicated by pulmonary hypertension (PH), by means of contemporary diagnostics and therapy.
Methods: We have examined 120 children with BPD aged from 1 month to 3 years. Functional class was determined on the basis of age-specific signs according to Panama classification of pulmonary hypertensive vascular disease in children (2011).
Results: Moderate PH (SPAP = 36-50 mm Hg) was diagnosed in 11 children and severe PH (SPAP > 50 mmHg)-in 12 children. Signs of cor pulmonale, considered as cavity dilatation or hypertrophy of the right ventricle free wall on EchoCG, were found in 9 children with severe PH. The development of cor pulmonale in examined children was related to severe PH ($p = 0.00364$) and severe chronic hypoxemia ($p = 0.01986$). In addition to prolonged oxygen therapy in all children with BPD at Sat O2 < 91%, and in the case of pulmonary hypertension with Sat O2 < 94%. 14 children were prescribed Capoten 0.5-1 mg/kg/day, 10 children were further prescribed Sildenafil at a dose of 0.5-2 mg/kg 3-4 times a day. 9 patients (90%) had hemodynamically significant effect in the form of SPAP decrease ($p \leq 0.005$). After the treatment, we observed a significant decrease in the absolute values of SBP (84 ± 36 vs. 36 ± 12 mmHg, $p = 0.005$), in the coefficient of SPAP/SBP by 20% from the baseline ($1 \pm 0{,}4$ vs. 0.94 ± 0.17, $p < 0.005$). To prevent RSV infection during the epidemic season, 76 patients with BPD, including 23 infants with PH, were passively immunized by Sinagis (1 injection per month). During immunization period in this group of children there were no cases of acute respiratory infections caused by RSV and requiring hospitalization.
Conclusions: It is necessary to monitor hypoxemia and SPAP level in patients with severe BPD for timely onset of treatment aimed at prevention of PH occurrence and adverse outcome. Immunoprophylaxis of RSV infection by Sinagis monoclonal antibody agent is effective in the group of the most severely ill preterm infants with PH at the background of BPD.

P1-102

Necrotizing Enterocolitis In Newborns Before And After Critical Congenital Heart Diseases Repair: Our Treatment Approaches and Outcomes

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Critical congenital heart disease (CCHD) course in newborns is commonly exacerbated with necrotising enterocolitis (NEC). NEC generally consequences of the hypoxic ischemic injury combined with alterations of the gastrointestinal microbiome.
The aim of our study was to discuss clinical approach to NEC treatment or prevention in newborns with CCHD.
Materials and methods: 321 newborns ($3.4 \pm 0{,}7$ days, 3.1 ± 0.8 kg) with CCHD underwent surgical repair in Neonates Department between 01.2013 and 06.2015. All patients required emergent surgical intervention due to CCHD incidence. At the hospital stage 23% (n = 73) of infants were diagnosed: 15% (n = 11) on admission were diagnosed with active NEC, 85% (n = 62) showed high probability signs of NEC development.
Results: The CCHD range in newborns with NEC was primarily represented by aortic obstructive lesions (n = 42, 57%), TGA (n = 18, 25%), pulmonary atresia (n = 6, 9%), an extreme form of TOF (n = 4, 5%), HLHS (n = 3, 4%). Of 62 of children with the impendence of NEC development the implementation of preventive treatment was started at the preoperative stage in 63% (n = 39) of cases, and in 37% (n = 23) of cases it was postponed to the postoperative period, due to the need for emergency surgery. Children with the current NEC received corresponding treatment before surgery with the purpose of the relief of this complication: 1) broad-spectrum antibiotic treatment 2) control of the intestinal tract condition, inflammation tests, hemostasis control; hemostatic therapy was applied if necessary; 3) enteral pause, parenteral nutrition with the exception of lipids in the acute phase; 4) the continuation of therapy started before the CCHD repair in the postoperative period — depending on the staging process.
36 patients (49%) were diagnosed with NEC development during the postop: 4 patients had NEC on admission, 9 patients showed high probability signs of NEC development during pre-op and received preventive treatment, 23 patients showed high probability signs of NEC development and did not receive preventive treatment 68 (93%) of patients were cured using medicinal treatment only, and 5 (7%) infants required surgical intervention dui to enterobrosia.
Conclusions: NEC treatment in CCHD patients should start preliminary to the surgical repair, and should be resumed in postoperative period.

P1-103

PDA Device Closure In 'Hypertensive PDA'

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Introduction: In developing countries, PDA still remain a cause of irreversible severe pulmonary hypertension due to late diagnosis as there is poverty, poor education and few cardiac centers to treat for the same. This study will highlight the role of interventional pediatric cardiologist to reduce these cases progressing towards 'irreversible pulmonary hypertension'.

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Objective: To assess the pulmonary artery pressures in patients with large Patent Ductus Arteriosus (PDA) and severe pulmonary hypertension undergoing device closure.
Material and Methods: This descriptive study was carried out at CPE Institute of Cardiology, Multan, Pakistan from 2007 to 2015. Patients who had undergone device closure for hypertensive PDA were included. Patients with mean PA pressures about 66% of systemic are labeled to have hypertensive PDA. Reversible severe pulmonary hypertension decided on clinical ground (normal room air saturation, cardiogegaly on x-ray chest). Patients with weight < 12 kg were excluded. Patient's record was analyzed retrospectively. Echocardiography was performed before and after the procedure. Pulmonary artery pressures were measured in catheterization laboratory before and after the procedure.
Results: A total of 15 patients who had 'hypertensive PDA' underwent PDA device closure. All patients were female. Mean age is 9.8+ 3.1 years. PDA size measured on lateral angiogram is 9.1+ 2.3 mm. Device size used 14+ 2 mm on aortic side. Mean PAP measured on catheterization is 64+ 12.38 mmHg before device closure and 30.8+ 6.34 mmHg after device closure. There is significant fall in mean PAP pressures after device closure in (p-value 0.002).
Conclusion: PDA device closure in hypertensive PDA significant reduced mean pulmonary artery pressures.
Key words: PDA device closure, hypertensive PDA, severe pulmonary hypertension.

P1-104
Spectrum of Cardiac Diseases in Children seen at a Tertiary Hospital in Northwestern Tanzania

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Background: Given an estimated incidence of congenital heart disease (CHD) of 8/1000 live births worldwide, the number of newborns with CHD in Tanzania (46 Mio people, annual birth rate 1.9 Mio) could be as high as 15.200 per year. Therefore, CHD together with acquired heart diseases (AHD) will significantly contribute to morbidity and mortality of Tanzanian children.
Objective: To describe and analyze the spectrum of cardiac diseases as presented to Bugando Medical Centre (BMC), Mwanza.
Methods: Descriptive analysis of patients with cardiac conditions seen at the paediatric department of BMC, one of the four tertiary hospitals in Tanzania mainland (catchment area 13-14 million people) and one out of two hospitals offering regular paediatric echocardiography services.
Results: From 09/2009 to 08/2015 a total of 5249 echocardiography studies for 3188 patients were performed. In 1477 patients (46.3%) congenital and acquired heart diseases were detected.
1106 patients (74.9%) were diagnosed with CHD, the remaining (25.1%) presented with acquired cardiac conditions, mainly rheumatic heart disease (RHD).
VSD was the leading CHD in 302 cases (27.3%), followed by PDA (20.1%) and AVSD (11.4%). Most common cyanotic heart disease was TOF (9.6%). Only 36.1% of patients with CHD were diagnosed in the first 6 months of life.
Indication for surgery or intervention was given in 722 cases (65.3%) of CHD. Unfortunately, nearly 10% of patients presented too late for any surgical or interventional treatment.
209 cases of RHD were diagnosed and indication for surgical treatment was already given in 174 patients (83.2%) at time of diagnosis.
Conclusion: Proportion of pathologic echocardiography studies is high and majority of children diagnosed with cardiac diseases are in need of surgery, which is not yet easily available in Tanzania.

P1-105
Prospective study of children with NSAA (Non Specific Aotroarteritis)

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Introduction: Non specific aortoarteritis (NSAA) or Takayasu's arteritis in children is a rare disease and no prospective study among children is available.
Methods: 28 children with newly diagnosed NSAA were enrolled. Interventions in the form of PTA (percutaneous transluminal aortoplasty) or PTRA (percutaneous transluminal renal angioplasty) were done wherever indicated. Patients were prospectively followed for a mean duration of 13.5 ± 6.9 months.
Results: The mean age at presentation was 10 ± 2.92 years (range 6-15 years) with male to female ratio of 1:1. Presenting complaints were very variable with acute decompensated heart failure (50%) being the most common mode of presentation. Classical findings like asymmetric pulses were rare. Uncommon presentations included B/L cataract, seizures and peripheral embolism. LV dysfunction was seen in 19(67.8%), with severe LV dysfunction in 15 (53.5%). Hypertension was found in 20 (71.4%) patients. Type 2 NSAA was seen in 18(64.28%) while 8(28.5%) had Type 3 disease. Most commonly involved vessel was DTA (descending thoracic aorta), seen in 13(46.4%) followed by the renal arteries in 12 (42.8%). Modified Ishikawa's criteria were met by 21(75%) patients.
Interventions were required in 26(92.8%) and were successful in 22(78.57%) patients. On follow up, the mean NYHA class and the LV EF improved.
Conclusions: NSAA in children is a rare disease and children present with myriad of symptoms and are generally sick at initial presentation. Clinical profile of children with this disease is probably very different from the one seen in adults, and hence poses challenges in recognition and management.

P1-106
Prevalence and Determinants of Hypertension in Apparently Normal School children in India: A four-center study

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Introduction: There is increasing interest in blood pressure (BP) in children as it is likely to affect cardiovascular events in later life. Hypertension (HT) in children is often under recognized in developing countries.
Objectives: To study apparently healthy schoolchildren for BP distribution, prevalence of HT and factors determining these.
Methods: The study was conducted in schools located in Haryana (north), Gujarat (west), Manipur, (northeast) and Goa (southwest) states of India. BP of apparently healthy school children of ages 5 to 15 years were recorded in addition to their age, gender, anthropometric parameters, type of school and season of BP measurement. Type of school (government, government-aided or

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private) was taken as indicator of socio-economic status (SES). Modified recommendations of Fourth report on the diagnosis, evaluation, and treatment of high blood pressure in children and adolescents (Pediatrics 2004) were used for diagnosis of HT.
Results: A total of 14,959 children with mean (±-SD) age of 10.8 (+ /-2.83) years were included. 55.4% were males. Mean (+ /-SD) BP was 108 mmHg (+ /-12.3) systolic, and 68.6 mmHg (+ /- 9.8) diastolic. The overall prevalence of HT (systolic, diastolic or both) was 23%. Systolic HT was present in 13.6%, diastolic HT in 15.3% and both systolic and diastolic HT was present in 5.9% children. Age, gender, weight (but not height), waist circumference, SES and season were predictors of presence of HT on both, univariate and multivariate analysis. HT was more prevalent in younger, female children with higher weight and waist circumference and in winters. HT was less common in children with government-aided (middle SES) schools (prevalence 10%) as compared with government (low SES) (20%) or private (high SES) schools (29%).
Conclusion: There is high prevalence of HT in apparently healthy schoolchildren in India. Regular monitoring of BP and lifestyle modification amongst children should be promoted in order to reduce this prevalence.

P1-107
Congenital aortic stenosis – Treatment outcomes in a nationwide survey

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Objective: To evaluate the treatment outcomes of pediatric valvar aortic stenosis in a nationwide follow-up.
Background: Balloon aortic valvuloplasty (BAV) has been a preferred choice of treatment for congenital aortic stenosis (AS) in Finland since year 2000.
Methods: A retrospective review of all children who underwent treatment for valvar AS in Finland from 2000 to 2014. The clinical records were reviewed for demographic and anatomical characteristics, surgical history, re-interventions and status at the latest follow-up.
Results: Sixty-four pediatric patients underwent either BAV (n = 54) or surgical treatment (n = 10) for valvar AS at a median age of 24 days (range 0.2 days to 16.9 years). Four patients in the surgical group were concurrently treated for aortic coarctation (CoA) and one patient in the BAV group had previously undergone CoA repair. Early mortality (before hospital discharge) was 6.3% and it was associated with critical AS in neonates. There was no late mortality during the follow-up (median 6.9 years, range 0.9-15.1 years). Adequate gradient reduction (residual gradient < 35 mmHg) without significant regurgitation (AR) was achieved in 80% of cases in both treatment groups. Freedom from re-intervention was 72%, 64% and 38% at 1, 5 and 10 years after BAV, respectively. Reason for the first re-intervention was restenosis in 43%, AR in 30% and combined aortic valve disease in 13% of cases. At the latest follow-up, AS mean gradient was 22 ± 12 mmHg and 45% of the patients had moderate or severe AR.
Conclusions: Majority of congenital AS patients require more than one intervention during childhood, both restenosis and AR causing the burden. In our center, BAV has provided good long-term outcome that is equal to our surgical results.

P1-108
A sociodemographic approach to congenital heart defects: epidemiology and mortality in Panama

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Introduction: It is estimated that 26.6% to 48% of children with birth defects die because of a congenital heart defect (CHD). Globally, the prevalence of CHD remains stable around 8 to 9 cases per 1000 live births, but it is know that the burden of CHD falls heavier in countries with higher fertility rates. In our region, large epidemiologic studies are needed to contribute to a wider understanding of CHD. Our main goal is to describe the main sociodemographic and clinical features of patients with CHD in a third world scenario and establish differences in mortality according to these attributes.
Methods: We conducted a retrospective observational study in children with CHD born between 2010 and 2014. A national database was used for data collection deriving from all the institutions with a pediatric cardiologist in Panama. An approximate of 2500 children were born with CHD during this time and our final sample size was 954. Exclusion criteria were preterm patients with a single patent ductus arteriosus, bicuspid aortic valve and Marfan syndrome.
Results: The most common defects found were patents ductus arteriosus in 362 cases (37.9%), followed by ventricular septal defect in 360 patients (37.7%) and atrial septal defect in 184 (19.3%). Total anomalous pulmonary venous drainage was the most common critical CHD, reported in 94 cases (10%). Maternal indigenous ethnicity showed to be a risk factor for this development of this latter pathology (OR = 4; 95% CI 2.64-6.07). A total of 284 (30%) of the patients died during this period. A survival analysis was perfomed between the indigenous and the non indigenous patients, and mortality was significantly higher in the first group (HR = 1.4; 95% CI 1.1-1.8).
Conclusions: Mortality seems to be higher in the more vulnerable groups. Poverty and lack of access to health service might play a critical role in the course of the disease in children from ethnic minorities. Further studies are needed for a more complex understanding of sociodemographic aspects of CHD and how they impact health outcomes and mortality.

P1-109
A case of pulmonary arteriovenous malformation as first sign of hereditary haemorragic teleangectasia

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We report the case of an 8-year-old girl who came to our Pediatric Department for urticaria and dermatitis. Blood exams indicated

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increase of haemoglobin and haematocrit (Hb 17.2 g/dl, Hct 53.6%); physical examination showed perioral cyanosis, digital clubbing and reduced oxygen saturation (86-90%). Electrocardiogram and conventional echocardiogram were normal. Chest X-ray showed a parenchymal consolidation with vascular ectasias at right lung, evocative of pulmonary arteriovenous malformations (PAVM). Transthoracic contrast echocardiography showed an extensive opacification of the left ventricle without outlining the endocardium (grade 3). Chest CT demonstrated a large high-flow PAVM (3.5 × 3.7 × 5 cm) at the right lower lobe and three small PAVM at left lower lobe. Therefore the patient was submitted to embolization of the large PAVM with an improvement of saturation until 100%. An accurate physical examination showed mucocutaneous telangectasias. Because of the presence of 2/4 Curacao criteria, we suspected hereditary haemorragic teleangectasia (HHT), confirmed by the presence of ENG gene mutation. Two years after the embolization, the reopening of the large PAVM required a lobectomy of the right lower lobe. Actually she is in a state of general health with a good oxygen saturation.

HHT is a rare systemic fibrovascular autosomal dominant dysplasia. Its prevalence is currently estimated at one in 5000 to 8000. Mutations in ENG and ALK1 genes have been reported to cause up to 85% of HHT. The signs and symptoms of HHT are nonspecific and are extremely variable within families. HHT is often difficult to diagnose on the basis of history and physical examination alone, especially in children. In our case, the presence of PAVM led us to think about HTT, even without nosebleeds and family history that are usually the most common criteria. Mutations in ENG gene are more frequently associated with PAVM, so these patients need of a careful follow up for the risk of progression of vascular disease.

P1-110
High Sensitivity Troponin T Levels in Healthy Newborns

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Introduction: Research on cardiac biomarkers and their clinical implications has intensified significantly in the past few years. With advances in technology, a new era in troponin assays has approached. High-sensitivity troponin T (TThs) assays represent an important advance with added sensitivity for cardiac myocyte necrosis. This method detects concentrations of the same proteins that conventional assays, just in much lower concentrations. To date, there is an insufficient data regarding TThs levels in neonates. The aim of this study is to assess TThs levels in healthy newborns.

Materials and methods: Following approval by the institutional review board, consecutively 454 healthy full termed newborns were enrolled in the study. Samples of cord blood were drawn and tested for TThs concentrations with high-sensitive assay. Two hundred and thirteen samples (47%) were excluded due to blood hemolysis of various degrees. Further, the group of 241 healthy newborns (birth weight 3423 g ± 429 g) was statistically analyzed. The 97.5 percentile of TThs concentration was assessed and correlation analysis was performed.

Results: The mean concentration of TThs in healthy newborns was 42.1 ± 16.4ng/ml, 97.5 percentile was 81.2 ng/ml (confidential interval 74.2-104.2 ng/ml). We found statistically higher TThs concentrations in boys when compared to girls (mean 44.6 ± 18.2ng/ml; 39.4 ± 13.6ng/ml, $p < 0.05$ respectively). TThs levels in newborns after cesarean section were lower than in children delivered vaginally (mean 38.9 ± 15.1ng/ml; 43.5 ± 16.8ng/ml, $p < 0.01$ respectively). Importantly, TThs concentrations were statistically decreased in hemolytic blood samples when compared to non-hemolytic samples (39.7 ± 18.9 ng/ml; 42.1 ± 16.4ng/ml, respectively $p < 0.01$).

Conclusion: This study confirmed the higher TThs concentrations in healthy newborns compared to adult population. The reason for this elevation is unclear. The rise may be related to the apoptosis of myocardial cells and troponin T isoforms transformation within fetal and newborn period. TThs concentrations may be decreased in hemolytic blood samples. One should take all these facts into consideration in clinical assessment of myocardial disease in newborns.

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P1-111
Prognostic value of cardiac biomarkers in congenital heart defects is time-dependent: umbilical cord versus newborn blood

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Title: Prognostic value of cardiac biomarkers in congenital heart defects is time-dependent: umbilical cord versus newborn blood

Aim: To compare whether there is a difference in cardiac biomarkers according to umbilical cord and newborn peripheral venous blood source.

Methods: Newborns with prenatal diagnosis of CHD were admitted consecutively at a Neonatal Intensive Care Unit, between August 2012 and March 2015. Plasma levels of BNP, troponin I, myoglobin and CK-MB were measured in the cord blood and in the peripheral venous blood of the newborn and compared.

Results: The cardiac biomarkers were significantly higher in the newborn than in the cord blood sample in newborns with congenital heart defects. Median (P25-P75) plasma levels in the newborn peripheral venous blood vs. cord blood of BNP were 86.2 (59.9-960.8) vs. 29.6 pg/mL (14.9-60.2), $p = 0.001$; CK-MB 8.2 (4.7-14.7) vs. 3.6 (2.9-4.7), $p = 0.002$; troponin I 0.04 (0.01-0.06) vs. 0.01 (0.01-0.01), $p = 0.001$ and myoglobin 104.7 (61.7-154.5) vs. 43.8 (32.9-55.6), $p < 0.001$, respectively.

Conclusions: The cord blood has the advantage of being noninvasive when compared with the newborn blood. However, the cardiac biomarkers levels are time-dependent in CHD, and the higher values in the newborn blood may reflect the impact of the hemodynamic changes that occur in the transition from fetal to post-natal circulation.

P1-112
Paediatric Infectious Endocarditis in University Hospital during Years 2010-2014

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The guidelines for infectious endocarditis (IE) prophylaxis have been modified during last years. Specialized pediatric cardiac care

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in Latvia as well as pediatric autopsies is carried out only in University Hospital for Children in Riga.
The objectives of the study were to analyse all cases of IE treated in our hospital during years 2010-2014 or discovered during autopsy in this period of time.
Methods: All the cases with IE were selected from hospitals database and/or autopsy register. We analyzed case histories to explore the course of disease, causative agents, echocardiographic findings, underlying conditions, previous hospitalizations and the outcome.
Results: There were 10 cases of IE: 5 girls and 5 boys, age 5.2 ± 4 years. 5 cases aortic valve endocarditis (50%), 2-mitral valve endocarditis (20%), 2 cases vegetations in contegra conduit (20%), 1-vegetation in right atrium, 1-vegetations on intracardiac pacemaker leads.In 50% of cases the causative agent was no found. 2 cases Staphylococcus aureus, 2 cases Haemophilus species, 1 case Streptococcus pyogenes and coagulase-negative Staphylococcus and Pseudomona. 50% pacients had previous recent hospitalization due to acute illness (2 cases pneumonia, 2 cases bacterial infection of unknown origine, 1 case arthritis). 60% of patients had underlying congenital heart disease, 40% had previously undergone cardiac surgery. 2 patients had undergone other operations (1 gangrenous appendectomy in patient without heart disease, 1 cleft palate plastics in patient with patent ductus). 30% of patients were in need of inotropic support, 2-ventilation. 1 patients was in need of transvasal pacemaker lead change and 1 patient underwent aortic valve repair 3 years later. In 90% (9) of cases diagnosis was suspected during hospitalization. There were 4 cases of death (40%):1 case of post-mortem diagnosis of IE in neonate with very late diagnosis of transposition of the great vessels that died of hypoxic multiorgane failure, 1-severe myocarditis with heart rhythm disorders and IE, 2-IE with septic shock.
Conclusions: IE remains serious condition with high mortality rates and the diagnosis is sometimes challenging. Every patient suspicious for IE should have several blood cultures prior to antibacterial therapy to find the most effective antimicrobial treatment as early as possible.

P1-113
Longitudinal myocardial deformation in children after the Fontan operation: feasibility and impact of ventricular morphology in a multicenter setting

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Introduction: Assessment of single ventricle (SV) function in patients after the Fontan operation is clinically important. Controversy exists whether SV anatomy (right ventricular [RV] versus left ventricular [LV] morphology) influences SV function. Two-dimensional echocardiography is commonly used to assess SV function, but quantification is difficult due to differences in SV geometry compared to normal hearts. Myocardial deformation by speckle tracking echocardiography (STE) is less geometry dependent.
Study aims: To assess feasibility of STE in patients after the Fontan operation and to investigate whether STE parameters differ between children with dominant RV and LV morphology.
Methods: Cross-sectional, prospective, multicenter study (n = 109 children, 66 boys). Apical SV echocardiographic views were acquired using General Electric (GE, Vivid 7) or Philips (iE33) platforms. STE peak longitudinal strain (ε) and systolic strain rate (SR) of the lateral wall of the dominant ventricle were measured using vendor dependent software (EchoPac and QLAB). Independent T test was used to compare groups. Intra- and inter-observer variability was assessed by coefficient of variation (COV) of STE parameters in twenty patients (10 GE/10 Philips).
Results: Mean age at study was 12,0 (range 9,7-14,6) years. Dominant morphology of the SV was LV in 70 (64%) and RV in 39 (36%) children. Feasibility for STE measurements was 83% for GE and 78% for Philips ($p > 0.05$). Intra- and inter observer COV ranged between 6 and 14% for both platforms. Peak global longitudinal ε was -19,1 ± 3,7% for GE and -18,2 ± 4,4% for Philips ($p = 0.33$). Global longitudinal systolic SR was -1,13 ± 0,34 1/s. for GE and -1,15 ± 0,38 1/s. for Philips ($p = 0.79$). Results for STE parameters in LV versus RV morphology are shown in the table.
Conclusions: Feasibility and reproducibility of STE in children with SV morphology after Fontan palliation is acceptable, results for longitudinal deformation are comparable between 2 vendors. Longitudinal deformation is lower in children with dominant RV's compared to dominant LV's.

	Longitudinal peak strain (%)			Longitudinal systolic SR (1/s.)		
	LV	RV	p-value	LV	RV	p-value
Apical-lateral	-19.3 ± 5.8	-17.2 ± 6.1	0.13	-1.15 ± 0.34	-1.00 ± 0.44	0.10
Mid-lateral	-20.9 ± 5.8	-17.6 ± 6.4	0.02	-1.18 ± 0.46	-1.07 ± 0.45	0.28
Basal-lateral	-19.6 ± 6.0	-18.6 ± 5.0	0.44	-1.26 ± 0.51	-1.10 ± 0.45	0.16
Global-lateral	-19.5 ± 3.5	-17.7 ± 4.2	0.04	-1.20 ± 0.35	-1.04 ± 0.33	0.04

P1-114
Improvement of ergometric outcomes after a personalized exercise and cardiac rehabilitation programme in postoperated Tetralogy of Fallot children

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Introduction: postoperated Tetralogy of Fallot patients (PTF) frequently associate disorders that impair exercise capacity (ExC). We developed a personalized cardiac rehabilitation program (CRP) conceived to counteract the deconditioning in PTF. Our aim: to characterize the effect of CRP through conventional exercise test (ExT).
Methods: 18 PTF patients were recruited. CRP started with clinical review, baseline echocardiography, submaximal ExT and post-exercise echocardiography. Documentation of nutrition, life-style and cardiac risk factors was obtained. CPR consisted in 1hr supervised exercise 2-3 times/week, 3 months, including respiratory training, column table, and aerobic exercise (at 75-85% of the maximal HR reached). After CRP, tests were repeated. Relevant cardiovascular outcomes pre/post CRP were:

- ergometric parameters: baseline and maximal HR and BP, HR reserve (HRR), endurance time (ET) and double product (DP).

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- echocardiographic systo/diastolic functional parameters.
- quality of life outcomes (questionnaires).

Results: Submaximal ExTs (limited by symptoms, including fatige) in treadmill were performed following Bruce protocol. The reason for termination was fatige.
13/18 increased their ET with a mean of +1.56 min, showing an improvement in ExC.
Prerehabilitation mean peak HR was 166bpm (minimal 124bpm, maximal 190bpm). Postrehabilitation mean peak HR was 171bpm (min 134, max 200). 10/18 improved their peak HR, with a mean of +16bpm (min 3, max 31) and increased +9.2% over their peak HR (min 1.6%, max 17.9%). This findings indicated an objective improvement in ExC, cardiac output (CO) and chronotropic response.
Baseline mean HRR was 72bpm (min 36, max 109), increasing after CRP to a mean of 79.4bpm (min 46, max 108). 13/18 increased HRR, with a mean of +10.5bpm (min 1, max 25).
Prerehabilitation mean DP was 197,88.4 (min 9900, max 30,400), increasing postrehabilitation to a mean of 216,92.4 (min 15,410, max 28,500). This findings suggested a rise in CO and a better myocardial oxigen consumption.
No subject had abnormal response to exercise (ischemia, arrhythmias, or excessive hypo/hypertensive response).
Conclusions: It is demonstrated that a 3-month CRP can significantly improve ExC of PTF. ExT provide objective quantitative information of patient's ExC. Routine use of formal CRPs may reduce morbidity in CHD.

P1-115
Paediatric acute myocarditis in a spanish reference center

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Introduction: Acute myocarditis is a rare disease, but may have adverse outcomes in paediatric population. The gold standard diagnosis is still histological but cardiovascular magnetic resonance (CMR) has an important role as a non-invasive diagnostic tool. The aim of our study is the description of a paediatric patient's serie with acute myocarditis and the evaluation of the diagnostic and prognostic utility of non-invasive tools in this population.
Methods: This retrospective study included all pediatric patients (0-16 years) with acute myocarditis admitted to our hospital from July 2007 to July 2015. All patients underwent an echocardiography at admittance. CMR was performed in hemodynamic stable subjects. Endomyocardial biopsy (EMB) was performed in selected cases. A clinical follow-up was performed in outpatient clinics. Adverse outcomes were defined as the composite of cardiovascular death, heart failure, or cardiac transplantation.
Results: 25 patients (11 females and 14 males) were included. The median age of the population was 19 months (2.5-99.5), presenting clinically as: heart failure (44%), cardiogenic shock (40%), chest pain (12%) and arrhythmias (4%). The most common pathogen was parvovirus B19 (31.2%). 15 patients (60%) showed an impaired left ventricular dysfunction (left ventricular function $< 35\%$) by echocardiogram and 10 of them (66.7%) had adverse outcomes, however only 3 patients (30%) with left ventricular function $\geq 35\%$, had adverse outcomes (p 0.022). CMR was performed in 17 patients, with a high correlation in terms of left ventricular function (r:0.92; $p < 0.001$). EMB was performed in 8 patients (32%). With a median follow-up of 22 months, 52% had full recovery, 20% progressed to dilated cardiomyopathy, 16% needed transplant and 12% died.
Conclusions: Acute myocarditis is a less frequent but serious illness in paediatric age. CMR is a noninvasive test that allows us to help the diagnosis. Left ventricular dysfunction was associated with poor outcomes, however no information from CMR was significally associated with outcomes.The use of endomyocardial biopsy is still recommended for selected cases.

P1-116
Spontaneous closure of isolated secundum atrial septal defect in children in Kurdistan

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Abstract: Atrial septal defect (ASD) is the second most common congenital heart defect in children in which ostium secundum ASD representing the majority of cases. Atrial septal defect. Isolated secundum ASD accounts for ≈7% of congenital heart defects.
Aim of the Study: To identify the frequency of spontaneous closure of isolated secundum atrial septal defect in children and to assess the factors that affect spontaneous closure.
Patients and Methods: This study is a retrospective revision of patient's record with ASD secundum diagnosed at echocardiographic unit of Sulaimanya Pediatric Teaching Hospital. They were examined during period of 3 years. They were 100 monitored for >3 months with serial echocardiography. Patients were divided into 4 groups based on ASD diameters (4–5 mm, 6–7 mm, 8–10 mm, or >10 mm).
Results: There was a strong association between ASD diameter at diagnosis and the frequency of spontaneous ASD closure highest frequency of spontaneous closure was among those within 4-5mm diameter at diagnosis. Age at diagnosis was associated with the spontaneous ASD closure.
Conclusions: In our study, 22% of ASD closed spontaneously while 13% regressed to ≤3mm. There is high frequency to close spontaneously among those with initial diameter of 4-5mm of which was 65%. Children ≤1 year of age at ASD diagnosis experienced spontaneous ASD closure in 38.5% of cases, whereas children >1 year of age experienced spontaneous ASD closure in 4.2% of cases.
Gender did not affect spontaneous closure of ASD secundum in our study.

P1-117
Relation between elevation of brain natriuretic peptide and right ventricular dysfunction can be estimated simply by routine echocardiography in patients with repaired tetralogy of Fallot

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Introduction: Right ventricular (RV) dysfunction resulting from pulmonary valve (PAV) regurgitation is serious problem in patients with reapared tetralogy of Fallot (TOF). Replacement of PAV is triggered by severe RV dysfunction, which has been estimated by magnetic resonance imaging. Brain natriuretic peptide (BNP) is reported to increase by RV dysfunction. Furthermore,

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RV dysfunction is reported to cause left ventricular (LV) dysfunction by interaction. We investigated whether relation between BNP-elevation and RV dysfunction could be evaluated simply by routine echocardiography in repaired-TOF patients.
Methods: Echocardiography and blood sampling were reviewed in 88 ambulatory patients with repaired-TOF. Levels of BNP in the top quartile of 88 patients were defined as BNP-elevation (≥42.6 pg/ml). Normal end-diastolic dimension of LV (N-LVDd) was calculated with use of body surface: N-LVDd = 4.1*(BSA)0.5*10.1mm. We defined %RVDd, %LVDd and %LVSd (end-systolic dimension of LV) as the ratio to N-LVDd. We used increase of %LVSd as index for reduced LV contraction.
Results: Patients ranged in age from 2 to 33 years. After multiple logistic regression analysis BNP-elevation was independently associated with odds ratio of 31.7 ($p = 0.029$) for moderate-to-severe PAV regurgitation (≥II/III), 6.5 ($p = 0.013$) for enlarged %LVSd (≥70%), 4.7 ($p = 0.044$) for non-stenosis of pulmonary artery (≤2.5 m/s), and 3.7 ($p = 0.04$) for enlarged %RVDd (≥62%). In monovariate analysis BNP-elevation was additionally related to enlarged %LVDd (≥ 113%) and reduced ejection fraction of LV (≤ 60%). There were no significant associations with following factors: degree of tricuspid valve regurgitation; repair strategy for outflow tract of RV. Of repaired-TOF patients with BNP-elevation, 95% patients had risk factors on RV side; 63% had factors on LV side. Each BNP-elevation in this study could be explained without exception by one or more factors described above.
Conclusions: Our study revealed that BNP-elevation in repaired-TOF patients was associated with RV related factors, and LV dysfunction which would be interacted with RV dysfunction. These dysfunctions colud be simply estimated by routine echocardiogram. All BNP-elevation could be accounted for by any one of these risk factors in this study. Based on this simple investigation we should evaluate RV dysfunction in young repaired-TOF patients by cardiac magnetic response.

P1-118
We should use NT-proBNP with care to decide on candidate for re-surgery in patients with tetralogy of Fallot after definitive repair

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Background: Patients with tetralogy of Fallot (TOF) after repair were sometimes redone right ventricle outflow reconstruction (re-RVOTR). We decide a candidate for re-surgery on the basis of RV overload measured by catheterization or magnetic response imaging, both of which cannot be performed frequently. Levels of NT-proBNP were got routinely on an outpatient basis. We investigated whether RV overload which met a requirement for re-RVOTR was predicted by levels of NT-proBNP.
Methods: The medical records of 101 repaired-TOF patients aged from 1 to 53 years were reviewed. Cardiac catheterization was performed to grasp hemodynamic status between 2010 and 2015. We stipulated that end-diastolic volume of RV was 150 ml/m2 or over (hRVEDVI) as indication for re-RVOTR. First, we sought cut-off value to approve them by square test. Second, we calculated each index for predicting indication of re-RVOTR: positive predictive value (PPV), negative predictive value (NPV), sensitivity (SEN), and specificity (SPE).
Results: Values of NT-proBNP were significantly elevated in hRVEDVI (967 vs. 340 pg/ml). We decided 945 pg/ml as cut-off value to distribute patients with hRVEDVI to one side most statistically (60% vs. 12%, $p = 0.00067$). Index values were follows: PPV 60%; NPV 88%; SEN 35%; SPE 95%. However, the area under ROC curve for predicting hRVEDVI was 0.55 (95% confidence interval: 0.37-0.73). Moreover, the ratios of patients with degeneration for RV, such as low ejection fraction or increased end-systolic volume, were significantly high in patients with NTpro-BNP over cut-off value. Similarly, the ratios of patients with degeneration for LV, such as increased volume on end-diastole or end-systole, or low ejection fraction, were significantly high in over-cut-off group.
Discussion: Our study showed significant cut-off value of NT-proBNP subsisted for predicting re-RVOTR candidate in repaired TOF patients. We could gain high SPE but low SEN with this cut-off value, which indicated that this cut-off value was not suitable for screening. Moreover, we could not distinguish candidate for re-RVOTR by ROC analysis, which pointed out low NT-proBNP values had constant distribution in hRVEDVI. Not only elevated RVEDVI but also other ventricular degeneration rising up values of NT-proBNP would throw out predicting re-RVOTR by NT-proBNP.

P1-119
Cardio-pulmonary factors in patients with 22q11.2del are equal to those in patients with normal chromosome after repair of tetralogy of Fallot

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Background: The ratio of 22q11.2del syndrome is 15% in patients with tetralogy of Fallot (TOF). Patients with 22q11.2del often have abnormality of pulmonary arteries (PA), such as pulmonary atresia, major aorto-pulmonary collateral artery (MAPCA), and high pulmonary vascular resistance. Cardiac and pulmonary-artery complications arise in repaired TOF patients, particularly on remote period. However, there were no reports about cardio-pulmonary functions in repaired TOF patients with 22q11.2del. We investigate cardio-pilmonary matters in repaired TOF patients with 22q11.2del.
Methods: The medical records of 121 repaired-TOF patients were reviewed aged from 1 to 53 years. We performed cardiac catheterization to grasp hemodynamic status between 2010 and 2015. Cardio-pulmonary performances were compared between patients with 22q11.2del and patients without chromosomal aberrancy.
Results: We excluded 10 patients with abnormal chromosome other than 22q11.2del, the rest of whom we divided into two groups: 22q11.2del group ($n = 11$) and normal chromosome group ($n = 100$). As type of PA in 111 patients pulmonary stenosis was 76, pulmonary atresia without MAPCA 24, and pulmonary atresia with MAPCA 11. The ratio of pulmonary atresia with or without MAPCA was different between 22q11.2del and normal chromosome (54% vs. 29%, $p = 0.01$); that of Rastelli type surgery was also different between two groups (72% vs. 37%, $p = 0.049$). Between 22q11.2del and normal chromosome, however, there were no significant differences in ratio of patients with severe PA valve regurgitation; reduced ejection fraction of RV; expanded RV volume; increased RV pressure. Similarly, estimations of pulmonary vascular bed were almost equal between two groups: mean pressure of PA, pulmonary vascular resistance, and pressure of pulmonary capillary wedge. As left ventricular factors, there were no significant differences between two groups. Clinically, the ratio of patients having symptoms was not different between two groups.
Conclusion: Contrary to our exception, TOF patients with 22q11.2del possessed pulmonary vascularization equivalent to those with normal chromosome, if they had completed definitive repair.

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Similarly, they owned both ventricular functions which were not inferior to those in patients with normal chromosome. After radical surgery of TOF we might observe cardiao-pulmonary problems of 22q11.2del in a similar way with normal chromosome.

P1-120
Increased pressure in a pulmonary artery: is it a contraindication to bidirectional cavopulmonary connection?

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Introduction: Bidirectional cavopulmonary connection (BCPC) is optimal intermediate stage for hemodynamic correction of congenital heart diseases (CHD) with single ventricle (SV). However, there is no identical opinion upon optimal criteria for BCPC performance.
Methods: Over the 2012 to 2014 period 89 patients with SV at the age from 2 months to 13 years old (Me = 0.64; IQR:0.5-1.18) were involved into the study. They were performed catheterization of heart cavities with pulmonary resistance calculation at different stages of hemodynamic correction. The patients were divided into two groups: the first group (n = 64) – children with initial pulmonary artery (PA) pressure under 15 mm Hg, the second group (n = 25) – patients with PA pressure above 15 mm Hg.
Results: The average PA pressure was 11 mm Hg (IQR:10.00–13.75), pulmonary blood flow volume indexed to lung volume (iQp)-3.57 (mL/Min)/m^2 (IQR;2.69-4.87), pulmonary resistance indexed to body surface area (iRp)-1.41 WU/m^2 (IQR;0.90-1.98) in the first group. In the second group the average PA pressure was 20 mm Hg (IQR:17.00-26.00), iQp 4.56 (mL/Min)/m^2 (IQR:3.17-6.04), iRp 2.36 WU/m^2 (IQR:1.85-3.10). Taking into consideration increased PA pressure, oxygen test was performed. A slight decrease of average PA pressure up to 19 mm Hg (IQR:17.00-30.00), p = 0.02, iQp increase up to 11.2 (mL/Min)/m^2 (IQR:5.85-15.6), p = 0.003) and iRp decrease up to 1.37 WU/m2 (IQR:0.72-1.95), p = 0 002 were marked after the test. All the patients were performed BCPC. Before the total cavopulmonary connection the average PA pressure was 8 mm Hg (IQR:7.00–9.13), iQp 2.69 (mL/Min)/m^2 (IQR;2.14-3.15), iRp was 1,04 WU/m^2 (IQR; 0.81-1.57), p = 0,1 in the first group. In the second group it was 8 mm Hg (IQR:6.38–11.25), iQp 2.46 (mL/Min)/m^2 (IQR;2.22-3.30), iRp was 1.23 WU/m^2 (IQR:0.86-2.3). The comparative analysis between two groups of patients after DCPC showed no significant difference of cardio-pulmonary hemodynamics, p > 0.05.
Conclusion: Thus, the pressure in a pulmonary artery higher than 15 mm Hg is not a contraindication to BCPC. An operability criterion is the index of pulmonary resistance that after 100% inspired oxygen should be lower than 2 WU/m^2.

P1-121
Distribution of allelic variants of hemostatic genes in patients with single ventricle

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Aim: To study the frequency of allelic variants of hemostatic genes associated with thrombophilia in patients with single ventricle.
Methods and materials: Molecular genetic testing with diagnostics of allelic variants of hemostatic genes was performed to 102 patients with single ventricle. Study material was whole blood. DNA samples were tested for single nucleotide polymorphism in hemostatic genes: F2:20 210 G > A (Factor II), F5:1691 G > A (Factor V, Leiden Mutation), FGB:-455 G > A (Factor I), ITGA2:807 C > T (platelets collagen receptor GP Ia-IIa), ITGB3:1565 T > C (platelets fibrinogenic receptor GP IIb-IIIa), PAI-1:-675 5 G > 4 G (plasminogen activator inhibitor I). Genotype was detected by polymerase chain reaction method using market reagent kit (DNA-Technology, Russia).
Results: 97.1%) of examined children had wild type (GG) of factor II gene, 2.9%-heterozygous (GA), 0% – homozygous (AA). Carriers of wild type factor gene (GG) were 94.1% of children, heterozygous (GA) – 5.9%, homozygous for a mutant allele (AA) – 0%. Polymorphism of factor FGB gene was the following in children with single ventricle: 52.0% – wild type (GG), 39.2% – heterozygous (GA), 8.8% – homozygous genotype (AA). Study of allelic variants of PAI-1 gene disclosed wild type (5 G5G) in 5 (14.7%) children with CHD, heterozygous (5G4G) – in 50 (49.0%), homozygous (4 G4G) genotype – in 37 (36.3%). Study of thrombocytic glycoproteins GP Ia-IIa gene discovered wild type (CC) in 45 (44.1%) children with single ventricle, heterozygous (CT) – in 41 (40.2%), homozygous (TT) variants – in 16 (15.7%). Polymorphism of GP IIa-IIIb was the following in children with single ventricle: wild type (TT) – in 77 (75.5%) pts, heterozygous (TC) – in 22 (21.6%) pts, homozygous (CC) genotype – in 3 (2.9%) pts.
Conclusion: The results of molecular genetic testing of allelic variants of hemostatic genes together with current diagnostic procedures can be used for risk prediction and early therapy management of thrombophilic state in patients with single ventricle.

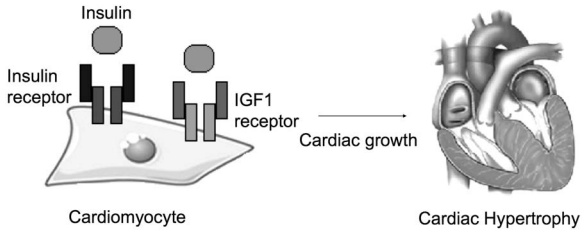
P1-122
The Etiology of Cardiac Hypertrophy in Infants

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Introduction: Cardiac hypertrophy in young childhood is a rare and heterogeneous disease. Unfortunately, often the etiology remains idiopathic. The pathophysiology and distribution of etiologies in infants are supposedly different from cardiac hypertrophy developing later in childhood.
Objectives: To investigate the distribution of etiologies, association with hyperinsulinism, distinctive (echocardiographic) variables and the prognosis of cardiac hypertrophy in infants.
Methods: This single center retrospective study included all patients born between 2005-2014 with cardiac hypertrophy measured by echocardiography (diastolic interventricular septum (IVSd) or left ventricular posterior wall (LVPWd) thickness with Z-score ≥ 2.0) below the age of 1 year. Children with cardiac hypertrophy due to congenital heart disease (CHD) or hypertension were excluded. Underlying diagnosis, echocardiographic data and clinical follow-up were extracted from patient files. Association with hyperinsulinism was reviewed for each underlying cause.
Results: Echocardiograms of 6941 infants were screened. Cardiac hypertrophy was reported in 205 cases. After exclusion of 141 children with Z-scores < 2.0 (n = 72), underlying CHD (n = 56) or hypertension (n = 13) 64 infants remained eligible for analysis. In two-thirds of these children (n = 44;69%) an etiology was identified. Malformation syndromes (n = 21;33%, including

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Noonan n = 10;16%) and maternal diabetes mellitus (n = 13;20%) were most common causes. Less common causes were sarcomeric disease (n = 4;6%), metabolic disease (n = 3;5%), congenital hyperinsulinism (n = 2;3%) and neuromuscular disease (n = 1;2%). In half of the identified causes the etiology was associated with hyperinsulinism (n = 21;33%). Cardiac hypertrophy by hyperinsulinism was diagnosed significantly earlier, had lower LVPWd (Z-scores), a higher IVSd:LVPWd ratio and a more often and faster normalization of cardiac hypertrophy compared with cases without hyperinsulinism (all $P < 0.05$). Metabolic disease, sarcomeric disease and malformation syndromes had higher mortality rates and worse survival than children with maternal diabetes mellitus, idiopathic cardiac hypertrophy, congenital hyperinsulinism or a neuromuscular disorder.



Conclusions: An etiology can be identified in most infants with cardiac hypertrophy. Hyperinsulinism is a causative factor in the development of cardiac hypertrophy in many infants. Echocardiographic variables may distinguish between the different causes of cardiac hypertrophy. Prognosis depends on the underlying cause.

P1-123

Cardiac Malformations Associated with Dextrocardia

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Introduction: Dextrocardia is a cardiac malposition in which the major axis of the heart points to the right in the right hemithorax. It is associated with various congenital cardiac anomalies. There are some studies determining the frequency and type of congenital cardiac malformations associated with dextrocardia. The aim of our study is to determining the congenital cardiac malformations in patients with dextrocardia by using echocardiography.
Method: All patients diagnosed to have dextrocardia in the pediatric cardiology departments of Yuzuncu Yil University Faculty of Medicine between March 2011 and December 2014 and Ipekyolu Women's and Children Hospital between December 2014 and December 2015 were retrospectively examined. 45 patients consist of 27 male (60%) and 18 famale (40%) were enrolled to study.
Results: The average age was 4.98 ± 4.91 years (10 day-17 years, median: 4.0 years). 11 patients (24%) had situs solitus dextrocardia (SSD) whereas 34 patients (76%) had situs inversus dextrocardia (SID). The frequency of congenital cardiac malformation was 72% (8/11) in SSD and 50% (17/34) in SID. 3 patients (%6.6) had atrial situs inversus (2 right atrial isomerism, 1 left atrial isomerism). Among all patients atrial septal defect and ventricular septal defect was the most seen congenital defect (40% and 37%, respectively), followed by double outlet right ventricle (20%), patent ductus arteriosus (17,7%), pulmonary stenosis (15,5%), d-transpositon of the great arteries (13,3%), complete atrioventricular septal defect (11,1%), tricuspid atresia (%11,1). Detected cardiac malformations are listed in table 1). One patient was diagnosed as acute lymphoblastic leukemia, one patient with down syndrome had dilated cardiomyopathia and one patient was received combined bosentan and iloprost theraphy with diagnosis of Eisenmenger Synodrme.
Conclusions: Present study, reconfirms that patients with dextrocardia have various congenital cardiac malformations including severe structural abnormalities that require interventional and surgical procedures. Color Doppler echocardiography can identify these subsets. As the frequency of concomitant cardiac structural abnormalities is high, all patients with dextrocardia have to be evaluated with echocardiography early and routinely for being able to early and appropriate surgical or interventional procedures to correct these complex abnormalities.

Table 1. Dextrocardia and associated cardiac defects

Cardiac Defect	SSD (n:11)	SID (n:34)	Total (n:45)	Cardiac Defect	SSD (n:11)	SID (n:34)	Total (n:45)
ASD	*8 (72%)*	*10 (29%)*	*18 (40%)*	PA	*2 (18%)*	*2 (5,8%)*	*4 (8,8%)*
VSD	*8 (72%)*	*9 (26%)*	*17 (37%)*	l-TGA	*1 (9%)*	*1 (2.9%)*	*2 (4.4%)*
DORV	*2 (18%)*	*7 (20%)*	*9 (20%)*	RAI	*0*	*2 (5.8%)*	*2 (4.4%)*
PDA	*2 (18%)*	*6 (17.6%)*	*8 (17.7%)*	LAI	*0*	*1 (2.9%)*	*1 (2.2%)*
PS	*2 (18%)*	*5 (14.7%)*	*7 (15.5%)*	V-INV	*0*	*1 (2.9%)*	*1 (2.2%)*
d-TGA	*1 (9%)*	*5 (14.7%)*	*6 (13.3%)*	DILV	*1 (9%)*	*0*	*1(2.2%)*
AVSD	*0*	*5 (14.7%)*	*5 (11.1%)*	C-TGA	*1 (9%)*	*0*	*1(2.2%)*
TA	*2 (18%)*	*3 (%8.8)*	*5 (11%)*	TAPVR	*0*	*1 (2.9%)*	*1 (2.2%)*

DORV: Double outlet right ventricle, PA: Pulmonary atresia, VSD: Ventricular septal defect, TA: Tricuspid atresia, TGA: Transposition of the great arteries, ASD: Atrial septal defect, PS: Pulmonary stenosis, PDA: Patent ductus arteriosus, RAI: Right atrial isomerism, AVSD: Atrioventricular septal defect, LAI: Left atrial isomerism, V-INV: Ventricular inversion, TAPVR: Total anomalous pulmonary venous return.

P1-124

Risk Factors for Congenital Heart Disease: A Case Control Study

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Introduction: Congenital Heart Disease (CHD) is the most common congenital problem that accounts for up to 25% of all congenital malformations that present in the neonatal period and is leading cause of neonatal and infant mortality. CHD is associated with multiple risk factors, consanguinity may be one such significant factor. This study was done to find out the risk factors for CHD.
Material and Methods: Study was conducted in the Paediatric department of a Rural Medical College in Central India over a period of 2 years (January 2012-February 2014).
It was a 1:2 matched case control study and for each CHD case two age and sex matched controls were recruited in the study. Cases included were 200 with 400 matched controls. All cases were evaluated on clinical basis and 2D ECHO was done for final diagnosis. Detailed history regarding risk factors was enquired.
Results: Out of the total 200 cases of CHD, 163 were acyanotic heart diseases constituting 81.5% and 37 were Cyanotic heart diseases constituting 18.5% of the total CHD cases. Out of the total cases of CHD 15% of the patients had associated anomalies out of which 30% had chromosomal anomalies and the rest 70% had other organ system anomalies. Significant association between

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prematurity and CHD was observed with OR-16.84. There was no significant association between low birth weight (< 2500 gm) and CHD. We got significant association between maternal age > 30 yrs and occurrence of CHD on univariate analysis. Significant association was observed between family history of CHD and occurrence of CHD. Significant association was observed between tobacco chewing and occurrence of CHD with $p < 0.05$ while there was no association with smoking and alcoholism ($p > 0.05$). History of consanguinity was present in 6% of the cases with CHD and there was statistically significant association between consanguinity and occurrence of CHD with OR -9.60(95%CI:2.97-30.96),$p < 0.001$.

Conclusion: Analyses of our results show that parental consanguinity, tobacco chewing, family history of CHD, maternal age > 30 yrs, and prematurity are independent risk factors for CHD.

P1-125

Organisation of perinatal care for fetuses with congenital heart disese

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Objectives: Perinatal care is important for children with prenatal diagnosis of congenital heart disease (CHD). Close co-operation between obstetrician and fetal cardiologist is needed, as typical obstetric monitoring (CTG) is not always diagnostic. Obstetric and neonatal care and the rate of prenatal diagnosis were evaluated for newborns born with CHD at a tertiary institution.

Methods: A retrospective review of patients born with CHD. Fetal echocardiography results, perinatal care and obstetric outcome was evaluated.

Results: 569 neonates with CHD were delivered between January 2006 and December 2014. 95% were diagnosed prenatally. 30 ventricular septal defects(VSD), 1 tetralogy of Fallot(TOF) and 1 common arterial trunk were missed prenatally. The most common CHD were: VSD14% (93/569), TOF10%(56/569), TGA 9%(50/569), AVSD8%(46/569), HLHS 8%(43/569). Extracardiac abnormalities were diagnosed in 8%(48/569). Rhythm disturbances were observed in 5%(31/569). 267(47%) fetuses were karyotyping, 103(18%) cases of genetic disorders were found with the most common Down Syndrome diagnosed in 8%(45/569). There were 8(1,4%) intrauterine deaths after 22 weeks of pregnancy. There were 79(14%) fetuses with intrauterine growth restriction. Preterm birth occurred in 39(7%).63%(355/569) were delivered vaginally, 22%(127/569) by urgent vaginal or cesarean section, 15%(85/569) by elective cesarean section. There was not electronic continuous monitoring during labor in 4%(21/569) lethally ill fetuses during vaginal deliveries. Perinatal palliative care was introduced for those families during pregnancy and perinatal period. Abnormal intrapartum fetal heart rate patterns according to the SOCG guidelines were observed in143(25%) during the first stage of labor. In 53(9%) newborns Apgar score was less than 7 at 5 minutes, and in 39(7%) umbilical cord blood pH values were lower or equal to 7.20. All other were born in good general condition.

Conclusions: The spectrum of congenital heart defects diagnosed in our institution is wide as patients are admitted from the whole country due to prenatal diagnosis. Majority were diagnosed prenatally what unable optimal perinatal care. There was not difference in neonatal conditions while delivered vaginally or planned CC, what proved that CHD is not an indication for CC. Isolated CHD did not change the result of intrapartum monitoring.

P1-126

ECG changes in mothers during transplacental treatment of fetal supraventricular tachycardia

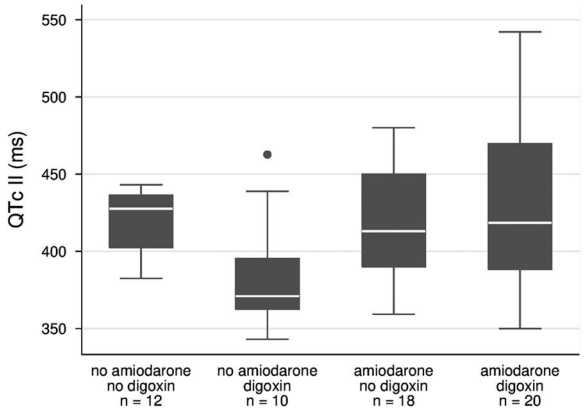
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Introduction: Transplacental treatment of fetal tachyarrhythmias was first introduced about 35 years ago. Due to its specific nature, applying this kind of treatment requires extended assessment of mother's medical history and regular monitoring. Fetal heart rate, mother's ECG, blood pressure and drug levels must be monitored

Methods: We have conducted a retrospective analysis of treatment related to 19 women who were admitted between January 2013 – December 2014. All had negative cardiac history. We studied 60 ECGs (before and during the treatment) with exam frequency related to one subject ranging between 1 and 7. Time span of observation ranged from 1 day up to 100 days. Among 19 investigated subjects 6 were treated with digoxin, 5 with amiodarone and 8 with both drugs. 2 patients required additional third-line propafenone administration.

In mothers' ECG we checked: 1) ST-segment changes (including 'sagging' ST-segment); 2) T-wave changes; 3) QTc; 4) PQ interval; 5) heart rate. We have also employed multivariate linear regression explaining QTc measured in II lead with 1) ST segment change, 2) treatment group in the first specification ($n = 60$) and additionally by 3) cumulative dose of amiodarone and digoxin in second specification ($n = 54$). Obtained estimates were corrected for ECG clusters over subjects.

Results: Among examined ECGs patients ever treated with digoxin, 28 (72%) were characterised by ST-segment changes including 23(59%) 'sagging' ST-segment, 9(23%)T-wave change and 3 (8%) featured first-degree atrioventricular block. None of them revealed bradycardia. In the multivariate regressions QTc is marginally negatively related to digoxin treatment (I specification p-value = 0.15, II specification p-value = 0.10). Amiodarone treatment seems to prolong QTc (I specification p-value = 0.02, II specification p-value = 0.019) with maximal QTc of 542ms. Reported summary statistics are supported by graphical illustration of QTc over treatment groups.



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Conclusions: Digoxin induces multiple ST-segment changes, rarely first-degree atrioventricular block. Amiodarone impacts positively maternal QTc. Treatment consisting in balanced combination of amiodarone and digoxin seems to exposure to various side effects. In spite of ECG changes applied treatment had not increased maternal risk, what is important information while transplacental therapy is necessary.

P1-127

Left ventricular diastolic dysfunction assessed by tissue Doppler imaging predicts clinical course in children with hypertrophic cardiomyopathy

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Background: Tissue Doppler imaging (TDI) parameters have become a sensitive measure of left ventricular diastolic dysfunction (LVDD), which reflects the clinical course in children with hypertrophic cardiomyopathy (HCM). The aim of the study was to determine whether diastolic TDI parameters are predictive of adverse clinical outcome in children with HCM.
Methods: Sixty-three children, median age 14.3years with HCM studied since 2010 to 2014 were enrolled and prospectively followed with respect to TDI results and to clinical endpoints. Patients underwent echocardiography, ECG, Holter ECG, CMR-LGE. The LVDD was diagnosed if one or more parameters were raised in relation to the standards values: transmitral septal/lateral E/E' (z-score > 2), LA dimension (z-score >2), LA volume index (>34 ml/m^2). The clinical endpoints were defined as cardiovascular events: sudden cardiac death (SCD), appropriate ICD discharges (ICDdx), nonsustained ventricular tachycardia (NSVT), supraventricular tachycardia (SVT), syncope, progression of heart failure to NYHA class III. We analyzed also other parameters: chest pain, QTc, the presence of myocardial fibrosis in CMR-LGE. All 63 pts were divided into groups:gI-40(63%) children with LVDD and gII-23(37%) with normal LV diastolic function.
Results: During a follow-up, median 2.5years, 21(33%) children reached the clinical endpoints. In group with LVDD (gI) 19pts achieved endpoints: SCD(n = 3), ICDdx(n = 2), NSVT(n = 3), SVT (n = 2), syncope(n = 2), progression to NYHA class III(n = 7) while in gII without LVDD only 2pts: NSVT(n = 1) and SVT(n = 1); (p = 0.0017).TDI parameters transmitral septal and lateral E/E' demonstrated increased values, furthermore septal E/E' ratio > 2.21 z-score was proved to be an independent predictor of adverse clinical outcome. Moreover significantly larger LA dimension (median z-score 2vs0.7;p = 0.02) and more often LA volume index > 34 ml/m^2 (57% vs 26% pts;p = 0.02) in children with clinical endpoints were found. In children with LVDD increased value of QTc (0.43vs0.41;p = 0.032) and significantly more frequent myocardial fibrosis (33/63 = 52%vs5/63 = 8%;p = 0.0001) were observed.
Conclusions: (1) TDI parameters are sensitive indicators of diastolic LV dysfunction, which is an important predictor of major clinical events and risk of death in children with HCM. (2) Measurement of transmitral septal E/E' should find a place in the routine clinical follow-up of children with HCM. (3) Myocardial fibrosis is highly significant predictor of LVDD in children with HCM.

P1-128

Impact of lung ultrasonography to assess increasing pulmonary blood flow of ventricular septal defect

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Introduction: The lung ultrasonography (LUS) has been recently introduced as a novel diagnostic tool to assess pulmonary congestion. Especially B-lines, ultrasound lung comets, consistent with classical Kerley's B line, suggest excess extravascular lung water. However, there has been few reports of LUS regarding congenital heart disease with left-to-right shunt. We evaluated whether B-lines could result from increasing pulmonary blood flow or not.
Subjects and Methods: We investigated six patients with ventricular septal defect (VSD group) (mean age; 1.2 years, mean Qp/Qs ratio; 2.1) and 11 aged matched control with no cardiac abnormality (control group). LUS was performed with sector transducers of EPIQ (Philips) and Vivid 7 (GE). Each thorax was divided into three areas and LUS finding were recorded on 6 regions. B predominance is defined as more than three positive B-lines. The number of B predominance areas and the presence of bilateral lung B predominance were examined in each group.
Results: The number of B predominance areas was significantly larger in VSD group than in control group (4.0+ /- 1.4 vs 0.5+ /- 0.8, p = 0.001), and bilateral lung B predominance was significantly higher in VSD than in control group (83% vs 18% p = 0.03). However, In VSD group, cardiac catheterization revealed that there was no elevation of pulmonary capillary wedge pressure(8.8+ /- 1.5 mmHg).
Conclusions: Our study suggested that bilateral lung B-lines may be induced not only by pulmonary venous congestion but high pulmonary blood flow. LUS may be helpful to find patients with high pulmonary blood flow due to VSD.

P1-129

Comparison of self- and parent-estimated health-related quality of life with cardiopulmonary exercise testing in Croatian children and adolescents with congenital heart disease

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Introduction: In adult medicine it's established that exercise tests and health-related quality of life (HRQOL) instruments should be used together to get an appropriate overview of the health status of patient with congenital heart disease (CHD). Information on that in children is lacking.
Study analyses the self- and parent-reported HRQOL among patients with CHD, treated in tertiary pediatric cardiology care center, with the aim to compare perception of HRQOL between patients and their parents, and both with cardiopulmonary exercise testing (CPET) parameter - peak oxygen uptake (pVO2)-as the best represent of exercise capacity and cardiopulmonary fitness.

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Methods: A cross-sectional, single-centre, observational study was performed on 63 patients aged 8 to 18 years (40 M, 23 F), with various CHD, under routine follow-up. A significant proportion of patients had previous Fontan procedure (32%). Patients and their parents completed a HRQOL questionnaire - PCQLI®.
We analysed answers on the general health perception item, given at 5-point Likert scale. Afterwards, CPET with Bruce treadmill protocol was performed under same conditions, conducted by one of the authors. Descriptive and inferential statistics were used for data analysis, including calculation of Spearman's rank correlation coefficients for the comparison of HRQOL and pVO2.
Results: To describe the study group, mean pVO2 were compared with reference values of healthy pediatric population, local and from the literature; by Student's t-test, patients value was significantly lower ($p < 0.01$) for both gender.
Excellent matching of self- and parent-estimated health-related quality of life was found: in 65% identical, in 20% parental perception was worse, while in 15% parents underestimate childrens' impairments.
Peak oxygen uptake correlated significantly with the general health perception - of patients ($r = 0.608$, $p < 0.05$), and of their parents ($r = 0.610$, $p < 0.05$).
Conclusions: In general, our patients had reduced exercise capacity. Peak oxygen uptake was in good correlation with the perception of health, made by patients and by their parents: better perception of health mainly means better pVO2. But, in case of mismatching, fatal outcomes can happen. So, complementary usage of these two methods should be helpful in advising patients on activities of daily life, sports participation, and choice of occupation.

P1-130
Cardiac Tamponade – Late Complication of Minimally Invasive (Nuss) Procedure for Pectus Excavatum
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Introduction: Nuss procedure is an established and preffered minimal-invasive technique for pectus excavatum correction. Complications related to the Nuss procedure are not unusual, but major complications rarely occur. Possible complications include pneumothorax, pleuritis, hemothorax, displacement of bar, pericardial effusion, pericarditis and cardiac injury. Pericarditis and pericardial effusion occur in 0.4-4.2% cases. Late cardiac complications have been reported caused by broken or displacement bar, and broken sternal wire, but late complications as cardiac tamponade or large pericardial effusion without clear cause are described very rarely.
Methods and Results: A total of 228 children underwent Nuss procedure in Department of Paediatric Surgery of University Children's Hospital Zagreb since 2001 to 2015 y. All these procedures were done without major cardiac complications except late cardiac complications in three boys (1,3%), 14-16 y old, as cardiac tamponade/ large pericardial effusion without a clear cause, 6 months after Nuss operation.The course of the operations was uncomplicated. Two patients developed pleural effusion, and marginal pneumothorax in the early postoperative period. After recovery from early postoperative complications, the further course was uneventful with normal echocardiography. Dyspnoa and chest pain developed gradually. Echocardiography revealed large pericardial effusion of 2.5 to 5 cm around the whole heart. Two boys had clinical and echocardiographic signs of heart tamponade and pericardiocentesis was necessary. The steel bars were undamaged and their position unchanged. Inflammatory parameters were normal and no microbiological agent was detected. The treatment was continued with nonsteroidal anti-inflammatory drugs. The complete clinical recovery and resolution of the effusion was reached after 8-10 weeks. No recurrence was detected in any of the patients, and there was no indication for the earlier metal bar removal. Two bars were removed two years after the procedure, one is still in place. Etiology and pathogenesis of this process remain controversial, with possible involvement of irritation of the pericardium, autoimmune and (auto)inflammatory pathways.
Conclusions: This cases suggest that surgeons should keep in mind the possibility of cardiac complications without common causes late after surgery.

P1-131
Reduced maximal exercise capacity in adults with small, unrepaired Ventricular Septum Defects
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Introduction: Ventricular septum defect (VSD) is the most common congenital heart defect with 30% closing spontaneously during childhood. The rest are considered as hemodynamically important and are closed surgically, or not hemodynamically important. Small VSDs considered to have no influence usually remain unrepaired, and long term results for living with a small VSD are still undetermined. Some studies have reported of complications occurring later in life, but common for these studies are that patients are all examined at rest. We exercise-test a group of adults with small, open VSDs and a group of healthy, age-matched controls in order to assess long-term physical condition.
Methods: Maximal exercise capacity is determined in participants on an upright bicycle with an incremental workload protocol, chosen individually on the basis of body mass, gender and exercise habits of the participant. Gas exchange is measured breath-by-breath with Jaeger MasterScreen CPX®. Participants are handed the International Physical Activity Questionnaire for assessment of habitual activity level. Prior to the exercise test, all participants undergo bioelectrical impedance analysis, in order to determine their lean body mass for later matching of the groups.
Results: So far, 14 patients and 16 controls have completed the exercise test, with a mean age of 25.4 ± 5 in patients and 25.5 ± 3 in the control group. No differences were found in age, gender or body mass index between the groups. From impedance analysis we found lean body mass in patients to be $73.4 \pm 9\%$, and $75.5 \pm 7\%$ in controls ($p = 0.5$). Both groups reached the targeted test time between 8 – 12 minutes, without differences in maximum heart rate and maximum workload. At maximum exercise level, patients reached a lower maximal oxygen uptake (ml O2/min/Kg) 35.4 ± 7, compared to the healthy controls 45.0 ± 7 ($p < 0.001$). Effort independent endpoint (aerobic capacity) was also lower in patients; 23.3 ± 7 compared to controls 31.1 ± 6 ($p < 0.003$). Neither of this could be explained by a difference in lean body mass, body mass index or the self-reported habitual

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physical activity level of the indivual, calculated from the questionnaire.
Conclusion: These results suggest a reduced cardiopulmonary exercise capacity in adults with small, unrepaired VSDs compared to healthy peers.

P1-132
Cardiopulmonary Capacity in Relation to Right Ventricular Function and Volume in Patients with Fallot Tetralogy
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Introduction: Cardiopulmonary capacity, right ventricular volume and function play a key role in determining the indication and timing for pulmonary valve intervention in patients with Fallot tetralogy.
Methods: Data of 47 patients (23 male, 49%) aged 26.6 (11-54) years, with Fallot tetralogy, who underwent cardiopulmonary exercise testing and MRI between May 2014 and September 2015 was analyzed. Measurements collected in MRI: right ventricular enddiastolic volume indexed to body-surface area (RV-EDV), RV-ejection fraction (RV-EF) and pulmonary valve regurgitation fraction (PV-RF). Maximum cardiopulmonary capacitiy (VO2max) was put in relation to predicted values according to Cooper and Weiler-Ravell.
Results: Mean VO2max of predicted was 71.8 ± 17.9% (range 43 – 122%). MRI data (mean and range: RV volume 114.3 (65.6-149.5) ml/m^2. RV-EF: 47.3 (31.6–61.8)% and PV-RF 19.1 (1.0–43.0). VO2max shows no correlation to RV-EDV: Pearson correlation 0.134, P = 0.379. Mean VO2max of predicted did not correlate to RV-EF: Pearson 0.028, P = 0.855 nor did it correlate to PV-RF Pearson: 0.171, P = 0.267.
Conclusions: RV endiastolic volume, RV Function and PV-Regurgitation measured in MRI are well established factors to make treatment decisions in patients with Fallot Tetralogy, but do not reflect the patient's present exercise tolerance in a group of patients with RV-EDV not exceeding 150 ml/m^2.

P1-133
Complex ACHD patients – 15 years of care in a specialized tertiary care
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In response to the increasing number of adults with congenital heart disease (ACHD), the first specialized tertiary care facility in Hungary developed in 2000. Patients with a complex severity (severe and moderate lesions) have been rigorously followed in our centre. Objectives. To characterize the major features of this largest tertiary ACHD centre in Hungary, with a focus on ambulatory care, hospital admissions and mortality data of those patients with complex disease. Methods: Prospectively collected data were analyzed from our database. Results: A total number of 1391 patients (mean age 31 ± 18 years) have complex congenital lesions (moderate, 30%; severe, 70%). Eighty-three percent of these patients had undergone one or more reparative surgeries. Age at transfer from pediatric to adult care has decreased over a 15 years period from 25-30 to 18-20 years. Compared with year 2000, by 2015 the number of new patients /year increased from 30 to 116; the outpatient visit/week has been 15 times higher, and the overall hospital admissions 7 times higher. Whether in 2000 hospital admissions meant mostly surgical interventions for residual lesion, nowadays the major issues are arrhythmias, heart failure, and pulmonary hypertension. Lapse of follow-up (> 3 years) has been present in 16% of patients with severe and 14% in moderate lesions. Overall, 52 complex patients died. Conclusions: This study gives an insight in the current trends in complex ACHD patients care, characterized by increasing number of patients, more committed patients to specialized follow-up, and better survival.

P1-134
Exercise capacity and lung function in patients late after fontan intervention and impact of physical activity
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Introduction: The Fontan operation have improved survival in patients with functionally univentricular heart. However their exercise tolerance is reduced with a wide range of maximum oxygen consumption. The determinants of such variability are not well understood.
The aim of our study is to evaluate the impact of physical activity on exercise capacity evaluated by cardiopulmonary test (CPT) in patients late after Fontan.
Methods: We retrospectively reviewed patients after Fontan palliation who underwent CPT cardiac magnetic resonance (CMR), and spirometry in our institute. 28 patients (age:17.8 ± 2.9) were included in the study. Self and/or parents-reported physical activity were reported and grouped into no physical activity (Group 0, n:9), scholar physical activity (Group 1 n:10) and regular aeorobic activity at least twice a week (Group 2: n:9). Medical and surgical history was recorded.
Result: No difference between groups was found for systemic ventricle type or for type of Fontan, ventricular volumes and function and cardiac index. There were no significant differences also in lung parameters at rest (FEV, FEV1/FVC, TLC, VT, VD/VT, VE, RR). While the following parameters at peak were significantly different between groups: Watt: Group 0 (72 ± 24) vs Group1 (96 ± 36) vs Group 2 (136 ± 20) $p < 0.01$; Vo2 peak (ml/Kg/min): Group 0 (16.8 ± 3,1) vs Group 1 (22.9 ± 3,9) $p < 0.001$ and vs Group 2 (28.3 ± 2,7) $p < 0.01$; Vo2 (% of predicted): Group 0 (36 ± 13) vs Group 1 (51.6 ± 6,3) and vs Group 2 (57.7 ± 7.5) $p < 0.01$, VE/VCO2 slope: group0 (34.7 ± 7), Group1 (35 ± 3,7) Group2 (29.6 ± 5,7) p = 0.02-Vo2/HR: Group 0 (median 6: 5.2-8) vs Group1 (median 8: 5.8-13): p = 0.004, basal VE/peak VE at peak was higher in the Group 2 (median 64: 54-72) vs Group 1 (median 42: 34-93), p = 0.05 and vs Group 0 (median 37: 34-47,5), p = 0.006. Basal VT/peak VT: group 0 (median 1.02:0.6-1.36), group 1 (median 1.18: 0.78-1.8), group 2 (median 1.83: 1.44-2.1) p = 0.001.
Conclusion: patients after Fontan palliation perform lower physical activity probably due to functional capacity and imposed limitations, and/or psychosocial factors. From our data exercise performance of patients after Fontan palliation is mainly related to physical activity regardless more traditional cardiovascular and pulmonary parameters at rest.

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P1-135

Is pregnancy a risk factor for aortic complications in Marfan syndrome?

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Introduction: Marfan syndrome (MFS) is a pleiotropic disease affecting the skeletal, ocular and cardiovascular organ systems. The cardinal cardiovascular complication is progressive aortic root (AoR) dilatation, entailing a risk for aortic dissection. Whether pregnancy leads to excessive AoR growth or triggers dissection in MFS women under surveillance is unclear.
Objective: We aimed to study AoR growth and the incidence of aortic dissection during pregnancy and during follow up in our MFS patient population.
Methods: We selected all women with molecularly confirmed MFS who had been pregnant between Nov 2011 and June 2015. We retrospectively collected demographic, clinical and echocardiographic data of these patients before and during pregnancy and during follow up. Furthermore we selected a matched group of nulliparous MFS women to analyse aortic root growth during follow up.
Results: There were thirty-seven women aged 20-45years at the moment of the study. Fifteen pregnancies took place in eleven patients. Mean AoR diameter before pregnancy was 36,19mm (IC 95% 32.85-39.53 mm); z-score: 2.27 (IC 95% 0.31-4.23). One woman had previously undergone prophylactic AoR replacement. Six women received beta-blockers throughout eight pregnancies. There were no type A or type B aortic dissections in our cohort. There was a non-significant AoR growth during pregnancy (AoR diameter before pregnancy: 36,5mm vs 38,14mm after pregnancy; $p = 0\,157$). When compared to the matched nulliparous group, the parous cohort showed a significantly faster AoR growth rate (0.96 vs 0.05 mm/yr; $p = 0.002$) during follow up (mean: 5.29 years- IC 95% 1.63-8.95 years).
Conclusions: Pregnancy seems to be safe in terms of risk for aortic dissection in patients with MFS who are under surveillance but can affect AoR growth on the long term. Our study however was retrospective in a limited number of patients. Effect of multiple pregnancies and safety of pregnancy after AoR replacement could therefore not been addressed.

P1-136

Transfemoral Implantation of an Edwards Sapien 3 Valve in the Damus-Kaye-Stansel Anastomosis in a Patient with Fontan Circulation

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Transcatheter pulmonary valve replacement with the Melody valve has fast become an important adjunct in the treatment of children and adults with failing right ventricular outflow tract conduits. Recently, the Melody valve has also been successfully implanted in the tricuspid, mitral, and aortic positions, typically within a failing bioprosthetic valve. The Edwards Sapien valve has been developed for treatment of calcified stenotic aortic valves in elderly patients at high surgical risk. There is limited data on the use of this valve in other positions.
We present a 22-year old patient with univentricular physiology who had already undergone 5 open heart surgeries including palliation with a Damus-Kaye-Stansel (DKS) procedure, Fontan completion and tricuspid valve replacement. In addition, epimyocardial pacemaker implantation and revisions had been necessary. He developed symptomatic free regurgitation of the pulmonary portion of his DKS-anastomosis. To avoid additional high-risk open heart surgery, we successfully implanted an Edwards-Sapien 3 valve transfemorally in the pulmonary portion of the DKS anastomosis relieving the insufficiency. To our knowledge, this is the first successful use of a transcatheter valve in a DKS anastomosis in a patient with single-ventricle physiology. This intervention may increase functional longevity of single-ventricle palliation, postpone the need further surgery, and potentially for orthotopic heart transplantation.

P1-137

Two different types of restrictive physiology in adolescents and adult with repaired tetralogy of Fallot

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Objectives: Restrictive right ventricular physiology (r-RV) has been reported to be favorite late outcome, because it limits pulmonary regurgitation, resulting in less RV dilation and less prolongation of QRS in repaired tetralogy of Fallot (TOF). However, the beneficial effects of r-RV are controversial. The aim of the study was to characterize the clinical differences between with and without r-RV in adolescents and adults with repaired TOF.
Methods: Sixty patients over 15 years old who underwent cardiac magnetic resonance and/or catheterization were reviewed. Blood levels of B-type and atrial natriuretic peptides (BNP, ANP) were measured. Consistent antegrade late diastolic flow in the pulmonary artery by Doppler echocardiography was defined as r-RV.
Results: Median age (range) at the time of evaluation was 25.6 (15.2~62.8) years. The r-RV was identified 18 (30%) of 60 patients. The patients with r-RV had higher levels of ANP, pressure gradients between right atrial pressure (RAp) and diastolic pulmonary artery pressure (PAdp)(RAp – PAdp). There were no differences in RV and LV end-diastolic volumes, ejection fraction (EF), systolic RV pressure, serum levels of BNP, cardiothoracic ratio (CTR) and QRS duration of ECG. Ten of 18 r-RVs had smaller (<150 ml/m2) RV volume. Compared to r-RV with larger RV (≥ 150 ml/2), r-RV with smaller RV volumes had better RV and LVEF, less prolongation of QRS and smaller CTR.
Conclusions: There are two different types of r-RV defined as the antegrade late diastolic flow in the pulmonary artery. The r-RV with small RV may be associated with favorite late outcome, but not r-RV with large RV.

P1-138

Outcomes and prognosis of Total Cavopulmonary Connection performed at adult age

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Purpose: The aim of this study was to investigate the clinical features, assess the clinical course and outcomes, and identify predictive factors for mortality in patients undergoing total cavopulmonary connection (TCPC) at adulthood.
Methods: Single center retrospective analysis of all patients who underwent TCPC at 18 years of age or older, between 1990 and 2015. Patients were classified according to a previous

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atrio-pulmonary connection (TCPC-Conv group), previous palliative surgery (TCPC-Pall group) or absence of previous surgery or bidirectional Glenn anastomosis only (TCPC-Nopall group). Vital status, clinical events during follow up, functional status at last medical visit and quality of life were recorded.
Results: Thirty five adult patients underwent total cavo-pulmonary connection, 19 in TCPC-Pall group, 7 in TCPC-NoPall group, 9 in TCPC-Conv group. Median follow up was 7 ± 6,7 years. Actual survival was 80, 73,2 and 68,3% at respectively 1, 5 and 10 years. There was no significant difference in mortality between surgical groups (22% in TCPC-Conv, 14% in TCPC-NoPall, 42% in TCPC-Pall, p = 0,3). Early mortality was 17%. Mortality was significantly higher in patients in which a fenestration was performed at the time of surgery (100% vs. 22,5%, p = 0 005) and albumin blood levels at last follow up was significantly lower among deceased patients (mean 31.3 g/l versus 41.9 g/l, p = 0.04). TCPC resulted in mean NYHA improvement in every groups (TCPC-Conv: 2,5 ± 0,5 vs. 1,5 ± 0,8 p = 0.01; TCPC-NoPall: 2,7 ± 0,7 vs. 1,7 ± 0,9 p = 0 004; TCPC-Pall: 2,6 ± 0,6 vs. 1,9 ± 0,8 p = 0.02). The frequency of arrhythmia episodes decreased from 60.6% to 22.2% at latest follow-up (p = 0.5), but preoperative PLE in conversion group did not resolved.
Conclusion: Total cavo-pulmonary connection in adults can provide a benefit in long-term survival and symptoms improvement in selected patients. However high postoperative mortality may lead to renunciation or foreseeing postoperative care in high-risk patients as fenestration did not improve outcome

P1-139
Impact of conversion from classic Fontan to total cavopulmonary connection on adults with single-ventricle short and long-term outcomes
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Objective: The aim of this study was to assess the beneficial effects of conversion from classic Fontan to total cavopulmonary connection (TCPC) in adults with univentricular heart.
Methods: All patients who underwent conversion from atrio-pulmonary to total cavopulmonary connection. Preoperative and postoperative clinical, echocardiographic data were collected. Long term outcome was assessed.
Results: Nine patients (7 males), all with tricuspid atresia, were converted from Fontan to TCPC at a mean age of 26.8 years. Heart failure was present in 33% of the cases, and arrhythmias occurred in 100% before conversion. Preoperative ventricular ejection fraction was 54.7%. Duration of bypass, CICU stay, hospital stay and postoperative drainage was respectively 145mn, 5.3days, 24 days and 8 days. Median follow-up after surgery was 7 years. Two early deaths occurred (22%), no late death. Survival at 1 and 10 years was 80%. NYHA class I-II patients increased from 44% to 84%, NYHA class III decreased from 56% to 17%, while heart failure decreased from 33% of the patients to 17%. Arrhythmias frequency lessened from 100% of the cases before conversion to 57% postoperatively and 33% of the patients in later follow-up. Only one patient suffered from protein losing enteropathy.
Conclusion: Despite an early postoperative mortality of 22%, conversion to TCPC in adulthood can substantially decrease arrhythmias and heart failure frequency, and improve NYHA class.

P1-140
Multivalve surgery in adults with congenital heart disease
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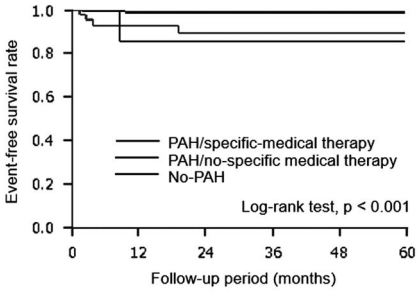
Introduction: Grown-up congenital heart (GUCH) patients are a growing population formed by youth people with high complexity related to the particular type of cardiopathy itself and previous sugeries. During the last 12 years in our GUCH unit we have operated 232 patients with extracorporeal circulation (ECC). Multivalve surgery is growing along time, reaching 18,9% of our whole activity. We present here our experience with these patients requiring simultaneous surgery in 2 or more cardiac valves.
Methods: Retrospective study of 44 patients operated in our GUCH unit (surgery in 2 or more cardiac valves at the same time), in the period (December-2003-November-2015). Statistical study with SPSS-15.0
Results: Mean age: 40 ± 12 years (range: 18,75-74), 54,5% women. Main heart disease: Fallot in 52,3% and pulmonary stenosis in 15,9%. Previous surgeries in 95,5%. Preoperative functional class III-61,5% and IV-12,8%, history of arrhythmia in 69,8%. Principal indications for surgery related to valve incompetence: pulmonary-79,1%, mitral-14%, aortic-7%. Surgical techniques employed: 89% with 2-valve surgery, 11% with 3-valve surgery, more frequent surgery performed: pulmonary bioprosthesis + tricuspid plasty/bioprosthesis in 23 patients (52%). Associated surgery in 26 patients (59%) mainly consisted in closing a residual septal defect. Median ECC time was 194 minutes (IQR: 135-292), and aortic cross-clamp time was 136 minutes (IQR: 99-152). Hospital mortality: 2(4,5%), was better than expected with the preoperative risk scales (97,7% in RASCHS category-3, mean EACTS: 7,5 ± 0.81, and mean Euroscore: 9,7 ± 7,7). Hospital morbidity: 14(33%).
Follow-up was complete, with mean 3,9 ± 3,9 years (maximum 12,5). Late mortality 2(4,7%), without new reoperations with ECC. Nowadays, functional class is I-37,5% and II-53,1%.
Conclusions: Multivalve disease in GUCH patients is a challenge with high risk. We operate these patients in our GUCH unit with good results (low mortality and improvement of clinical status), in low and medium-time follow-up.

P1-141
Long-Term Efficacy of Treat and Repair Strategy for Atrial Septal Defect with Pulmonary Artery Hypertension
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Introduction: Therapeutic strategies for atrial septal defect (ASD) with pulmonary artery hypertension (PAH) are controversial. Recently, the efficacy of treat and repair strategy (PAH specific-medical therapy and transcatheter ASD closure) is introduced, however, the long-term effects remain unknown. The purpose of this study was to evaluate the long-term efficacy of the treat and repair strategy for ASD with significant PAH (mean pulmonary artery pressure (mPAP) ≥ 25 mmHg and PVR ≥ 3 wood units).
Methods: A total of 616 adult patients who underwent transcatheter ASD closure were divided into 3 groups: PAH/specific-medical therapy (n = 11), PAH/no-specific-medical therapy (n = 43), no-PAH (n = 562). The endpoint was defined as cardiovascular mortality and hospitalization for heart failure.

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Results: Mean pulmonary artery pressure (PAP) before the treat and repair strategy was 56 ± 21 mmHg in PAH/specific-medical therapy group. Initially, the PHM group had higher PVR compared with non-PHM group (9.6 ± 3.8 vs. 4.2 ± 1.0 Wood units, P < 0.01). After treatment with PAH-specific medications, PVR in this group decreased to 4.0 ± 0.8 Wood units (P < 0.01). During a median follow-up of 24 months (1-110 months), the event-free survival rate in PAH/specific-medical therapy group was inferior compared to that in no-PAH group (log-rant test, p < 0.01), however it was not different from that in PAH/no-specific-medical therapy group (p = 0.87). More than 90% of patients with PAH/specific-medical therapy had no cardiovascular events. In the PHM group, during a treatment period of 52 ± 48 months, the World Health Organization Functional Classification significantly improved (3.0 ± 0.5 to 2.0 ± 0.0, P < 0.01), as well as in the non-PHM group (2.1 ± 0.6 to 1.5 ± 0.5, P < 0.01).



Patient at risk						
PAH/specific-medical therapy	11	5	3	1	1	1
PAH/specific medical therapy	43	33	21	7	3	1
No-PAH	562	317	182	115	8	2

Conclusions: Treat and repair strategy for ASD with severe PAH can be considered as a safe and valuable therapeutic option even in patients complicated with significant PAH.

P1-142

Haemodynamic impact of pulmonary vasodilators on Fontan circulation

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Introduction: The Fontan circulation is the current palliation for patients with a single ventricle physiology. The absence of a subpulmonary ventricle makes low pulmonary vascular resistance and optimal systemic ventricular function the essential elements of a successful Fontan circulation. The aim of this retrospective study was to investigate the potential effect of pulmonary vasodilators on pulmonary vasculature in Fontan patients.
Methods: Eighteen single ventricle patients in therapy with pulmonary vasodilators from 6 months or more were enrolled. Nine of these patients were in therapy after Glenn procedure or just after the Fontan completion (Group A) and nine more than 5 years after Fontan completion (Group B). Ten patients without pulmonary vasodilators who underwent two catheterization at a maximal distance of two years (Group C).
Data from two different right catheterizations (before starting therapy and 6 to 24 months after) were collected: mean pulmonary pressure (mPAP), arteriolar pulmonary vascular resistances (PVR), pulmonary capillary wedge pressure (PCW), transpulmonary pressure gradient (dP), Cardiac output (CO), pulmonary blood flow (PBF), right pulmonary artery (RPA) and left pulmonary artery (LPA) diameter, Nakata index and pulmonary arterial compliance (PAC).
Results: After starting therapy, PBF (p = 0.04), PVR (p < 0.001), RPA and LPA diameters (p < 0.001) and Nakata index (p = 0.001) improved after starting therapy. These results were confirmed analyzing the subgroups A and B independently.
At baseline Group A had a smaller Nakata Index (p = 0.047) and a tendency to higher mPAP and higher PVR than Group C. At the second catheterization the PCW decreased only in Group C (p = 0.039). dP (p = 0.025) and PVR (p = 0.009) decreased in group A. Nakata Index (p = 0.003) and PAC (p = 0.001) increased in group A.
Conclusions: Pulmonary vasodilators reduce PVR and increase PAC, Nakata index and CO. Pulmonary vasodilators could be used before Fontan completion in selected patients as 'bridge to Fontainability'. In adults the best effect was seen in patients with higher dP and lower PAC. In patients with low dP and high PCW the treatment increased cardiac output but also wedge pressure raised. For these reasons, given also the costs of the therapy, we suggest to perform the heart catheterization before starting this therapy.

P1-143

Risk factor for Heart failure admissions in adults with congenital heart disease in monocentric tertiary center

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Adult with CHD are now more important than children with CHD, and this population is a rapidly growing population. Heart failure (HF) is a serious complication in the long-term follow-up, and is one of the main causes of death. Therefore, a substantial increase in hospitalizations because of HFin ACHD is observed, requiring specialized care and making this problem an important public health issue
Aims: We aimed to characterize HF in ACHD and its management in a tertiary center. We also wanted to identify risk factors of first HF-admission.
Methods: We retrospectively assessed the medical records of 408 admissions of ACHD in our center. Risk factors for HF-admission were assessed using regression logistic models.
Results: 408 patients were admitted during a median follow up period of 14 months. HF criteria were met by 29 patients (7,1%). ACHD with HF were significantly older than other ACHD patients admitted (median age was 43 vs 33 years, p = 0.001). They had more complex CHD (62%), they were mainly patients with history of RV outflow tract surgery, single ventricle and PAH (p = 0.007). The aetiologies of HF were myocardial dysfunction (n = 15), valvular disease (n = 5), pulmonary hypertension (n = 4), arrhythmia (n = 3) and infective endocarditis (n = 2). Mean hospital stay of ACHD patients with HF was longer (13 days vs 5 days, p < 0.0001). Ten percent (3/29) died at a mean period of 23 days after their admission, one patient required circulatory support, and 2 patients were listed for heart transplantation. Independent risk factors for HF-admission were history of stroke (OR: 7,6 95%CI[2,4-23,8], p), abnormal rhythm conduction (OR: 4,2 95%CI[1,5-11,6], heart failure (OR = 4,7 95%[1,6-13,7], p < 0.01)), and atrial arrhythmia (OR: 3,5 95%CI[1,4 to 9,2]). Number of cardiac surgeries was not a risk factor of HF. At admission systemic ventricle ejection fraction was the factor the most strongly associated with HF.

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Conclusions: Mortality risk is substantially increased after HF-admission, which emphasizes the importance to identify patients at high risk of HF-admission. These patients might benefit from closer follow-up and earlier medical interventions. This may add in care of patients with ACHD in the community and streamline care at tertiary centers.

P1-144
Pregnancy in women with percutaneous pulmonary valve implantation

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Introduction: Pulmonary valve replacement (PVR) is indicated in adult patients with dysfunctional right ventricular outflow tract. There is little data on pregnancy following PVR, particularly after percutaneous pulmonary valve implantation (PPVI). The aim of this study was to analyze pregnancy outcome in these patients.
Methods: Among 106 adult women with PVR followed in our center, 13 were pregnant. PPVI was performed in 7 of them, 6 ± 2 years before pregnancy (Melody® valve). We retrospectively collected obstetric and cardiologicdata.
Results: 7 patients with PPVI had 10 pregnancies at a mean age of 29 ± 6 (ranged from 17 to 38). Five led to a delivery after 20 weeks gestation (WG), and 3 had an abortion. No miscarriage occurred. 3/5 pregnancies were delivered by cesarean section, for obstetrical indications. Obstetric complications occurred in 2/5 complete pregnancies: one severe preeclampsia leading to a premature birth at 30WG, and one spontaneous preterm labor at 35 WG. There was no small for gestational age neonates. The mean gestational age at birth was 36 ± 3WG. No congenital heart disease was diagnosed in the newborns and there was no neonatal or fetal death. During complete pregnancies no maternal cardiac complication occurred. One patient died from endocarditis in the aftermath of an abortion. All patients were treated by aspirin throughout pregnancy and received antibiotic prophylaxis at delivery. The patient who died after abortion received antibiotics prophylaxis but had no aspirin.
At median follow-up of 14 months, there was no change in the trans-pulmonary valve maximal gradient (31 vs 27 mmHg) and no pulmonary regurgitation.
Conclusion: This small first series of pregnancies with PPVI seems reassuring for the maternal and neonatal outcomes, except for the risk of infection, which needs careful monitoring by experienced teams.

P1-145
New hybrid approach for complex re-coarctation involving supra-aortic branch

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Introduction: Stent repair of aortic re-coarctation is become a valid alternative to surgical correction. Several anatomical characteristics of the aortic re-coarctation involve the origin of a supra-aortic trunk, composing the clinical situation known as 'complex aortic re-coarctation' which may determine challenging situation for percutaneous treatment. We describe a new hybrid approach for treatment of stenosis involving the supra-aortic branch in two cases of complex aortic re-coarctation after previous surgical correction. In both patients the stenosis involved left subclavian artery.
Methods: A trans-isthmic gradient of 35 and 18 mmHg, was respectively detected in the 2 patients. The hybrid approach was performed within the operating room by a surgical vascular step characterized by a carotid-subclavian bypass in both patients, followed by an interventional concomitant step. The Interventional step was different for the 2 patients:

- A thoracic endoprosthesis GORE C-TAG 21-21-10, was implanted in the 41-year-old female pt, in order to exclude a chronic dissection of the aorta distal to the coarctation site, plus a 39 mm CP bare metal stent dilated with a NuMedInc Balloon-in-Balloon 14×45 mm.
- A covered CP stent 45 mm long dilated with aNuMedInc Balloon-in-Balloon 16×45 mm was implanted in the 15-year-old male patient.

Results: Both intervention had immediate treatment success achieving a final pressure gradient < 15 mm Hg. Patients were discharged without post procedure minor and/or major complications. At 6-month follow-up both patients were asymptomatic with neither diastolic run-off nor carotid-subclavian bypass significant gradient at transthoracic echocardiogram.
Conclusions: Our experience supports the idea that an hybrid approach to complex re-coarctation involving supra-aortic branch is a safety and efficacy tecnique to treat it.

P1-146
Factors Associated with Renal Dysfunction in Adults with Fontan Circulation

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Introduction: It is not uncommon for adults with Fontan circulation to have multi-organ complications. However, only a few studies have described the frequency and degree of renal dysfunction in adult Fontan patients. This study aimed to elucidate factors associated with renal function in adult Fontan patients.
Methods: We performed a retrospective chart review of 21 adult Fontan patients (age 29.9 ± 7.2 years, 13 males) who had undergone postoperative cardiac catheterization. Renal function was evaluated by calculating estimated glomerular filtration rate (eGFR) based on serum creatinine level at the cardiac catheterization. Demographic data and laboratory results including platelet counts, serum levels of total bilirubin and gamma-glutamyltranspeptidase, plasma brain natriuretic peptide (BNP) concentration, and arterial oxygen saturation (SaO_2) were collected. The hemodynamic measurements obtained by cardiac catheterization included central venous pressure (CVP), cardiac index (CI), and pulmonary vascular resistance (PVR). The correlation between eGFR and the aforementioned parameters was assessed.
Results: Fontan procedures (4 atriopulmonary connection, 8 extracardiac conduit including 4 converted from atriopulmonary connection, and 9 lateral tunnel) were performed at the mean age of 8.9 ± 5.1 years. The time interval since the Fontan procedure was 20.3 ± 4.5 years. No patient had symptomatic renal failure, and eGFR was 94.6 ± 17.6 mL/min/1.73 m^2. Fourteen patients (67%) were on either angiotensin converting enzyme inhibitors or angiotensin receptor blockers. Platelet counts were $140 \pm 37 \times 10^9$ /L, total

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bilirubin was 1.1 ± 0.5 mg/dL, gamma-glutamyltranspeptidase was 120.5 ± 98.3 IU/mL, BNP was 67.8 ± 142.6 pg/mL, and SaO_2 was 92.1 ± 3.6%. As for hemodynamic measurements, CVP was 11.6 ± 2.2 mmHg, CI was 2.5 ± 0.7 L/min/m^2, and PVR was 1.9 ± 1.0 Wood units·m^2. A statistically significant correlation was found between eGFR and age at examination (r = –0.51, p = 0.017), the time interval since the Fontan procedure (r = –0.55, p = 0.010), CVP (r = 0.56, p = 0.008), and CI (r = 0.60, p = 0.003). There is no significant correlation between renal function and liver function.
Conclusions: In adult Fontan patients, renal dysfunction is mild and rarely symptomatic. The degree of renal dysfunction is associated with hemodynamic integrity of Fontan circulation that is reflected in CVP and CI.

P1-147
A Rare Form of Transposition of Great Arteries: Two Cases of Posterior Aorta

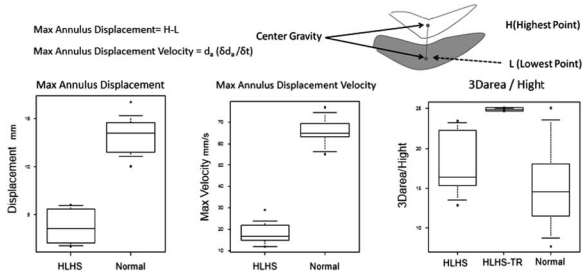
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Introduction: Transposition of the great arteries (TGA) with a posterior aorta is a very rare form of TGA characterized by a right-sided and posterior aorta arising from the right ventricle.
Case reports: A 3 days old male patient with a 3.2 kg of body weight admitted to the emergency department with cyanosis and tachypnea. The oxygen saturation was 78%. There was a 2/6 murmur on left sternal side on physical examination. The second case was a 2 months old infant weighing 4.3 kg who admitted to the clinic with tachypnea. Similarly on echocardiographic evaluation of both patients, there was a concordant atrioventricular but discordant ventriculoarterial relation. The relation between aorta and pulmonary artery was normal where aorta was posterior and to the right of the pulmonary artery. A doubly committed ventricular septal defect, patent foramen ovale and systemic pulmonary hypertension were demonstrated in both cases. Aortic arch hypoplasia was present additionally in the second case. An arterial switch without a Lecompte maneuvre, pericardial patchplasty of VSD, PDA division was performed in both patients where additionally aortic arch reconstruction was performed in second patient. Moderate degree pulmonary stenosis was demonstrated on follow up echocardiographic evaluations of the second patient.
Conclusion: TGA with posterior aorta is a rare entity, without a known reported incidence. We want to emphasize that the diagnosis of TGA can be difficult when pulmonary trunk is anterior and also during the operation although a lecompte maneuvre was not needed, the posterior location and the length of the left coronary artery can be an obstacle in its transfer.

P1-148
The impact of three-dimensional morphology of tricuspid valve on tricuspid regurgitation in patients with hypoplastic left heart syndrome

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Aim: To investigate the impact of the three-dimensional(3D) morphology and functions of tricuspid valve (TV) on tricuspid valve regurgitation (TR) in patients with hypo plastic left heart syndrome(HLHS).
Methods: The subjects were 9pts with HLHS after Fontan procedure (mean age of 10.8y) and normal healthy children(10.2y). The 3D volume data of TV was obtained from apical four chamber view by iE33 /X5-1 probe with 6 beats full volume data acquisition and stored. The acquired data was analyzed by off-line soft ware (MV-Assessment 2.3 of Tom-Tec Inc.) for 3D morphology of TV and functions.
Results: The 3D area of TV was larger in HLHS than in N (10.6 ± 3.1:8.9 ± 3.2 cm2), the height (Ht) was lower (0.5 ± 0.1: 0.6 ± 0.08 cm), 3Darea/Ht was higher (19.7 ± 4.7: 15.3 ± 5.3 cm). The displacement of TV (19.7 ± 4.7: 15.3 ± 5.3 cm) and annular velocity (18.3 ± 5.7: 65.7 ± 7.2 mm/s) were lower in HLHS than in N.
In conclusion: The 3D morphology of TV in HLHS is different from those in N, and the restricted mobility and lower annular velocity of TV might be prone to develop valve regurgitation.



P1-149
CHD Variant Dependence on Toxic Substances Content in Cardiovascular Tissue

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Intoduction: Etiology of congenital heart disease remains unsolved problem of pediatric cardiology. There are about 30 metals and metalloids that are potentially toxic to humans. Some of them, according to the experimental data, able to penetrate the placental barrier and violent cardiogenesis.
Methods: We had determined content of toxic substances in 107 biosubstrates of heart and great vessels of 55 patients with CHD: 34 boys and 21 girls in age from 14 days till 17 years. Among 31 CHD variants 17.7% of patients had heart malformations, 22.6% – great vessels, the combination of heart and vascular malformations – 59.7%; simple defects – 21.8%, combined (2 to 8 variants) – 78.2%. 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 content in heart and great vessels tissue by methods of the atomic emission spectrometry in the inductively coupled plasma and atomic absorption spectrometry with electrothermal atomization.
Results: The presence of 12 toxic metals and metalloids (Al, Ba, Li, Ni, As, Sr, Pb, Zr, Sn, Ti, Ag, W) was documented in the

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cardiovascular tissues of patients with severe combined CHD, compared with children who had simple defects (9 substances). The average concentration of Ni, W, Ti and Ag ($p \leq 0.05$) was higher in patients with two or more malformations compared with children who had one. The presence of 3 and more toxic metals in pathological concentrations had 31.8% patients of complex combinative CHD, which were statistically significantly more higher than in patients with simple defects (7.1%), $p < 0.05$. The greater amount of toxic metals in pathological concentration and a high concentration of Ba, As, Pb, Ag, Ti, Zr, Sn was revealed in patients with combined defects (heart and great vessels) compared with patients with isolated heart or great vessel defects.
Conclusions: Our results had determinate dependence on CHD complexity of toxic metals and metalloids quantity and concentration in cardiovascular tissue, that may indicate possible pathological impact of toxic substances on cardiogenesis violation in children.

P1-150
Portal vein anatomy and pulmonary complications of polysplenia syndrome: difference between patients with biventricular and univentricular heart disease
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Objectives: Polysplenia syndrome is associated with a variety of cardiac anomalies and often accompanied by abdominal and pulmonary vessel abnormalities including portosystemic shunt (PSS), portal vein hypoplasia, pulmonary arteriovenous fistula (PAVF) and pulmonary arterial hypertension (PAH). PSS and PAVF may be congenital or acquired, which makes pathologic condition more complicated. The purpose of this study was to explore the relationship of the type of heart disease and PSS, portal vein hypoplasia, PAVF, and PAH in polysplenia syndrome.
Methods: We conducted a retrospective observational study including 14 consecutive heterotaxia patients with interruption of IVC and azygos or hemiazygos vein connection admitted to our institution during 2002 and 2014. Abdominal and pulmonary vessels were evaluated using ultrasonography, 320-raw multi-detector computed tomography and angiography.
Results: Seven patients (age 1-19 years) had biventricular (BV) heart disease and 7 patients (age 0.3-14 years) had univentricular (UV) heart disease. Both BV and UV patients were often complicated by anomalous renal and splenic venous return. In the UV group, PSS was found in 4 patients, PAVFs in 3 patients and PAH in one patient. Two patients had both PSS and PAVFs. The other two patients with PSS did not have PAVF. One patient with PAVF was not accompanied with PSS but demonstrated abnormal pulmonary distribution of hepatic venous flow after Fontan operation with extracardiac total cavopulmonary connenction. Hypoplastic intrahepatic portal vein was found in an infant with the most severe form of diffuse bilateral PAVFs associated with large PSS and PAH prior to any surgical intervention. Ligation of PPS in this patient resulted in incomplete resolution of PAVFs without improvement in PAH. Neither PSS, portal vein hypoplasia, PAVF, nor PAH were found in the BV group.
Conclusions: PAVFs were found in both congenital and postoperative secondary forms. PSS, together with abnormal pulmonary distribution of hepatic venous flow in cavopulmonary circuit in UV heart disease, may contribute to the development of PAVF. A combination of hypoplastic portal vein and PSS may lead to congenital PAVF. Detailed evaluation of the abdominal venous anatomy is crucial in patients with polysplenia syndrome.

P2-1
Pulmonary Hypertension in Paediatric Patients: data from the COMPERA-KIDS registry
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Background: Pulmonary hypertension (PH / PAH) can have many possible causes in childhood. The aim of the COMPERA registry is the characterization of patients of all ages with PH / PAH and their treatment patterns.
Methods: Since June 2013, paediatric patients can be included in the COMPERA registry (ClinTrials.gov: NCT01347216) which has originally been established for adult patients with pulmonary hypertension in 2007.
Results: Until November 2015, 102 patients < 18 years with pulmonary hypertension were enrolled. Of these paediatric patients, 65.7% had PAH due to congenital heart disease (PAH-CHD), 18.6% had idiopathic PAH (iPAH), 6 had persistent PH of the newborn (PPHN), and 2 had PH associated with interstitial lung disease, valvular heart disease or congenital malformations, respectively. The patients were 5.2 ± 5.9 years old, 58 female; NYHA functional class I / II in 59%, III in 36%, and IV in 5%. The average disease duration after diagnosis was 31.7 ± 52.0 months. Invasive measurement data by right heart catheterization were available for 78% of the patients. The mean pulmonary artery pressure was 45 ± 20 mmHg, the right atrial pressure was 7.9 ± 8.0 mmHg, cardiac index was 3.7 ± 1.3 l/min/m2. Monotherapy was received by 66% of the patients, 28% of the patients had a dual and 4% a triple therapy. Phosphodiesterase-5 inhibitors were administered to 81% of the patients, 33% of the patients received endothelin receptor antagonists and 3% received prostacyclins.
Conclusion: The most common form of PH in this study cohort is PAH due to congenital heart disease, followed by idiopathic PAH. Invasively measured right heart pressure data were available for 80% of the paediatric patients. Treatment options for children primarily comprise phosphodiesterase-5 inhibitors, although endothelin receptor antagonists are increasingly used. Only a small number of paediatric patients receive anticoagulation therapy.

P2-2
Everolimus – Initial side effects of everolimus should not exclude patients from long-term therapy – observation in 63 patients after pediatric heart transplantation
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Background: Everolimus, the modern mTor-inhibitor, has the potential benefit of reduced side effects such as nephrotoxicity, risk of infections and risk of secondary malignancies. As many patients suffer from relevant side effects (such as stomatitis and

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gastrointestinal problems) during initial treatment, many patients after pediatric heart transplantation might be excluded from the potential benefit of everolimus.
Methods: Retrospective analysis of all of our patients under immunosuppressive therapy with everolimus. We focused on the reasons for switch of the immunosuppressive therapy to everolimus and the side effects that led to an interruption or termination of the therapy with everolimus.
Results: 63 patients after pediatric heart transplantation were switched to an everolimus based immunosuppressive therapy between January 2007 and October 2015. The therapy with everolimus was started in average 5.9 years after transplantation. Only five patients were switched to calcineurin inhibitor free therapy, the remaining patients had a combination therapy with cyclosporine A (n = 2) or with tacrolimus (n = 56). After 2013 everolimus was standard therapy for all patients (n = 20). Other reasons for therapy with everolimus were vasculopathy (n = 15), renal insufficiency (n = 11) and gastrointestinal problems (n = 10). There was a mild improvement of the renal function and gastrointestinal problems especially 6 months after change of the immunosuppressive medication. The majority showed a stabilisation or even a mild improvement of the vasculopathy under treatment with everolimus.
However, almost one third of the patients (n = 18) discontinued the treatment with everolimus. The most important reasons were stomatitis (n = 6), gastrointestinal (n = 5) and haematological problems (n = 3). Based on the excellent clinical effect of everolimus we could convince 14 of 18 patients to restart everolimus about one year later. Thereafter 70 percent (n = 10) tolerated everolimus without the previously reported side effects.
Conclusion: The immunosuppressive therapy with everolimus is a save possibility even in pediatric patients who do not tolerate everolimus during the initial treatment.

P2-3
Tacrolimus once daily – Safe and effective after pediatric heart transplantation

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Background: Tacrolimus twice daily is the commonly used tacrolimus formulation after pediatric heart transplantation. In general, decreased adherence to regular intake of medication is a huge problem in pediatrics. To improve the situation tacrolimus once daily was developed. Several studies could verify the safety and effectiveness of tacrolimus once daily in adults; however, mild increase of the daily dose is required to achieve the target levels. If these findings also apply to pediatric patients is unknown yet.
Methods: Retrospective analysis of all patients after pediatric heart transplantation treated with tacrolimus once daily, special focus on the absolute daily dose required.
Results: Between 2012 and October 2015 44 patients after pediatric heart transplantation were switched from tacrolimus twice daily (n = 42), cyclosporine A (n = 1) and everolimus monotherapy (n = 1) to tacrolimus once daily. This change of the medication was in average 6.7 years after transplantation. In 25 cases, an evaluation of the daily tacrolimus dose before, one month and six months after change of the medication was possible. These patients needed an average daily tacrolimus dose of 2.5 mg under tacrolimus twice daily. The mean daily dose increased to 2.8 mg one month and to 3.0 mg six months after switch of the medication. Altogether, the development of the daily dose of tacrolimus was quite different. Six months after switch of the medication the daily dose of tacrolimus ranged between a decrease of 38 percent and an increase of 317 percent in comparison of the daily dose of tacrolimus before switch of the medication. None of the patients suffered from new side effects. Only one patient showed fluctuating tacrolimus trough levels, so that reinstitution of tacrolimus twice daily was necessary. In addition, patient compliance to the medication and quality of life were assessed by a patient questionnaire.
Conclusion: Based on our results we can recommend tacrolimus once daily as a safe and effective regime after pediatric heart transplantation. Single daily dose improves the adherence to the medication and the quality of life. Careful evaluation of the blood levels of tacrolimus is necessary during initiation of the medication.

P2-4
The analysis of myocardial deformation and function in patients with Kawasaki disease by speckle tracking and tissue Doppler methods

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Objectives: It was aimed to investigate myocardial deformation and function at acute and convalescent phases of Kawasaki disease (KD) in children.
Methods: The speckle tracking echocardiography (STE) and tissue Doppler imaging (TDI) were performed in 9 children with KD (mean age 5.4 years) and 10 healthy children (mean age 5.9 years). In the patient group, data were obtained during acute and convalescent phases of Kawasaki disease. The analysis of myocardial deformation [strain (S) and strain rate (SR)] was performed globally in two planes [longitudinal (L) and mid-circumferential (C)] at both left ventricle (LVLGS, LVLGSR, LVCGS, LVCGSR) and right ventricle (RVLGS, RVLGSR). The tissue Doppler studies [Sm, Em, Am, isovolumic contraction time (ICT), isovolumic relaxation time (IRT) and ejection time (ET)] were performed at base of interventricular septum (IVS), LV and RV.
Results: Among tissue Doppler parameters, Em and ET at IVS, ET at RV obtained before treatment were significantly lower in patients compared to controls (11.3 cm/s, 210.8 ms and 193.8 ms vs. 13.1 cm/s, 214.3 ms and 242.1 ms, respectively). After therapy, it was seen that Em at IVS remained significantly lower in patients compared to controls (11,2 vs. 13.1 cm/s).
The LVLGS, LVLGSR and LVCGSR values obtained before therapy were significantly lower in patients compared to controls (-20%, 0.2 s-1, 0.43 s-1 vs.-23.7%, 0.65 s-1, 0.91 s-1, respectively). At the end of the convalescent phase, longitudinal and circumferential global strain and strain rate values were found to be increased compared to acute phase. Besides, no significant difference was detected for LVLGS, LVLGSR, LVCGS and LVCGSR after therapy compared to controls (-24.3%, 0.45 s-1,-24.6%, 0.61 s-1 vs.-23.7%, 0.65 s-1,-26.2%, 0.91 s-1, respectively). There were no significant differences in RVLGS and RVLGSR values between patients before therapy and controls (-27.2%, 0.51 s-1 vs.-24%, 0.76 s-1, respectively).
Conclusions: STE and TDI that evaluates myocardial deformation and function can be used for the detection of myocardial dysfunction in patients with KD. This study showed spared myocardial functions at RV and gradual improvements in myocardial functions at LV during therapy in KD.

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P2-5

Right Atrial and Ventricular Function After Surgical and Percutaneous Closure of Atrial Septal Defect: A Strain Rate Imaging Study

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Basics: Two-dimensional (2D) strain analysis is a new tool to assess myocardial function. Strain and strain rate(SR) can quantify local myocardial function independent of the heart motion. The aim of this study was to compare the effects of surgical and device closure of atrial septal defects(ASD) on atrial and ventricular performance assessed by using strain and SR imaging.
Methods: In all, our study consisted of 45 patients : 15 patients after successful ASD device closure (ASD-D, atrial septal defect device closure group) aged 7.8+ 4.3 years ; 15 patients after successful ASD surgical closure (ASD-S, atrial septal defect surgical closure group) aged 7.5+ 4.6 years and 15 healthy subjects of similar age distribution and characteristics as control group. All patients underwent ASD correction at least 6 months before the study. Peak right ventricular (RV) longitudunal strain, RV lateral and septal strain, peak atrial longitudinal strain (es) and SR during systole (SRs), SR during early RV filling (SRe) and late RV filling (SRa) were measured.
Results: In the ASD-D group there was no significant difference in both RA and RV deformation properties when compared with control subjects. In the ASD-S group the peak systolic strain and SR values were significantly reduced in RA and RV when compared with control and ASD-D groups.
Conclusion: Strain and SR imaging provide clinically acceptable a deep inspection on regional changes in atrial and ventricular function for patients with ASD. Our results showed that right atrial and ventricular regional performance assessed by 2D strain analysis is reduced after surgical closure, but not after transcatheter atrial septal defect closure.In contrast to surgery, transcatheter closure of atrial septal defect preserves atrial and right ventricular function.

P2-6

Evaluation of Cardiac Functions in Brucellosis Patients without Overt Cardiac Involvement

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Objectives: Brucellosis is an important systemic infectious disease, especially in developing countries. Every organ and system of the human body can be affected in brucellosis. Cardiovascular complications of brucellosis are rare. This study aimed to assess cardiac functions in acute brucellosis patients without endocarditis.
Methods: This cross-sectional study enrolled 67 children with brucellosis and 40 healthy children. We performed detailed echocardiography in individuals with in brucellosis patients without overt cardiac involvement. Diagnosis was established by the Rose-Bengal test, positive Brucella standart tube agglutination test, and Coombs STA and/or isolation Brucella species from blood.
Results: Both groups were similar in terms of age, sex, and body mass index. Echocardiography revealed no difference among the two groups regarding ejection fraction, mitral and tricuspid annular plane systolic excursion, Pulsed-wave Doppler derived E/A ratios in mitral and tricuspid valves. Deceleration time of early mitral inflow was prolonged in patients with brucellosis. Mitral and tricuspid annulus Ea velocity were significantly lower in children with brucellosis. Ea, Aa, and Ea/Aa ratios in the interventricular septum, left ventricle (LV) posterior wall, and right ventricle (RV) free wall were lower in patients with brucellosis than in the control group. The E/Ea ratio was greater in patients with brucellosis than in the control group. Isovolumic relaxation time and RV and LV myocardial performance indices (MPIs) were greater in patients with brucellosis. There was also significant correlation between the inflammmatory parameters and MPIs.
Conclusion: This study showed the diastolic dysfunction in patients with acute brucellosis patients without overt cardiac involvement. In addition, we detected increased LV and RV MPI.

P2-7

Evaluation of treatment outcomes in patients with acute viral myocarditis by speckle tracking and tissue Doppler methods

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Objectives: The aim of this study was to assess the myocardial deformation and function by speckle tracking echocardiography (STE) and tissue Doppler imaging (TDI) in patients with acute viral myocarditis.
Methods: Seven patients (mean age 12 years, 6 male) diagnosed acute viral myocarditis and ten healthy children (mean age 11.9 years, 9 male) were studied prospectively. The STE and TDI were performed in patients before and after IVIG treatment. The left ventricular longitudinal global strain (LVLGS) and strain rate (LVLGSR), left ventricular circumferential global strain (LVCGS) and strain rate (LVCGSR) and right ventricular longitudinal global strain (RVLGS) and strain rate (RVLGSR) were examined by STE. The myocardial velocities (Sm, Em and Am) and time intevals [isovolumic contraction time (ICT), isovolumic relaxation time (IRT) and ejection time (ET)] at interventricular septum (IVS), left ventricular posterior wall (LVPW) and right ventricular lateral wall (RVLW) were examined by TDI.
Results: Sm (5.2 vs. 8.5 cm/s) and Em (11.1 vs. 14.6 cm/s) at IVS, Sm (4.7 vs. 8.2 cm/s) and Em (11.2 vs 15.8 cm/s) at LV, ET (223.4 vs. 261.7 ms) at IVS and ET (220.5 vs. 267.7 ms) at RV were significantly lower in patients before treatment than controls ($p < 0.05$). LVLGS (-18.4 vs.-23.3%), LVLGSR (0.17 vs. 0.83 s-1), LVCGS (-15.6 vs.-27.5%) and LVCGSR (0.3 vs. 1 s-1) were significantly decreased in patients before treatment than controls ($p < 0.05$ for LVLGS and LVLGSR, $p = 0.001$ for LVCGS and LVCGSR). There were significant improvements for LVCGS ($p = 0.001$) and LVCGSR ($p = 0.001$) in patients after treatment. Sm (5.2 vs. 6.2 cm/s) at IVS, LVCGS (-15.6 vs.-21.9%) and LVCGSR (0.3 vs. 0.6 s-1) were significantly lower in patients before treatment than in patients after treatment ($p < 0.05$). Inspite of improvements, Sm (6.2 vs. 8.5 cm/s) and ET (226.7 vs. 261.7 ms) at IVS, LVCGS (-21.8 vs.-27.5%) and LVCGSR (0.6 vs. 1 s-1) were significantly lower in patients after treatment than controls ($p < 0.05$).
Conclusions: The STE and TDI were useful methods for the evaluation of treatment outcomes in patients with acute viral myocarditis. The LVCGS and LVCGSR especially adds important information supporting both clinical and laboratory improvements.

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P2-8

Vendor Independent Evaluation of Three Dimensional Echocardiography of Right Ventricular Size and Function in Children

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Introduction: Quantification of right ventricular (RV) function has been difficult with 2-dimensional echocardiography; however, three-dimensional echocardiography (3DE) has emerged as a non-invasive tool to quantitate RV function and has been validated with cardiac magnetic resonance imaging. 4D-RV function 2 is a TomTec software that is vendor neutral and can generate RV size and functional parameters. This software has not been validated between different vendors in children. We sought to use this software on different vendors and to evaluate whether or not there are differences between vendors using this software in normal pediatric patients.
Methods: Thirteen children (median age 9yrs (5-14 yrs)) underwent 3DE evaluation using Philips EPIQ machine (X7 and X5 probes), GE E9 machine (4 V probe), and Siemen's machine (4 V probe) on the same day. 3D dataset were acquired and transferred onto the TomTec 4D-RV function 2 software to generate RV size and functional parameters. Analysis of variance was performed on these echocardiographic parameters.
Results: There were statistically significant differences found in the tricuspid annular plane systolic excursion (TAPSE) between Philips and the two other vendors (GE and Siemens) respectively. There were no statistically significant differences between vendors in 3DE parameters, RV strain, and fractional area change (FAC).

Table 1. RV size and functional parameters

	Philips	Siemens	GE	p-value
3D EDV	63.28 ± 41.11	62.75 ± 41.51	62.27 ± 41.22	0.6600
3D ESV	30.84 ± 20.81	30.50 ± 21.12	30.29 ± 21.29	0.7212
3D SV	32.47 ± 20.43	32.27 ± 20.53	31.98 ± 19.99	0.7451
3D EF	51.54 ± 2.85	51.81 ± 3.59	51.81 ± 2.32	0.5704
RVLS septum	19.55 ± 3.27	19.10 ± 3.58	18.10 ± 3.08	0.0624
RVLS free wall	26.51 ± 4.29	26.44 ± 2.87	26.22 ± 2.10	0.9366
TAPSE	16.14 ± 2.29	15.23 ± 1.74	14.86 ± 1.46	**0.0017†,‡**
FAC	45.45 ± 6.30	46.21 ± 4.99	45.35 ± 2.79	0.7205

†Philips is significantly different from GE
‡Philips is significantly different from Siemens

Conclusion: RV size and function using the vendor neutral software in children can be generated from three different vendors. Although statistically significant differences were found in TAPSE between vendors, the differences are clinically not significant and can be explained by the low resolution from the big probe. This study highlights the need to develop pediatric 3D probe from different vendors and TomTec 4D-RV function 2 can be used in the quantification of right ventricle in children.

P2-9

Correlation between Basic Echocardiogram and Cardiac Magnetic Resonance Parameter for the Right Ventricle Evaluation 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 and LV dimensions were measured in long axis and short axis during diastole and indexed by body surface area. The RV and LV echocardiographic measurements and the RV/LV ratio were 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, RV long- axis length index, and RV/LV long axis length ratio (RV/LV ratio) were correlated with the RV end diastolic volume index (RVEDVi) obtained by CMR. The RV/LV ratio is more correlated with the RVEDVi ($r = 0.71$, $p < 0.001$) in comparison with the RVEDD and RV long axis($r = 0.47$, $p = 0.01$ and $r = 0.27$, $p = 0.01$).
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 RV/LV has a better prediction of the RV volume in comparison with the RVEDD and RV long-axis length index.

P2-10

Real-Time 3D-Echocardiography of the Pediatric Left Ventricle - influence of different quantification software

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Background: Determining left ventricular (LV) 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 LV assessment, however there is no systematic study comparing the inherent influence of different evaluation software on the resulting measurements.
Methods: 497 healthy children and adolescents (range, 1 day-216 months) underwent RT3DE imaging of the LV (IE33, Philips, Andover, USA). 370/497 (74.4%) 3D data sets could be quantified using two different semiautomatic border detection software (Qlab9.0, Philips and TomTec LV2.7, Unterschleissheim, Germany). Using TomTec, the influence of changes of contour sensitivity was tested by 75 as well as 30 intensity units ($TomTec_{75}$ vs. $TomTec_{30}$).
Results: Analysis of identical 3D-data with the same software (TomTec) but different automatic contour finding sensitivity had a significant impact. Use of higher ($TomTec_{75}$) instead of lower ($Tom\ Tec_{30}$) sensitivity resulted in significant smaller enddiastolic, endsystolic and stroke volumes (EDV, ESV, SV) (Fig1a, Table1; $p < 0.001$). Comparing QLab to $TomTec_{30}$, Bland-Altman analysis showed moderate bias with slightly smaller EDV using QLab ($p = 0.07$) and larger ESV ($p = 0.05$; Table2). Comparing QLab to $TomTec_{70,}$ using a higher contour sensitivity, the bias between both software systems was significant higher ($p < 0.001$ for all parameters, Fig. 1b).

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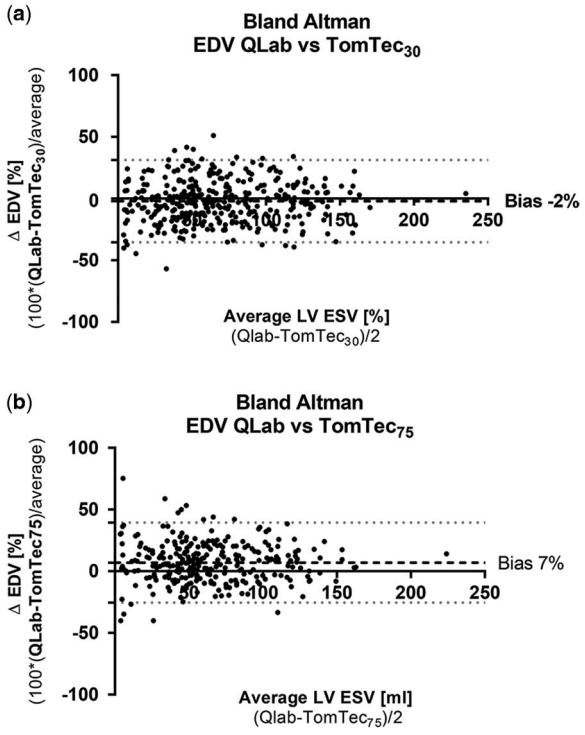
Intra- and interobserver-variation for EDV, ESV and SV were excellent both in Qlab and TomTec with intraclass correlation coefficients (ICC) between 0.91–0.99.

Table 1. 3D Volume Analysis of the Left Ventricle in Children

Agreement between high vs low contour detection sensitivity ($TomTec_{75}$ vs $TomTec_{30}$)				
Para-meter	**Bias (ml)**	**Bias (%)**	**SD (%)**	**Limit of agreement (ml)**
ESV	-1.9	8.1	14.4	-8.3 to 4.5
EDV	-5.4	7.0	10,5	-17.3 to 6.6
SV	-3.7	6.7	5.5	-14.5 to 7.1
EF	0.1%		4.3	-8.2 to 8.4%

Table 2. RT3D Volume Analysis of the Left Ventricle in Children

Agreement between QLab and $TomTec_{30}$				
Para-meter	**Bias (ml)**	**Bias (%)**	**SD (%)**	**Limit of agreement (ml)**
ESV	0.7	-0.8	6,5	-12 to 13,32
EDV	-1,2	-2.2	12,78	-26,3 to 23.8
SV	-2.0	-4.2	9,8	-21.2 to 17,2
EF	-1%		6,6	-14.8-11.7%



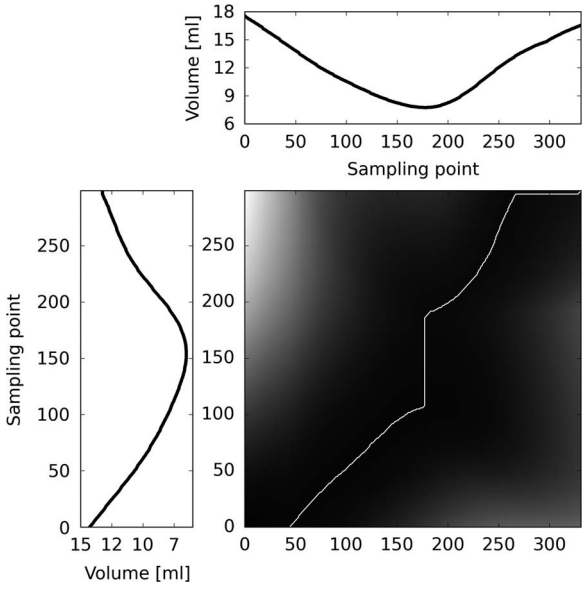
Conclusions: 3D-volumetric assessment of the LV is possible using different algorithms. However, resulting measurements differ depending on the software itself as well as different algorithms within the software. Therefore a set of reference values for the pediatric heart is urgently required. General agreements on analysis would be needed to overcome interobserver as well as interstudy variability.

P2-11

Statistical Analysis of Volume Time Curves for Multiple Imaging Methods

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Introduction: Real-Time-3D-Echocardiography (RT3DE) offers the opportunity to determine cardiac volumes non-invasively and display their behaviour during the cardiac cycle. Volume time curves (VTC) express volume trends over time and comprise important information about systolic and diastolic function. However, data interpretation and statistical comparisons of VTCs derived from different devices such as cardiac magnetic resonance imaging (CMR), RT3DE or conductance technology and comparison between different software tools is complex. Standard statistical methods have not been developed so far. Objective of our study is to develop a mathematical approach to analyse, characterise and compare VTC.
Methods: First, we want to apply pattern recognition and statistical algorithms to compare different VTC. We use a cluster analysis to detect similar VTC and group them into clusters. Subsequently, they are compared using the Euclidean distance and the dynamic time warping (DTW) algorithm. Each of the resulting clusters will be analysed with respect to the patients' data, such as heart size and anatomy. Additionally, a discrete Fourier transform decomposes the VTC into complex sinusoids, which are resembled by a list of coefficients. These coefficients are features of the VTC, which are easy to compare quantitatively. For the investigation of the coefficients, classification and filtering algorithms can be applied to group VTC regarding their frequencies.
Results: A first clustering with a set of 16 different VTC derived from RT3DE has been established. Furthermore, the DTW algorithm has been used to detect the optimal non-linear alignment between two VTC and calculates a cost matrix and warp path, which are shown in the figure.
Conclusions: This approach for data analysis can be used to recognize patterns and features of VTC. Characterization of systolic and diastolic functionality can be used to described and assess pathologies or dysfunctions of the heart. Furthermore, it could allow to compare different measurements from CRT, RT3DE or conductance technology to evaluate the quality of each device. In terms of 'big data' it paves the way for modelling of heart failures based on non-invasive methods and provides a fast data analysis.



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P2-12

Assessment of cardiac function in children with type 1 Diabetes Mellitus

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Introduction: Impairment of cardiac function in patients with type1 Diabetes (T1DM) represents one of the serious complications that may affect the quality of life and prognosis of the disease.
Objective: To evaluate the cardiac function in children with T1DM by conventional and tissue Doppler echocardiography.
Patients and methods: A prospective study included 40 T1DM patients (age between 6 and 16 years) with more than 5 years duration and 20 age and sex matched healthy children as controls. The patients were subjected to detailed history taking, thorough clinical examination and laboratory investigations including Glycosylated hemoglobin A1C, serum lipids & lipoproteins (serum total cholesterol, triglycerides, LDL cholesterol and HDL cholesterol). Conventional and tissue Doppler echocardiography were done to both patients and controls.
Results: The study included 16 females and 24 males with mean age ± SD of 12.1 ± 2.39 years and mean duration of diabetes 6.63 ± 1.89 years. Patients had larger dimensions of the aorta (AO), left ventricular end diastolic dimension (LVIDd) and left ventricular end systolic diameter (LVIDs) (P-value 0.047, 0.009 and 0.001 respectively). The early diastolic filling velocity of the tricuspid valve (E wave), late diastolic filling velocity (A wave) and E wave velocity of the mitral valve were found significantly lower in diabetic patients than controls (P-value of 0.023, 0.006 and 0.019 respectively). Diabetic patients had significantly longer Isovolumic Relaxation time (IRT) (P-value of 0.001).
Five patients had right ventricular diastolic dysfunction and another 5 patients had left ventricular diastolic dysfunction. Only one patient had both right and left diastolic dysfynction. There were 28 patients with poor glycemic control but no significant differences between them and those with good glycemic control as regards the echocardiographic data. No significant relationship existed between the duration of DM or HbA1c and the echocardiographic parameters.
Conclusion: Diabetic children have evidence of diastolic dysfunctions. This highlights the importance of periodic cardiac evaluation with both conventional and tissue Doppler echocardiography for early detection of this dysfunction.

P2-13

Cardiac ultrasound findings in children with mucopolysaccharidosis

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Introduction: Cardiac involvement in mucopolysaccharidosis (MPS) is variable, consisting in severe cardiac valve disease and ventricular hypertrophy and has a major contribution into the morbidity and mortality of these patients. The aim of the study was to characterise the echocardiographic abnormalities in children with different types of mucopolysaccharidosis and their evolution after 12 months of enzyme replacement therapy.
Methods: We evaluated the function of the mitral and aortic valves, left ventricular chamber dimensions, septal and posterior wall thicknesses and ventricular function in 20 patients (5 patients with MPS type I, 14 patients with MPS type II and one patient with MPS type IV), aged 1-16 years.
Results: At diagnosis, all patients presented echocardiographic alterations. Mitral valve thickening with variable grades of regurgitation was diagnosed in all patients; mitral stenosis was present in 10% of patients. Aortic regurgitation was present in 68% of patients and aortic stenosis in 5%. Left ventricular hypertrophy was diagnosed in 40% of patients and there was no systolic dysfunction. Mild pulmonary hypertension was present in 26% of the patients. Thirteen patients (3 patients with MPS type I and 10 patients with MPS type II) underwent enzyme replacement therapy. After 12 months of treatment we obtained stabilization of cardiac valvular disease in 69% of patients, mild improvement in 8% and worsening of disease in 23% of patients.
Conclusions: Left valve lesions, ventricular hypertrophy, and pulmonary hypertension are the most common findings in children with mucopolysaccharidosis. Enzyme replacement therapy had little effect on cardiac valve disease.

P2-14

Usefulness of myocardial strain imaging in Spinal Muscular Atrophy

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Background: In paediatric patients with Spinal Muscular Atrophy (SMA), cardiac systolic function is generally described to be within the normal range. Some studies have suggested the presence of subclinical dysfunction in congenital muscular diseases using tissue Doppler measurements and myocardial velocity gradients. This has not been described for SMA. The aim of this study was to further assess regional myocardial function in paediatric patients with SMA using myocardial velocity and deformation imaging.
Methods: Thirty-one patients with SMA (mean age, 7.2 years; range, 0-12 years) and 29 age-matched normal controls were studied with echocardiography. Standard echocardiographic measurements of left ventricular (LV) systolic and diastolic function were performed. Myocardial velocity and deformation data, including peak systolic and early and late diastolic myocardial velocities, peak systolic strain rate (SR), and peak systolic strain, were calculated in the radial direction in the inferolateral LV wall and in the longitudinal direction in the interventricular septum, the LV anterolateral wall, and the right ventricular (RV) free wall.
Results: Higher heart rates and increased LV end-systolic dimensions were seen in patients with SMA compared with controls. Significant decreases in radial and longitudinal peak systolic SR, peak systolic strain, and peak systolic and early diastolic myocardial velocities were found in the LV inferolateral and anterolateral walls in patients with SMA. No significant differences in longitudinal function could be found in the interventricular septum or in the RV free wall.
Conclusion: In young patients with SMA who have global normal systolic function, reductions in systolic deformation parameters as well as reduced early diastolic myocardial velocities can be detected in the anterolateral and inferolateral LV walls. The prognostic significance of these findings warrants further longitudinal follow-up.

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P2-15

Current qualification criteria for echocardiography examination by a pediatric cardiologist in neonatal screening yields 19% pre-test probability of significant congenital heart disease

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Neonatal echocardiographic screening remains an open debate. Clinical feasibility is obvious but cost-effectiveness remains of a concern. Proper qualification based on reasonable indications, and rational rules of follow-up avoiding both missed diagnoses and unnecessary costs are to be aimed at. This summarizes 20 months of neonatal screening in tertiary university center starting January 2014. The studied population encompassed 4758 life born newborns.

There were 1467 studies carried out in, and 1152 patients were examined. The mean birth weight was 2979+ /-963g, range 600-5220 g. 720 studies were ordered before the lapse of the second day and carried out before the third day of life. The most common indications for study were prematurity 27%, murmur 20%, positive pulsoximetry test 14%, prenatal diagnosis of congenital heart disease 13%, diabetes mellitus of the mater 8%, cyanosis/desaturation 5%. Other included respiratory distress, non-cardiac congenital and rhythm disturbances.61% newborns were assessed to be normal or to have physiological findings. 38% had PFO, 3% had ASDII and in 8% the defect had in-between size. PDA was present in 25%cases but only 5% were significant. VSD was found in 98 (9%). CoA was suspected or mild in 12 (1%) cases and was critical in 9 (1%) cases, 5 of which with hipoplasia of the arch. There were 3 cases of balanced, complete AVSD and 1 partial. There were 6 (0,5%) pulmonary stenosis (3 significant), 13 (12%) cases of bicuspid AV, 13 mild aortic insufficiencies,1 aortic stenosis, 18 moderate and 3 severe TR. There were 30 (2,6%) cases of complex defects: 6 ToF, 2 TAC I, 6 TGA, 4 HLHS, 2 PA, and single cases of Taussig-Bing, DORV, IAA-B, IAA-C, R-ISO, TA, PAPVR, TAPVR, Ebstein anomaly. There were 24 cases of ventricular hypertrophy related to diabetic fetopathy.

The total of significant congenital heart diseases was 225 and this yields decent 19% pre-test probability. On the other hand, a very early qualification for the echo study and normal or physiological findings in the vast majority of cases calls for further analysis whether the current qualification criteria can by fine-tuned to increase the performance of the whole screening without risking missed diagnoses.

P2-16

Increased aortic stiffness in prepubertal girls with Turner syndrome

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Introduction: Aortic dilation and dissection contribute highly to the increased mortality of Turner syndrome (TS) but the exact pathophysiology is not completely understood.

Methods: We investigated aortic diameters and aortic wall properties in young prepubertal TS girls. Fifteen prepubertal TS girls (median age 10.64, IQ 8.31-11.04) with a tricuspid (TAV n = 9) or a bicuspid (BAV n = 6) aortic valve, and 31 sex, age and height matched healthy controls underwent a cardiac and vascular ultrasound to evaluate aortic dimensions and elastic properties of the aortic wall.

Results: TS BAV had significantly larger ascending aortic diameters than controls for absolute diameter 22.2 ± 5.10mm vs 18.7 ± 1.92mm ($p = 0.017$) and z-score 1.8 ± 2.05 vs 0.2 ± 0.73 ($p = 0.004$). Distensibility of the ascending aorta was lower in the TS than in controls (40.2 10-3kPa-1, IQ 31.3-56.2 vs 62.9 10-3kPa-1, IQ 55.5-76.5, $p = 0.002$) both for TS TAV ($p = 0.012$) and BAV ($p = 0.003$).Stiffness index was higher in TS than in controls (5.26, IQ 3.34-5.26 vs 3.23, IQ 2.55-3.24, $p = 0.004$) both for TS TAV ($p = 0.027$) and TS BAV ($p = 0.004$). Pulse wave velocity along the whole aortic arch was not different between groups. There was no correlation between stiffness and z-score of the ascending aortic diameter.

Conclusions: We concluded that in prepubertal TS girls stiffness of the ascending aorta is increased in patients with a BAV and TAV while dilation of the ascending aorta is more frequent in BAV. This suggest an intrinsic aortic wall abnormality making all TS patients at increased risk for severe aortic complications although the risk is the highest for TS with BAV.

P2-17

Right atrial enlargement in Children with Atrial Septal Defect or Pulmonary Hypertension with Congenital Heart Disease: comparison to normative values

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Objectives: Right atrial (RA) size may become a very useful, easily obtainable, echocardiographic variable in patients with congenital heart disease (CHD) with right heart dysfunction, however according studies in children are lacking. We investigated growth related changes of RA dimensions in healthy children. Moreover, we determined the predictive value of RA variables in both children with secundum atrial septal defect (ASD) and children with pulmonary hypertension (PH) secondary to CHD (PH-CHD).

Methods: Prospective study in 516 healthy children, in 80 children with a secundum ASD (> 7 mm superior-inferior dimension), and in 42 children with PH-CHD. We determined three RA variables: end-systolic major-axis length, end-systolic minor-axis length, and end-systolic area, stratified by age, body weight (BW), length (BL), and surface area (BSA).

Results: RA end-systolic length and area z-scores were increased in children with ASD and PH-CHD when compared to those variables in the healthy control population. Using the Youden Index to determine the best cut-off scores in sex and age-specific RA dimensions we observed a sensitivity and specificity up to 94%, and 91%, respectively, in ASD children and 98%, and 94%, respectively, in PH-CHD children.

Conclusions: Normal values for RA size, and area in a representative, large pediatric cohort are provided. Enlarged RA variables with scores > +2 were predictive of secundum ASD and PH-CHD. Echocardiographic determination of RA size can identify enlarged RAs in the setting of high volume load (ASD) or pressure load (PH-CHD).

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P2-18

High sensitivity Troponin T in Pediatric patients with Congenital Heart Disease

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Background: High sensitivity troponin T (hsTnT) detects the myocardial injury and predicts poor outcomes in adults with acquired and congenital heart disease (CHD). However, few reports describe the usefulness of hsTnT in pediatric patients (pts) with CHD.
Objective: We sought to determine whether hemodynamic load and hypoxia induce the myocardial injury and the hsTnT as a merker of myocardial injury pedict the adverse events such as the cardiac arrest, death, or lethal arrythymiay in pediatric pts with CHD.
Methods: We assessed the hsTnT levels in 86 pts with CHD who underwent cardiac catheterization. We analyzed the relation between the levels of hsTnT and hemodynamic variables and assess the adverse events during follow-up.
Results: Age of pts was 2.8 ± 4.2 yrs. The hsTnT levels were significantly higher in caynotic CHD (median 0.0161ng/ml) than in non caynotic CHD (median 0.004ng/ml). Mutiple regression analysis showed that hsTnT levels correlated with the ratio of right to left ventricular pressure (RVP/LVP) ($r = 0.73$, $P < 0.001$) and systmic aortic saturation (SaO2) ($r = 0.69$, $p < 0.001$), but not the ratio of pulmonary to systemic fow and pressure (Qp/Qs and Pp/Ps). Median follow up period after the measurement of hsTnT was 2.0 yras. Four pts occurred adversed events during the follow up period. Pts wih elevated hsTnT levels (>0.015) occurred adverse events more frequently than in pts with normal hsTnT (75% vs 26%, $p < 0.05$).
Conclusion: Pressure overload to right ventricle and hypoxia induce the myocardial injury. The high levels of hsTnT may predict poor outcomes in pediatritrc patients with CHD.

P2-19

Does Tissue Motion Annular Displacement of the Tricuspid Valve measuring by two dimensional Speckle Tracking Echocardiography Predict the Right Ventricular Ejection Fraction?

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Background: Now magnetic resonance imaging (MRI) has become new gold standard for determining the right ventricular ejection fraction. A few of previous studies showed that mitral valve tissue motion annular displacement (TMAD) measured by two dimensional speckle tracking echocardiography (2DSTE) is a simple, effective, and highly reproducible method of assessing the left ventricular ejection fraction in normal children. We performed TMAD for tricuspid valve in patients with repaired Tetralogy of Fallot (TOF) patients and healthy peers. We try to determine any correlation between cardiac MRI (cMRI) derived right ventricular ejection fraction and tricuspid valve TMAD measurements.
Method: The study consisted of 20 patients with repaired TOF and 22 age and body surface area matched healthy peers. Tissue motion annular displacement was measured by 2DSTE. Septal, lateral and midpoint displacement of the tricuspid valve was measured. Right ventricular ejection fraction was determined by cMRI in 12 of study patients. Correlation analysis was performed to evaluate the relationship between cMRI derived right ventricular ejection fraction and percentage of TMAD measurements in patients with repaired TOF.
Results: The mean ejection fraction derived from cMRI was $43.6 \pm 9.11\%$ in 12 of study patients. We found a negative correlation between tricuspid TMAD midpoint measurements and cMRI-derived ejection fraction ($r = -0.68$, $p = 0.016$). We found significantly difference between study and control groups in terms of septal (10.92 ± 4.57 and 8.07 ± 4.02, respectively) ($p = 0.039$), lateral (7.40 ± 4.14 and 12.24 ± 3.31 respectively) ($p < 0.01$) and percentage of total change ($11.01 \pm 5.13\%$ and $14.09 \pm 4.19\%$, respectively) ($p = 0.041$) values.
Conclusion: Tissue motion annular displacement of the tricuspid valve is a simple, effective, and highly reproducible method of assessing the ejection fraction of right ventricle in patients with TOF. It has an highly strong negative correlation with cMRI-derived ejection fraction.

P2-20

Effect of oral high-dose cholecalciferol on cardiac mechanics in children with chronic kidney disease

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Background: As cardiovascular factors are the leading cause of mortality in chronic kidney disease (CKD) and as vitamin D deficiency is prevalent in this population, we aimed to examine the effect of oral cholecalciferol on cardiac mechanics in children with CKD.
Methods: Forty-one children with CKD and 24 healthy subjects free of any underlying cardiac or renal disease with low 25-hydroxyvitamin D3 (25OHD) levels were evaluated by conventional, tissue-Doppler imaging (TDI) and 2D speckle tracking echocardiographic (2D STE) methods basally and following Stoss vitamin D supplementation. Left ventricular strain and strain rate values were compared among the groups.
Results: Initial longitudinal and radial strain and strain rate values of the left ventricle were significantly lower in patients. After vitamin D supplementation, these improved significantly in patients, while no significant change was observed for the healthy group.
Conclusions: Our interventional study showed that while conventional and TDI methods could not determine any affect, 2D STE revealed the favorable effects of high-dose cholecalciferol on cardiac mechanics, implying the importance of vitamin D supplementation in children with CKD.

P2-21

Left ventricular mechanics are affected in children with celiac disease - a study of 2D speckle tracking echocardiography

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Background: Cardiomyopathy associated with celiac disease is reported, though not frequently. We aimed to examine the effect

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on cardiac mechanics in children with celiac disease (CD) by 2D speckle tracking echocardiography (2DSTE).
Methods: Eighty-one children with CD were compared with a control group of 51 healthy children by an echocardiographic examination. Children with CD were divided into two different groups: Group 1-positive serum anti-tissue transglutaminase antibody (n = 48), and Group 2-negative serum anti-tissue transglutaminase antibody (n = 33). Cardiac functions of all the children were evaluated by conventional, tissue-Doppler imaging (TDI) and 2DSTE methods.
Results: The mean ages and male/female ratio of children with CD were 10.1 ± 4.0 years and 26/55 (67% female) and were not different from controls. Patients were diagnosed at a mean age of 7.9 ± 4.1 years and mean follow-up time was 2.37 ± 2.98 years. Conventional echocardiography and tissue Doppler measurements did not differ between groups. The left ventricular longitudinal and radial strain and strain rate values were significantly lower in children with positive serum anti-tissue transglutaminase antibody (group 1) when compared control group.
Conclusions: Our results suggest that left ventricle longitudinal and radial myocardial deformation is decreased in children with coeliac disease who have positive serum anti-tissue transglutaminase antibody. Further follow-up is necessary to precisely determine the clinical significance of these myocardial changes detected by 2DSTE in children with CD.

P2-22
Clinical accuracy of annual cardiac computed tomography angiography in pediatric heart transplant recipients
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Introduction: Cardiac allograft vasculopathy (CAV) is a major cause of late heart graft failure. Because the transplanted heart often remains denervated, the first symptom of myocardial ischemia can be sudden death and routine periodic screening is thus recommended. Cardiac computed tomography angiography (CCTA) demonstrated a good sensibility and negative predictive value compared to coronary angiography in this indication, and is used for annual screening in our institution since September 2003.
The aim of this study was to evaluate clinical accuracy of CCTA annual screening in pediatric heart transplant recipients.
Method & Results: From September 2003 to November 2015, 175 CCTA were performed annually in 36 patients transplanted < 18 years between 1988 and 2014. Mean age at transplantation was 10+ /- 3.6 years; ischemic time was 208+ /- 44.3 minutes; 9 patients had CMV mismatch. All had immunosuppressive tritherapy. Mean follow-up was 11.8+ /- 6.8 years. Age at first CCTA was 14.9+ /- 4.2 years; volume of contrast agent was 1.4+ /- 0.4 ml/kg; product length dose was 179.1+ /- 109.4 mGy.cm. All CCTA were interpretable but 11 with uncertainties: poor visibility of distal coronary network in 7, artifacts in 4 (respiratory in 3, implantable cardiac defibrillator electrode in 1). During follow-up, 5 patients died, none had retransplantation, 8 had at least 1 episode of graft rejection, and 10 had chronic renal insufficiency due to anticalcineurin toxicity, with renal transplantation in 3.
No lesion was found on iterative CCTA in 32 patients (89%). Among them, none had coronary-related death, nor acute coronary syndrome. Coronary lesions occurred in 4 patients 2.9, 14.5, 22.6 and 24.7 years after cardiac transplantation. Coronary selective angiography confirmed CCTA lesions in 3; the fourth patient refused angiography. None needed coronary angioplasty; one died of acute graft rejection.
Conclusion: CAV is very uncommon in our cohort of pediatric heart transplant patients. No coronary-related events occurred in patients with negative CCTA. There was no false positive diagnosis although CCTA tends to overestimate the severity of lesions. Because of the low incidence of CAV in pediatric patients, we suggest that CCTA screening might be spaced out every 2 years.

P2-23
Pulmonary perfusion in patients with HLHS after Fontan operation and pulmonary artery stenting
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Introduction: Left pulmonary artery (LPA) stenosis is a common complication during multistage palliation of patients with hypoplastic left heart syndrome (HLHS).
Fontan operation results in diminished pulsatility of pulmonary blood flow which affects blood distribution in the lungs.
Aim: Lung perfusion assessment in patients with HLHS after Fontan operation using planar lung perfusion scintigraphy.
Material and Methods: 73 patients (52 boys) at a median age of 7,5 years (3,3-19,5 years) took part in the study. 57 patients (78%) had LPA stenting. All patients underwent planar lung perfusion scintigraphy. Two injections of 99mTc – MAA into the right upper and then into the right lower extremity were performed. Webster's formula was used to calculate the activity and Hawkeye hybrid gamma camera to perform lung perfusion studies, processed on Xeleris workstation. Percentage share of every lobe and lung in a global lung perfusion was calculated after averaging of counts in anterior and posterior projection for inflow from superior and then from inferior cavopulmonary anastomosis.
Results: Mean left and right lung contribution to global lung perfusion was: 38% (upper lobe 15%; lower lobe 23%) and 62% (upper lobe 8%; middle lobe 29%, lower lobe 25%) respectively. Severe left lung hypoperfusion, defined as percentage ≤ 35% of global lung perfusion, was noted in 30 patients (41%). In two patients without prior LPA stenting we found severe left lung hypoperfusion – 12% and 9% respectively. Both patients were asymptomatic. We diagnosed severe LPA hypoplasia, first patient had also left diaphragmatic paralysis. After successful LPA stenting, left lung perfusion increased up to 24% and 30% respectively. Postoperative diaphragmatic paralysis causing decrease of one of the lungs was noted in 8 patients (11%). Radionuclide activity in kidneys and brain suggested pulmonary arterio-venous fistulas (PAVF) in 5 patients (7%).
Conclusions: 1.Patients with HLHS after Fontan operation have pulmonary perfusion abnormalities due to: non-physiological pulmonary blood flow, pulmonary artery obstruction and postoperative diaphragmatic paralysis.
2.Planar lung perfusion scintigraphy is an useful method in postoperative evaluation of patients after Fontan completion.
3.Hypoperfusion of the left lung is the most common finding in spite of successful LPA stenting.

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P2-24

Is the lateral tethering angle associated with the severity of tricuspid valve regurgitation in hypoplastic left heart syndrome? A 3D echo and MRI study

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Objectives: The degree of tricuspid valve regurgitation (TR) is a significant prognostic factor for patients with hypoplastic left heart syndrome (HLHS) undergoing single-ventricle palliation. The lateral tethering angle (LTA) of the anterior papillary muscle (APM) of the tricuspid valve (TV) is the angle between the TV annulus and the APM and has been previously correlated with the severity of TR. However these studies have depended on subjective echo assessment of TR and RV function. We present data in respect of the relationship between LTA, MRI derived RV ejection fraction (RVEF), TR regurgitant fraction (TRRF) and indexed RV end-diastolic volume (iRVEDV).
Subjects and Methods: Patients with HLHS at different stages of single ventricle palliation. Echocardiography: LTA was measured using Tomtec Echoview software on transthoracic 3D datasets obtained using the Philips iE33 ultrasound system. Indexed RVEDV, TRRF and RVEF were recorded from MRI. MRI imaging was performed within 4 weeks of the echo in 26/31 cases. 31 paired MRI/echo studies from 27 patients (aged 35 days to 13 years) were analysed. 18 studies were prior to Hemifontan, 12 prior to Fontan, 1 post Fontan. Based on data from our own institution the patients were separated into groups depending on the surgical stage, TRRF (trivial/mild: RF < 20%, moderate/severe: RF ≥ 20%), RVEF (good ≥ 50%, impaired < 50%), iRVEDV (pre Hemifontan normal ≤ 133.6 ml/m2, pre Fontan normal ≤ 109.6 ml/m2).
Results: The LTA for the different groups was: 1) Pre Hemifontan (n = 18): 93.5° ± 10.1°, pre Fontan (n = 12): 93.7° ± 8.96° (p = 0.949). 2) TRRF < 20% (n = 21): 93.8° ± 9.6°, TRRF ≥ 20% (n = 10): 91.2° ± 10.8° (p = 0.502). 3) RVEF ≥ 50% (n = 25): 93.5° ± 10.7°, RVEF < 50% (n = 6): 90.9° ± 5.3° (p = 0.572). 4) Normal iRVEDV (n = 26): 93.7° ± 9.9°, enlarged iRVEDV (n = 5): 89.4 ° ± 9.9° (p = 0.378).
Conclusions: Our study did not show any statistically significant difference in the LTA for different surgical stages, severity of TR, MRI-derived RVEF or iRVEDV. It can be used as part of the assessment of the TV but should be interpreted in the context of the whole echocardiographic and MRI findings. Further studies and larger number of patients are required to define this technique further.

P2-25

Diffuse fibrosis in the ventricles of patients with transposition of great arteries late after atrial switch

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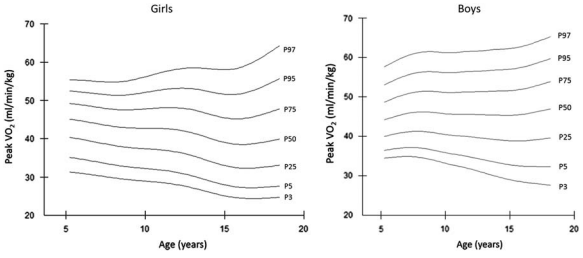
Objectives: In adult patients with transposition of great arteries late after atrial switch (Mustard or Senning), the subaortic morphologic right ventricle (RV) is hypertrophic, while the subpulmonary morphologic left ventricle (LV), is usually hypotrophic.
The extent of diffuse fibrosis in the RV and LV in these patients remains unclear. Therefore, the aim of this study was to determine myocardial extracellullar volume (ECV) in both ventricles in these patients.
Methods: We determined ECV by cardiac magnetic resonance (CMR) in 10 patients (36.8 ± 5.3 years old), without relevant pulmonary stenosis, late after atrial switch by acquiring T1-maps of the myocardium before and 10 minutes after injection of Gadolinium-based contrast agent. Furthermore, we obtained interobserver agreement of determining ECV.
Results: ECV of the LV was significantly increased compared to the RV (36 ± 4% vs. 27 ± 3%, p < 0.01).
Conclusions: In patients late after atrial switch, ECV of the hypotrophic, subpulmonary morphologic LV is significantly increased compared to ECV of the hypertrophic, subaortic morphologic RV. ECV of the RV is in the upper normal range. Increased ECV in the LV may be due to diffuse fibrosis, induced by long time reduced activity of LV, or simply due to relatively higher ECV surrounding hypotrophic myocardial cells of the LV.

P2-26

Contemporary reference values for peak oxygen uptake in healthy European children and adolescents

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Background: The aim of the present study was to establish prediction equations and to describe contemporary reference values for healthy boys and girls from West-Europe.
Design: Cross sectional multicenter study.
Methods: Between January 2000 and April 2015, 1580 healthy children (1010 boys; mean age 12.4 years; range 5-18) completed a maximal graded exercise test until volitional exhaustion either on a bicycle or on a treadmill. Minute ventilation, oxygen uptake (VO2), and carbon dioxide (CO2) production were measured on a breath-by-breath basis and continuous electrocardiography was performed. Peak VO2 was expressed per kg bodyweight. Prediction equations based on sex, age and weight were calculated by means of regression analysis and cross-validated through concordance correlation coefficient determination and Bland-Altman analyses. LMS chart maker was used to provide age-related percentile curves.
Results: Multivariate regression analysis revealed age, sex and weight as statistically significant determinants of peak VO2. Hence, following prediction equation was established and cross-validated: peak VO2 = 44.5-218*sex+ 27.1*age+ 37.4*weight (p ≤ 0.001; sex = 0 for boys and sex = 1 for girls). Finally, the distribution of peak VO2 per kg body weight with age was descripted by percentile curves (Fig. 1 and 2) and reference values were established for boys and girls separately.



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Conclusions: Our study is the largest and most recent population-based study to provide sex-specific reference data and a prediction equation for peak VO2 for a West-European population of healthy children between the ages of 6-18 years.

P2-27
New technologies for the study of left ventricular mechanics in pediatric heart transplantation recipients

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Introduction: Recently some studies dealing with assessment of cardiac function by Speckle Tracking Imaging (STI) in heart transplant (HTX) recipients have been published, an integrated assessment of all myocardial properties of deformation is lacking.
Methods: The aim of this study is to perform, by using Speckle Tracking Imaging, an integrated study of LV mechanics (longitudinal, radial, circumferential regional deformation, twist, untwist onset and diastolic function) in HTX recipients with preserved ejection fraction and good clinical conditions (NYHA I). We enrolled 20 HTX subjects without symptoms (NYHA I) and with preserved ejection fraction (HTX group), and 40 normal subjects as control group (CTRL group).
Results: HTX patients were characterized by an early diastolic dysfunction, detectable by an increased value of E/E' ratio (HTX 10,58+ /- 2,94; CTRL 5,3+ /- 1,5; $p < 0.0001$). Compared to control group our HTX recipients showed: a) impaired longitudinal and radial strain values with normal circumferenzial strain values b) LV rotation was preserved at apex and impaired at basal level (bas rot CTRL -7,35+ /- 1,6; HTX =-5.05+ /- 2,89 $p < 0.001$; ap rot CTRL 6.92°+ /- 2.56°; HTX = 9,73+ /- 4,38 $p < 0.0001$) with normal LV twist (CTRL 14,22+ /- 3.4; HTX = 14,78+ /- 5,58; p value not significant); c) delayed untwisting due to prolonged twist (time to peak twist/systolic time: CTRL 80,48 + /- 13,69, HTX 103,45+ /- 18,42 $p < 0.0001$).
Conclusions: Of interest 'healthy' HTX patients, even in presence of widespread impairment of regional myocardial deformation, normal apical rotation, which account for normal amplitude of twist and in turn global ejection fraction.
In addition delayed untwisting (after aortic valve closure) due to prolonged twist could be responsible of an early impaired LV diastolic filling.

P2-28
Cardiac MRI and 3D Echocardiography in acute rheumatic carditis

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Introduction: The purpose of this study was to evaluate the usefulness of 3D echocardiography and cardiac MRI, to assess the structural and functional changes in mitral valve apparatus during an episode of acute rheumatic fever.
Methods: Twenty two consecutive cases of acute rheumatic fever (mean age 12.15 ± 4.2 years) with carditis were enrolled in the study. Baseline investigations and a 3D echocardiographic study were done. The follow-up echocardiograms were repeated after 4 weeks of steroid therapy in 16 patients. Age-matched chronic rheumatic mitral regurgitation patients were included as controls. Seven successive patients with acute rheumatic carditis also underwent cardiovascular MRI with gadolinium enhancement.
Results: Twelve (55%) patients had evidence of rheumatic nodules on echocardiography, while none of the controls had rheumatic nodules. 3D echocardiography showed a diffuse nodular appearance of mitral leaflet surface during activity in contrast to the glistening appearance seen in chronic RHD. The thickness of leaflets at tip, mid part and base of anterior/posterior leaflets during acute activity was higher than the control. The thickness of the mid-part of anterior mitral leaflet was 5.1 ± 0.6 mm in the study group initially and 4.9 ± 0.96 mm on follow up, while the control group had a thickness of 3.87 ± 0.8 mm ($p = 0.008$).
On cardiac MRI, global increase in signal intensity in T2W imaging was found in 28.5% (2/7) of our patients of acute rheumatic fever. All 7 patients have shown late gadolinium enhancement (LGE) indicating the presence of myocardial damage. The LGE was seen in all the patients, in posterior wall of left atrium and mitral valve, involvement of tricuspid valve was seen in 57.1% (4/7 cases) while one patient showed LGE in LV endocardium.
Conclusions: 3D echocardiography complements 2D echocardiography in the evaluation of patients presenting with rheumatic fever. Nodular appearance and leaflet thickening are important echocardiograpic features of rheumatic carditis. Cardiac MRI shows some characteristic features in rheumatic carditis. Whether these finding are helpful in differentiating acute carditis from chronic RHD needs to be investigated further.

P2-29
Tissue Doppler and speckle tracking deformation measurements are not interchangeable in term neonates

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Introduction: Conventional echocardiographic indices often fail to detect impaired heart function in neonates. Strain and strain rate are new indices of heart function that describes deformation of the myocardium. It is possible to obtain these indices by colour tissue Doppler echocardiography (cTDE) and 2D-strain speckle tracking echocardiography (STE). cTDE enables high time resolution but gives false low values in poor-quality images. STE assess deformation within two dimensions. In this study, we compared left ventricle deformation indices by cTDE and STE in term neonates.
Methods: From apical four-chamber views, we assessed peak systolic strain (PSS), peak systolic strain rate (PSSR), early diastolic strain rate (ESR) and strain rate in atrial systole (ASR) by cTDE and by STE in the left ventricle at the same examination in 88 neonates the first day of life. We averaged measurements from three heart cycles and assessed indices by cTDE in one large segment from the septum and one large segment from the left wall and indices by STE (2D-strain) from one large region of interest covering the septum and the left lateral wall. We calculated left ventricle cTDE indices as the average of septum and left lateral wall measurements.
Results: Median frame rate (/s) was 192 (cTDE) and 77 (STE), and median heart rate was 116/min.
Conclusions: We found all cTDE and STE deformation indices significantly different in pairwise comparisons. It is therefore not feasible to use indices by cTDE and STE interchangeably in neonates.

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Table 32. P2-29 Table: Pairwise comparisons of strain and strain rate indices by cTDE and STE.

	cTDE		STE		
	Mean	SEM	Mean	SEM	p
PSS (%)	-15.2	0.5	-17.3	0.5	<0.001
PSSR (/s)	-1.21	0.05	-1.34	0.04	0.004
ESR (/s)	1.51	0.08	2.01	0.10	<0.001
ASR (/s)	1.47	0.07	1.22	0.05	<0.001

SEM: Standard error of the mean. p: p-value for difference between indices by cTDE and STE.

P2-30

Predictive value of right ventricle diameter´s Z score to detect normal versus enlarged right ventricles in postoperated Tetralogy of Fallot children

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Introduction: accurate assessment of right ventricle (RV) size is important in treatment and follow up of congenital heart disease. The aim of the study was to assess the predictive value of RV Z-score diameters in identifying enlarged RV in postoperated Tetralogy of Fallot patients (PTF) according to indexed end-diastolic volumes (RVDVi) from cardiac magnetic resonance (CMR).
Methods: 50 PTF were enrolled (24 patients with RVDVi ≤110 ml/m^2, 16 had 111-150 and 10 were ≥ 150). RV end-diastolic diameter: basal, medial and longitudinal were measured. These parameters were tested with RV diameter´s Z-score made with a 641 healthy pediatric patients cohort (0-18 years), using body surface area (BSA, Haycock formula) and satisfying the assumption of homoscedasticidy and normality of residuals, also considering confounders as gender and inter/intraobserver variability when computed.
Results: PTF mean age was 11 ± 5 years, mean BSA was 1.1 ± 0.4 m^2, mean RVDV was 118 ± 38 ml/m^2, mean Z-score for basal diameter was 36 ± 7.6 mm, for midcavity was 34 ± 6.7 mm and for longitudinal diameter 70 ± 12 mm. There was weak correlation between diameters and VDVDi (Rho Spearman < 33%). But diameter´s Z-scores in ROC curves showed a very good discriminating capacity with AUC >0.8, able to determine which RV were normal sized and which were not (dilated PTF). A cut-off point of >+ 2 z-score provided high specificity (>97%).
Conclusions: Diagnostic validity of RV diameter´s Z-scores to discriminate between normal and dilated RV was significant. Diameter´s Z-scores can accurately predict RV enlargement in PTF patients. As therapeutic decisions may depend on RV progressive dilatation, we offer an easy tool to guide follow up and decision making in PTF children.

P2-31

Reference values and Z scores for right ventricle diameters in caucasian children

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Introduction: quantitative measurements of the right ventricle (RV) is needed in order to detect abnormal RV dimensions in children with congenital heart disease (CHD). RV diameters represent an easy, fast and reproducible way to determine its size. Data relating RV normalized diameters is scant in pediatric population. Reference values and Z scores of RV diameters are provided.
Methods: 641 healthy children (0 days to 18 years old) were enrolled. RV end-diastolic diameters (basal, midcavity and longitudinal) were measured in a 4-chamber focused view and indexed using body surface area (Haycock formula)[1]. Z scores were computed according to literature requirements[2].
Results: RV diameters increased from neonates to adolescents in a nonlinear way. Exponential and square root regression models resulted in the best fit with R^2 of 0.81, 0.82 and 0.9 respectively. They all satisfied the assumption of homoscedasticidy and normality of residuals (Bresch-Pagan and Saphiro-Wilk tests performed). Confounders as gender and inter/intraobserver variability were considered. Predicted Z scores of basal, midcavity and longitudinal diameters plus plots of standardized residuals against BSA are presented. For basal, midcavity and longitudinal diameters and 0.1 m^2 BSA ± 2 Z-score were 8 and 14.7, 6 and 10.6 and 14.7 and 22.8mm respectively. For BSA 0.5 m^2: 12.7 and 23, 12 and 22, 31.7 and 49mm. For BSA 1 m^2: 18 and 32.5, 16.6 and 30, 43.3 and 69mm. For BSA 1.5 m^2: 23 and 42, 20 and 36.5, 53.8 and 83.5mm.
Conclusions: echocardiographic reference values for RV diameters are provided, derived from a large population of healthy children using a rigorous statistical design. These data cover a gap in actual echocardiographic assessment and represent a valid diagnostic tool for RV quantification in children. Determination of RV internal parameters in children with CHDs and its comparison with normal Z scores could provide a new insight in follow up and decision making in CHD.

P2-32

Towards a more accurate planning of invasive procedures: the emerging 3D technologies offer a new approach to the management of complex congenital heart disease

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Background: Three-dimensional (3D) printing technology can be successfully used to create heart prototypes of congenital heart disease (CHD). The use of 3D cardiac models allow for previously unavailable visualization, understanding, handling, and analysis of anatomy of complex cardiac disease. Here we present a virtual cardiac laboratory we recently created in collaboration with the Politecnico University in Milan to facilitate surgical and interventional procedure.
Methods: Based on CT and MRI images performed in selected patients with complex CHD, the cardiac anatomy was segmented using the Mimics software (Materialise, Leuven, Belgium). Out of each patient's data set we generated a virtual computer model and an STL-file that was printed as a 3D plaster model.

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Results: We printed three 3D models and created nine virtual models of patients with CHD. Two patients had a complex intracardiac anatomy (one patient with DORV and one with isolated ventricular inversion with VSD and severe PS) and the models helped the echocardiographers to better understand the spatial orientation of the intracardiac structures, and the cardiac surgeons to identify risk structures, and to plan in detail the surgical strategy reducing the on-pump time. Virtual models of 9 patients with right ventricular outflow tract dysfunction candidate to percutaneous pulmonary valve implantations (PPVI) were created. Data were sheared with our team of engineers to set up a computer simulation of PPVI release and implantation that could predict distortion of the cardiac anatomy and risk of coronaries compression.
Conclusions: The clinical use of 3D models allows to assess, plan, and simulate in-vitro interventions necessary in case of complex congenital malformations. These skills are especially important for young doctors and surgeons in training. In our experience, the printed 3D models had helped the medical-surgical team to gain a better understanding of the technical problems that had to be addressed improving the surgical procedure. The simulation program for PPVI is a useful tool to improve patients' selection and to eliminate any potential surprise during the procedure in vivo. Multi-center studies should be set up to evaluate the impact of cardiac 3D models on medical decisions, the quality of care, and the patients' outcome.

P2-33
Cardiac Abnormalities Determined by Tissue Doppler Imaging and Arrhythmias in Adolescents with Anorexia Nervosa

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Background: Anorexia Nervosa (AN) has a prevalence of 0.5-3% in the adolescent population. Cardiac anomalies including arrhythmias, pericardial effusion and myocardial dysfunction are risk factors in these patients. Tissue Doppler imaging echocardiography (TDI) is an accepted tool in the evaluation of cardiomyopathies. Our objective is to describe cardiovascular anomalies, particularly in TDI in patients with AN.
Methods: Retrospective review of ECG, Holter and echocardiography in 28 patients diagnosed with AN; 20 females and 8 males with a mean age of 14.6 years (range 11 to 19), and mean disease duration of 10.3 months (range 1 to 60).
Results: The ECG was abnormal in 71%: sinus bradycardia (< 60/m) was observed in 16/28 (57%), and prolonged QTc (>460ms), low voltage, and ectopic beats in 4/28 (14%) each. Holter confirmed sinus bradycardia without significant pauses. Wenckebach AV block was observed in one patient. SVT or VT were not observed.
Echocardiography showed structurally normal hearts in all patients. Pericardial effusion was seen in 2/28 (7%). LV mass was decreased (< 44 g/m2) in 3/28 (11%). Contractility was normal with a mean fractional shortening of 38.4% (26-48.9%), except in one patient with 26%, and mean ejection fraction of 0.73 (0.59 to 0.87). TDI evaluation revealed systolic and diastolic dysfunction with decreased S', e' and a' velocities (< -2SD) in the septal and lateral basal segments in two patients (7%), both with severely decreased LV mass. They had a disease duration of 9 and 17 months, no different from the rest. TDI also showed a trend towards low basal segments velocities (S', e', a') between -1SD and -2SD, in 10/28 patients (36%).
Conclusion: Sinus bradycardia was frequent but severe rhythm disorders were not seen in our study. The most relevant finding was that of abnormal TDI velocities indicating systolic and diastolic dysfunction, associated with significant reduction in ventricular mass and independent of disease duration in 2 Pts (7%). A trend for decreased TDI velocities was seen in 36% additional patients. Cardiac complications are a significant cause of morbidity and mortality in AN. Early diagnosis of myocardial dysfunction with TDI could be an indicator for aggressive nutritional intervention.

P2-34
Stereo-Lithographic Models in children with congenital heart disease: an Up-to-Date

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Background: Congenital heart diseases (CHD) represent a very attractive field of application for three-dimensional (3D) printed models. The purpose of this study is to provide a critical up- to-date of current researchers on stereo-lithographic models in children with CHD.
Methods: A systematic search in the National Library of Medicine was conducted in December 2015 starting from the key words: 3d/Stereo-Lithographic models, cardiac, congenital heart disease. The search was further refined by adding the key words children, magnetic resonance imaging (MRI), computed tomography (CT) and 3D-echocardiography.
Results: 17 works were selected for the final analysis, reporting 3d-models reconstruction in 97 cases of CHD. Our research revealed how consistent steps have made in the last decades. Stereo-lithographic representations are able to reproduce complex cardiac/extra-cardiac anatomy including small details with very limited range of errors (< 1 mm). However a series of limitations remain yet. Several steps in the building of 3D models remain highly operator dependent (i.e. images acquisition and their selection, the segmentation process, the choice of materials for printing). In fact not surprisingly in a very innovative field there is a lack of standardization in: 1) procedures employed to collect images (i.e. different techniques-MRI/CT-often used interchangeably, differences in the sequences/projections used, 3D-echo rarely used), 2) in segmentation process (i.e. different software used for DICOM conversion into printable formats, use of manual vs semi-automated/automated segmentation, whether to use systole or diastole not defined); 3) in printers and materials employed for printing. Current 3-D models may be rigid or rubber like flexible (that may be used for surgical simulation) but remain unable to reproduce physiological variations occurring during the cardiac cycle. Furthermore the high costs, particularly those related to software able to convert DICOM files into printable formats and cost of print itself, greatly limit a more extensive use of 3D models.
Conclusions: stereo-lithographic representation of complex CHD anatomy are feasible and very accurate thus may be helpful to plan surgical/percutaneous management in selected cases. Extensive studies to test feasibility, diagnostic accuracy, and cost-effectiveness of stereo-lithographic models in pediatric cardiac surgery/interventional cardiology are warranted.

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Table 1. 3D-print in CHD: Some of the major works.

Author		*Sample size and Age*	*CHD*
Ngan 2 006, Canada	CT	6 cases (6 mths -2 yrs)	PA + VSD + MAPCAs
Noecker 2006 USA	CT	11 cases (age, 3 years; 2 days -13 yrs)	With and without CHD
Schmauss Germany 2014	CT /MRI	4 cases (3 month-16 yrs)	1 vascular ring; 1 VSD, 2 failing SV
Schievano 2007 London	MR	12 cases (20 yrs; 9–39 yrs)	Pts requiring PV implant
Shiraishi 2010 Japan	CT	8 cases (4 days-4 years)	Aco, HLHS, DORV, AVSD, APVER. PDA
Olivieri 2015 USA	3D echo	9 cases	8 VSD, 1 peri-valvular leak
	Software for segmentation	***Printer***	***Materials***
Ngan 2 006, Canada	Mimics, Materialize, Lueven, Belgium	Stratasys Prodigy Plus (Stratasy Inc, Minn)	Solid acrylic or plastic model
Noecker 2006 USA	Stereolithography machine (SLA 250/30 A, 3D Sys, Valencia, USA)	Z printer 310, Z Corporation, Burlington MA, USA	polyurethane + silicone rubber
Schmauss Germany 2014	Amira (Mercury Comp. Systems, MA, USA) cellulose + elastomeric urethane resin	Spectrum Z™ 510, Z Corporation, Burlington, MA, USA	starch/
Schievano 2007 London	Not reported	P1500, Stratasys, (Stratasy Inc, Minn)	Thermoplastic resin
Shiraishi 2010 Japan	Stereolithography biomodelling company (Japan)	Not reported	Rubber like models urethane
Olivieri 2015 USA	Mimics, Materialize, Lueven, Belgium	Object 500, Connex, Polyjet	Not reported

P2-35
Digital assessment of endothelial function and its association with clinical variable in patients with Fontan operation

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Background: Endothelial dysfunction has been demonstrated in prior studies of Fontan survivors. However it's clinical determinates remains unclear.
Purpose: To clarify the clinical determinates of endothelial dysfunction in Fontan patients.
Methods and Results: We measured endothelial function using reactive hyperemia-peripheral arterial tonometry (RH-PAT) as RH-PAT index (RHI) in 26 Fontan patients (aged 15 to 32 years, male gender 62%) and 6 healthy control (aged 29 to 38 years, male gender 66%), and compared the result with the clinical variables, including hemodynamics, plasma level of natriuretic peptide, lipid profile, glucose tolerance, and exercise capacity. RHI was converted into a natural logarithmic form. The mean RHI was 0.56+ /- 0.26 in Fontan patients and 0.78+ /- 0.31 in control ($p = 0.09$). RHI in Fontan patients was associated with diastolic blood pressure ($r = -0.37$, $p < 0.05$), heart rate ($r = -0.49$, $p < 0.05$), and hemoglobin A1c level ($r = -0.60$, $p < 0.01$). Medication use, central venous pressure, cardiac output, exercise capacity, plasma level of natriuretic peptide and lipid profile did not associated with RHI.
Conclusion: Endothelial function assessed using RH-PAT in Fontan patients was associated with abnormal glucose tolerance and arterial stiffness rather than hemodynamics and heart failure severity. Glucose regulation might be a potential target for the treatment of endothelial dysfunction in Fontan patients.

P2-36
Right ventricular longitudinal strain assessment in children with severe pulmonary arterial hypertension associated with congenital heart defects

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Objectives: Right ventricular function has been identified as an important prognostic factor in children with pulmonary arterial hypertension. The aim of the study was to assess the right ventricular longitudinal strain in children with severe pulmonary arterial hypertension associated with congenital heart defect.
Methods: We evaluated prospectively 37 children (16 children with severe pulmonary arterial hypertension associated with congenital heart defects; and 21 age and sex match controls) using conventional and speckle-tracking echocardiography (iE33, Q LAB 10), brain natriuretic peptide level and clinical parameters (WHO functional class, 6-minute walk test). Right ventricular free wall strain was measured by averaging three (basal, medial, apical) regional peak systolic longituginal strain/ strain rate.
Results: Right ventricular free wall longitudinal strain and strain rate were significantly lower in patients group compared with controls ($p < 0.05$); both has been correlated with right ventricular fractional area change, myocardial performance index, left ventricular eccentricity index and brain natriuretic peptide level ($r = -0.49$, $r = 0.58$, $r = 0.64$, $r = 0.41$ respectively $r = -0.38$, $r = 0.44$, $r = 0.46$, $r = 0.41$) with a $p < 0.05$.
Conclusions: Right ventricular longitudinal strain/strain rate assessment is useful in noninvasive evaluation of right ventricular function in children with pulmonary arterial hypertension associated with congenital heart defects.
Aknowledgement: The present study was carried out in the research project no 27/11.12.2013, financed by UMF Tg. Mures
Key words: right ventricle, longitudinal strain, congenital heart defects, pulmonary arterial hypertension, children

P2-37
Cardiac Mechanics in Young Patients After Arterial Switch Operation for d-Transposition of the Great Arteries and Gothic Aortic Arch Morphology

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Background: In patients who have undergone arterial switch operation (ASO), aortic arch has been found to be more acute and associated with abnormal bioelastic properties. Our study aims were to assess in young patients after successful ASO 1- the occurrence of Gothic aortic arch (GA); 2- the association between GA and aortic stiffness; and 3- the impact of GA on left ventricular (LV) function using standard echocardiography and speckle tracking echocardiography (STE).
Methods and Results: We studied eighty consecutive asymptomatic patients, who have undergone first stage ASO for simple d-TGA, with normal left ventricular ejection fraction (LVEF $\geq$ 53%).

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Forty-two (52%) patients showed an GA (mean age 10.8 ± 6.6 years, 26 males) while thirty-eight (48%) patients (mean age 9.2 ± 6.1 years, 27 males) did not present GA. There were no significant differences for age, sex, BSA, and blood pressure values between groups. In group GA left atrial volume indexed (Group AAA = 17.8 ± 9.7 vs 13.2 ± 8.4ml, p = 0.025) and aortic stiffness index (Group AAA = 2.05 ± 1.2 vs 1.5 ± 0.7, p = 0.016) were significantly increased, while basal LV longitudinal strains were significantly reduced .

Conclusions: GA is responsible for significant increase in aortic stiffness, dilated left atrial volume, and impaired LV longitudinal systolic deformation in the basal segments in young patients after ASO.

P2-38

Evaluation of ventricular function in Fontan patients undergoing feature tracking magnetic resonance strain

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Introduction: Feature tracking strain (FTS) is a new technique to assess cardiac function from cardiac magnetic resonance (CMR). We compared FTS with conventional function parameters in single ventricle subjects with Fontan physiology undergoing CMR.

Methods: 18 Fontan subjects (mean age 17.6+ /- 9.2 years, post Fontan period 14.2+ /- 8.2 years, 13/18 morphologic right ventricle, 5/18 morphologic left ventricle) underwent a CMR study. Ventricular volumes and cardiac output were calculated off-line (Medis QMass advanced edition, the Netherlands). Off-line strain and strain rate (SR) analyses were performed (TomTec Image Arena, Germany) on the 4-chamber and short-axis views at the basal, mid, and apical levels of the ventricle.

Results: Basal endocardial circumferential strain/SR (-11+ /- 9% and -0.8+ /- 0.5 1/s) were lower than it at the mid (-17+ /- 6% and -1.1+ / -0.5 1/s; p = 0.02 and 0.02) and apical (-26+ /- 9% and -1.9+ /- 1.0 1/s; p = 0.001 and 0.003) levels. There was correlation between average endocardial longitudinal strain /SR and ventricular end-diastolic volume (r = 0.73 and 0.68). At the mid and apical levels, there was correlation between average endocardial circumferential strain/SR and ventricular end-systolic volume, and ejection fraction (r = 0.53 to 0.85). There was also correlation between average endocardial radial strain/SR and ventricular stroke volume at the mid level (r = 0.69 and 0.77).

Conclusions: There is correlation between strain/SR and measures of ventricular volume, ejection fraction and cardiac output in single ventricle subjects with Fontan physiology. FTS is independent of inadequate acoustic windows unlike echocardiography and could have clinical relevance. Analysis of regional strain may helpful in understanding myocardial mechanics in the single ventricle in further studies.

P2-39

Relationship between Systolic and Diastolic Function during Exercise in a Healthy Pediatric Cohort

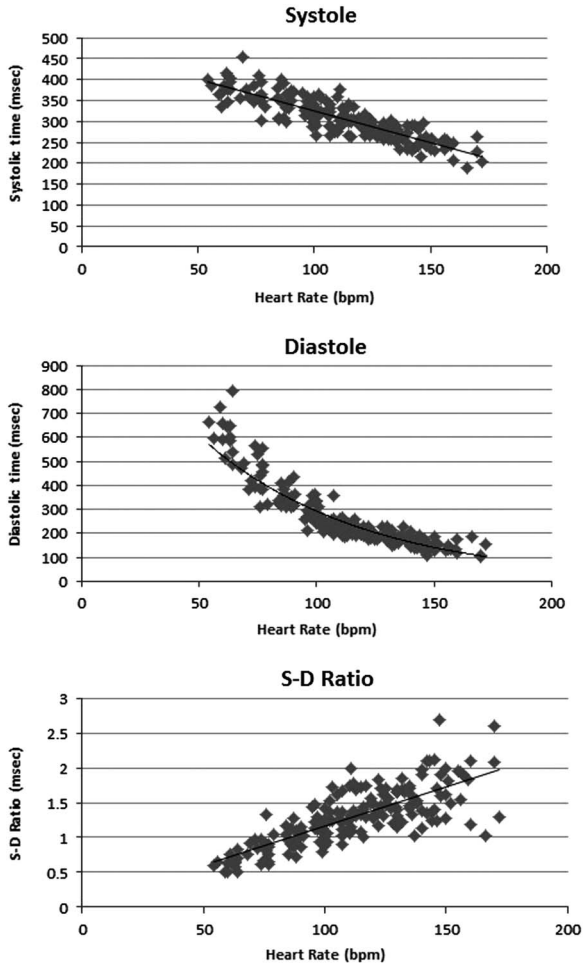
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Introduction: The normal relation between systole and diastole in response to exercise has not been previously described in a pediatric population. The aim of the current study was to evaluate systolic and diastolic durations by pulsed tissue Doppler imaging and the resultant systolic to diastolic duration ratio (S:D ratio) during exercise in a normal pediatric cohort using semi-supine cycle ergometry stress echocardiography (SSCE).

Methods: A total of 38 healthy children (22 females) were recruited. Median age at the study was 14 years (range 8 to18 years). A stepwise SSCE protocol was used. Systolic and diastolic duration was measured at rest and at incremental HR from pulsed tissue Doppler sampled at the lateral mitral annulus. S:D ratio was calculated. The relationship with increasing HR was evaluated for each parameter by plotting the values at each stage of exercise versus HR.

Results: Pulsed tissue Doppler images could be measured in all subjects during exercise. A linear relationship was observed between HR systolic duration and S:D ratio, but the relation between diastolic duration and HR was exponential (see figure).



Conclusions: This study provides the normal variability of systolic and diastolic time duration, and of the derived S:D ratio, measured at incremental HR during exercise in a healthy pediatric cohort. These data can be used as reference to evaluate alterations of systole/diastole relationship with increasing HR in different pediatric disease conditions.

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P2-40

Dramatic dose reduction in three-dimensional rotational angiography after implementation of a simple dose reduction protocol

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Introduction: Three-dimensional rotational angiography (3DRA) is a relatively new imaging modality in the paediatric catheterization laboratory (cathlab). Previously, we reported a mean effective radiation dose of 2.0 mSv per 3DRA run in children (normal dose cohort). The objective of this study was to evaluate the effect of implementation of a dose reduction protocol on effective radiation dose (ED) and image quality in 3DRA. Furthermore possible correlations between ED and readily available parameters at the cathlab were studied.
Methods: The dose reduction protocol consisted of several dose reducing techniques: frame reduction (60 f/s to 30 f/s), active collimation of the X-ray beam, usage of a readily available low dosage program and a pre-3DRA run check. Biomedical data required for calculation was collected. ED's were calculated with Monte Carlo PCXMC 2.0. 3DRA Image quality of both the dose reduction cohort as well as our normal dose cohort was blindly assessed by one observer.
Results: Between October 2014 and October 2015 84 patients (mean age 6.4 years) underwent a total of 105 3DRAs with a mean ED of 0.67 mSv ± 0.45 SD using the dose reduction protocol. Mean ED in the normal dose cohort (17 3DRA's) was 2.0 ± 1.4 SD. Image quality in the dose reduction cohort remained excellent. Correlations between ED and dose area product (DAP) and ED and dose (mGy) were found with a R^2 of 0.7936, and 0.76164 respectively.
Conclusions: Introduction of a simple protocol led to 67% dose reduction in 3DRA. Mean ED in 3DRA in the dose reduction group was 0.67 mSv and image quality in this group remained excellent.
In 3DRA ED correlates well to DAP and mGy, parameters readily available at the cathlab.

P2-41

4D flow CMR and CT of the left ventricular outflow tract and neo-aorta after the arterial switch operation

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Introduction: Neo-aortic root dilatation, a common complication after the arterial switch operation (ASO) for transposition of the great arteries (TGA), could theoretically be related to the changed geometric and hemodynamic characteristics of the reconstructed left ventricular outflow tract (LVOT) after ASO. The aim of this study was to determine if geometric characteristics of the LVOT and related blood flow patterns are associated with neo-aortic root dilatation by using four-dimensional (4D) flow cardiovascular magnetic resonance (CMR) and cardiovascular computed tomography (CCT).
Methods: The study cohort consisted of 59 TGA patients who underwent ASO between 1978 and 2001. The following neo-aortic root measurements were performed using CCT: (1) surface area of aortic valve annulus indexed by Z-score, (2) surface area at level of aortic root sinuses indexed by Z-score and (3) spatial relationship between aortic root and pulmonary trunk in the transverse plane. (4D flow) CMR was used to assess (4) the geometric relationship between LV and ascending aorta, expressed by 4 angles between the LV and LVOT as seen in Figure 1 and (5) degree of flow eccentricity by measuring the angle between peak systolic flow direction and aortic valve annulus. Correlations were determined using Pearson's correlation coefficient.
Results: Neo-aortic root dimensions were increased (mean ± SD for *Z* annulus: 2.98 ± 1.60, *Z* sinus: 2.43 ± 1.41). Neo-aortic root dimensions were not correlated with any of the four angles as described (4) ($r < 0.06$, $p > 0.05$), nor to the spatial relationship between aortic root and pulmonary trunk (3) ($r = -0.20$, $p = 0.14$) or flow eccentricity (5) ($r = 0.04$, $p = 0.79$). Patients with pre-operative ventricular septal defect (VSD) (n = 16, 27%) had similar aortic root dimensions as patients without and the presence of pre-operative VSD was not associated with aortic root dilatation. ASO patients had good ventricular systolic function (mean LVEF 56.8% (95% CI[55.3, 58.3])).

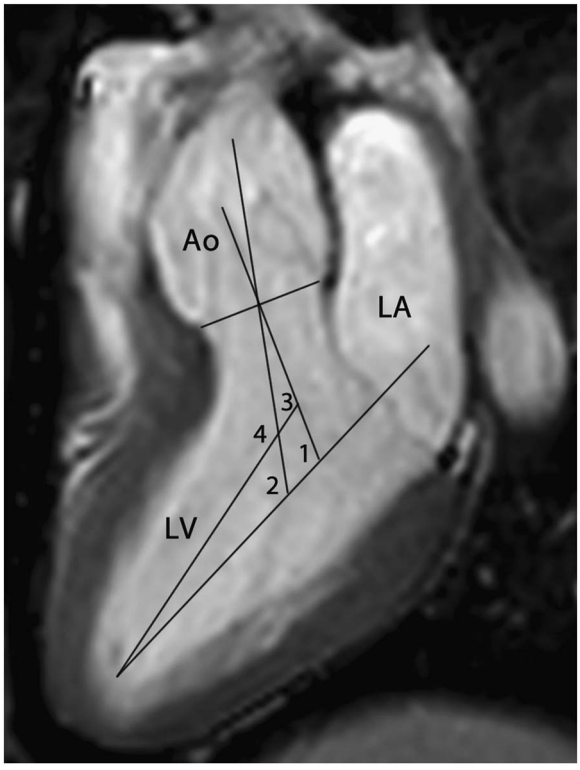


Figure 1
CMR image of a 3 chamber LV view during systole with (1) angle between LV inflow (line bisecting mitral valve annulus to LV apex, based on expected intracardiac blood flow) and line perpendicular to aortic valve annulus; (2) angle between LV inflow and aortic root long axis (line bisecting aortic valve annulus); (3) angle between LV intracardiac outflow during systole (based on expected intracardiac blood flow) and line perpendicular to aortic valve annulus; (4) angle between intracardiac outflow during systole and aortic root long axis. Ao, aorta; LA, left atrium; LV, left ventricle.

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Conclusions: Neo-aortic root dilatation in TGA patients is not related to the geometric characteristics of the LVOT after ASO, nor to the pre-operative presence of a VSD. Neo-aortic root dilatation is likely to be explained by other factors like abnormal aorticopulmonary septation and surgical manipulations.

P2-42

Evaluation of Left and Right Ventricular Functions Using Conventional and Tissue Doppler Echocardiography in Children with Type 1 Diabetes Mellitus

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Introduction: Diabetes mellitus (DM) is one of the most common chronic diseases occurring in childhood. Doppler echocardiography has become common in clinical practice, due to its non-invasive nature and reproducibility of the results, ease of application. The aim of the present study was to evaluate the effects of glycemic control on left and right ventricular functions patients with type 1 diabetes mellitus.
Methods: The study was conducted in the Yüzüncü Yıl University between September 2013 and October 2014. The patient group comprised of 86 patients with type 1 diabetes aged between five and 17 years. The patients with HgA1c levels of 9% or lower were allocated into the well-controlled diabetes group and patients with HgA1c above 9% were allocated into the poorly controlled diabetes group. The control group was consisted from 54 healthy children. M-mode, 2-D, PW Doppler examinations and tissue Doppler imaging studies were performed for all subjects.
Results: Mitral annulus PW tissue Doppler showed significantly lower Em and Em/Am ratio in the well-controlled group. Am, IVRT and MPI were found to be significantly higher in the poorly controlled. IVCT, Sm and ET did not significantly differ between groups. The E to Em ratio of the mitral valve was significantly higher in the well-controlled and poorly controlled group. PW Doppler examinations showed that, six patients (13.6% in the well-controlled group and 13 patients (31%) in the poorly controlled group had type 1 diastolic dysfunction. Tissue Doppler examination showed that eight patients (18%) in the well-controlled group and 17 patients (40.4%) in the poorly controlled group had type 1 diastolic dysfunction. None of these patients exhibited a false normal filling pattern.
Conclusion: The present study showed that left ventricular systolic functions and right ventricular systolic and diastolic functions are preserved, although left ventricular diastolic function is impaired, during the early period in the poorly controlled group.

P2-43

Myocardial deformation characteristics of the systemic right ventricle after atrial switch operation for transposition of the great arteries

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Introduction: The atrial switch operation (Senning) has been the main surgical repair technique for d-transposition of the great arteries for many years. The Senning procedure results in a subsystemic morphologic right ventricle (RV) and a subpulmonary morphologic left ventricle (LV). This can be regarded as a model for the effects of long-term pressure overload on the RV, and of ultimately decreased afterload on the LV. We sought to determine the impact of these chronically altered loading conditions on the myocardial deformation of the RV and LV.
Methods: 26 patients after Senning (age 28.4 ± 7.5y) and 18 normal controls (age 22.2 ± 11.4y; $p = 0.034$) underwent cardiac magnetic resonance (CMR) imaging. 2D SSFP cine images were acquired in an horizontal long axis and in a short axis covering both ventricles and post-processed with a feature tracking software (TomTec 2D CPA). Global circumferential strain was measured on a short axis mid-ventricular slice. Global longitudinal strain was measured in a long axis, separately for each ventricle.
Results: When comparing RV in either position, subsystemic circumferential strain was higher than subpulmonary circumferential strain ($-16.1 \pm 2.9\%$ vs.$-13.1 \pm 4.3\%$; $p < 0.01$), and subsystemic longitudinal strain was lower than subpulmonary longitudinal strain ($-12.8 \pm 3.3\%$ vs.$-18.3 \pm 3.8\%$; $p < 0.001$). In contrast, LV global strain in subsystemic vs. subpulmonary position was similar: LV circumferential strain ($-23 \pm 13.1\%$ vs.$-20.2 \pm 3.9\%$; n.s.); LV longitudinal strain ($-17.5 \pm 4.6\%$ vs.$-16.1 \pm 5.3\%$; n.s.).
The subsystemic RV showed lower circumferential ($-16.1 \pm 2.9\%$ vs.$-23 \pm 13.1\%$; $p < 0.05$) and lower longitudinal strains ($-12.8 \pm 3.3\%$ vs.$-17.5 \pm 4.6\%$; $p < 0.001$) than the subsystemic LV. The subpulmonary LV exerted greater circumferential strains ($-20.2 \pm 3.9\%$ vs.$-13.1 \pm 4.3\%$; $p < 0.001$) but similar longitudinal strains compared to the subpulmonary RV ($-16.1 \pm 5.3\%$ vs.$-18.3 \pm 3.8\%$; n.s.).
Conclusions: In discordant ventriculo-arterial connection, the subsystemic RV adapts to the increased afterload with an increase in circumferential strain and an impaired longitudinal deformation. This may represent the effect of a positive interventricular interaction due to the shared circumferential fibers, since the LV shows higher circumferential strain than the RV even in subpulmonary position.

P2-44

Use of the amplatzer vascular plug II device to occlude different types of patent ductus arteriosus in pediatric patients

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Objectives: To evaluate the outcome of the Amplatzer Vascular Plug II (AVP-II) for closure of the patent ductus arteriosus (PDA) in children.
Methods: All patients undergoing transcatheter closure of PDA with AVP II from June 2013 to November 2015 were

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retrospectively evaluated. Clinical, angiographic, and echocardiographic data were analyzed.
Results: Twenty procedures were performed in 19 patients. Median age was 18 months (6–202 months) with median weight 9.9 kg (5.1–63). The morphological PDA classification was Type A in 4/19 (21%), Type C in 14/19 (74%), and Type E in 1/21 (5%). The median minimum PDA diameter, maximum and length was 3.24 mm (1.7-4.7 mm), 8.47 mm (3.3-17 mm), and 12.79 mm (11-21 mm) respectively. The implanted device sizes were: 6 mm in 3/20 (15%), 8 mm in 9/20 (45%), and 10 mm in 8/20 procedures (45%), The implanted device was mean of 2.97 ± 0.87 times the ductus narrowest diameter and mean of 1.03 ± 0.36 times the ductus largest diameter in successful procedures. The mean procedure and fluoroscopy time was 35 minutes (15-60 minutes) and 8.4 minutes (3.1-12.8 minutes). In four patients closure was performed from the arterial side. All procedures except one were successful; among succesfull procedures 100% 'in-lab' and 100% closure on post-procedural echocardiogram was achived. No left pulmonary artery stenosis and aortic obstruction observed with a median follow-up duration of 9 months (0-29 months).
Complications: In one patient device had to remove due to pulmonary hypertensive crisis. And in the last patient 8 mm device was embolized in to the pulmonary artery due to ductal spasm which caused underestimation of the duct. And on the next day 8mm device was retrieved and a 10 mm device was implanted.
Conclusions: The AVP II seems to be an effective and safe device for PDA closure in children. It is particularly useful in type C and E ductus and in small infants where it eliminated the risk of device-related aortic obstruction.

P2-45
Stenting of the right ventricular outflow tract: single center experience

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Background: Neonates and infants with decreased pulmonary blood flow due right ventricular outflow tract (RVOT) obstruction may need shunt to treat hypoxemia. We report our experience about stenting RVOT as an alternative procedure to temporarily improve pulmonary blood flow.
Methods: Retrospective procedural chart review of patients undergoing stenting of the RVOT between July 2014 and July 2015 was performed.
Results: Nine patients underwent 11 RVOT stenting procedure. One patient had an RVOT stent when he was 3 days old and stent was dilated with another stent (stent-in-stent) 3 months later. The other patient who required second procedure needed ECMO because of cardiac arrest during the first procedure and 2 days later RVOT stenting could be performed under ECMO.
In 2 patients transient complete AV block occurred and one returned to sinus rhythm during the procedure. Although second patient's AV block continued for 2 days did not needed pacemaker (this patient was under ECMO). In one patient (5-year-old), who is also under ECMO, both RVOT stenting, left pulmonary artery stenting and recanalization of central shunt procedures were performed. None of the patients died after successful intervention.
In one patient procedure was canceled (long sheath could not be advanced to the RVOT) and in one patient stent embolized in to the ventricle and patient underwent surgical extraction of the stent and BT shunt.
Conclusion:
- Stenting of the RVOT can be a treatment option for selected patients with very reduced pulmonary blood flow.
- Interventionist should be aware of the complications.

P2-46
Transcatheter Retrieval Techniques for Foreign Bodies in Children

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Introduction: Many techniques for the endovascular retrieval of lost, embolized or misplaced foreign objects have been developed, and the removal of almost every foreign object has become possible. In this paper, we report our experience in retrieving foreign objects
Methods: This study was a retrospective analysis of the records of all patients in our institution for transcatheter retrieval of intravascular foreign objects.
Results: Between January 2001 and November 2015, 2270 cardiac catheterizations were performed. Among them, 9 patients underwent percutaneous intravascular foreign body retrieval. Patients ages were ranged between 15 days to 10 years.
The foreign bodies consisted of guidewire (2), fractured catheter (2), PDA device (2), ASD device (1), Gianturco coil (1), and stent (1). Five retrievals were performed with single-loop snares easily with usual methods.
In one patient, stent was embolized to the descending aorta during PDA stenting. Initially stent was snared distally and taken into the guiding catheter. Later proximal side was snared and elongated by another snare. Than it was taken into another guiding catheter which was advanced through the contralateral femoral artery.
Misplaced ASD closure device cannot be retrieved with snare or bioptomer catheters. It was retrieved by re-screwing method. But during placement of the delivery cable in the slot of the device, a telescopic method was used.
A 2100 gram premature infant, referred to our clinic 60 days after the embolization of umbilical artery catheter. During catheterization foreign body cannot be retrieved from the proximal part due to adhesion. Snare catheter was advanced distally and snared from distal part. Although we try to pull and push it, the proximal part did not move. So snare was opened at the distal part and gently withdrawn towards the proximal part. After reaching proximal part it was snared and pulled out.
Conclusion: Device embolization or misplacement may occur during transcatheter device deployment. Retrieval of misplaced or embolized devices can be performed percutaneously. Cardiologist dealing with intervention should have several plans to retrieve the devices before using them.

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P2-47

Transcatheter closure of right coronary artery fistula to the right ventricle

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Introduction: Coronary artery fistula (CAF) is an uncommon anomaly usually congenital but can be acquired. It accounts around 0.2-0.4% of congenital cardiac anomalies. Although, most of the patients are asymptomatic, some may present with Congested Heart Failure (CHF), infective endocarditis, myocardial ischemia (which may present as chest pain or ventricular arrhythmias due to steal from the native coronary vessels) or rupture. Some are picked up with an incidental heart murmur.
In the past, surgical ligation was the only option in the management of CAF, but since 1983, transcatheter closure of CAF has been increasing as an alternative to surgery. Elective closure of significant CAF in childhood has been advocated to prevent later complications.
Method and result: We report a 3 year old boy with a large right coronary artery fistula to the right ventricle who had a successful transcatheter closure. Our case is differs from other CAF in that the fistula was communicating the Right Coronary Artery (RCA) itself to the Right Ventricle (RV).
Conclusion: Transcatheter closure of CAF in children is a safe alternative procedure to surgical ligation specially if the fistula is communicating the Right Coronary Artery (RCA) itself to the Right Ventricle (RV).

P2-49

The 'Dog Bone Technique'-A novel easy and safe catheter maneuver for aortic arch and coarctation stenting

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Background: Various techniques are described to facilitate stable stent implantation in aortic arch stenosis or coarctation. We describe an alternative technique, which due to the unique appearance during stent implantation, we have named 'Dog Bone Technique' (DBT).
Technique: The stent/ balloon assembly is placed proximal the stenosis, the long sheath is retrieved to uncover the distal 20-50% of the stent. The balloon is inflated with the pressure inflator just to expand slightly the stent. Thereafter the proximal end is uncovered and partially inflated; therewith the assembly takes the typical 'dog bone' shape before complete inflation and final positioning. Repositioning of the stent and control angiography is possible at each time of this procedure if needed.
Results: Between 1/2010 and 12/2014 we implanted 91 stents in 87 patients (mean age 20.2 years). 71 patients had typical native or re-coarctations and 16 patients had transverse aortic arch stenosis. In 38 patients (44%) a pharmacological exercise test with Orciprenaline was performed during implantation resulting in high cardiac output. In none of the patients reduction of cardiac output by adenosine or a rapid pacing of the right ventricle was required for stable stent implantation. All stents were implanted in the targeted position without any displacement using this single balloon technique. There were no acute or short-term complications detected.
Conclusion: DBT is a safe and feasible technique for aortic stent implantation even at high cardiac output. Other additional techniques for stent placement are not necessary to obtain a stable final position in the target region.

P2-50

Fontan completion – does it really relieve cyanosis?

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Objectives: Total cavopulmonary connection (TCPC) aims for reduction of ventricular volume overload and cyanosis. However, due to the use of fenestration and the development of venovenous (vv.) collaterals, some patients remain cyanotic or desaturate during follow-up (FU). We analyzed the persistence or development of secondary cyanosis during long-term FU after TCPC.
Methods: We retrospectively studied 267 patients (median age 15 (3-57) years), who underwent TCPC at our institution between 1986 and 2015. 16 early deaths were excluded and 251 enrolled in the long-term analysis. During median FU of 6 (0.3-30) years after TCPC, all patients underwent routine investigations in our outpatient department, including 142 cardiac catheterizations for fenestration closure and/or hemodynamic evaluation if indications were given. Transcutaneous oxygen saturation (SaO2) was noted and compared longitudinally. Cyanosis was defined as $SaO2 \leq 93\%$.
Results: Total long-term survival was 90% ($N = 226$). Total long-term mortality ($N = 25$) significantly correlates with cyanosis during FU (20 cyanotic vs. 5 non-cyanotic patients, $p < 0.001$). On discharge median SaO2 of the total cohort was 94% (78-100%), 95% (80-100%) in patients without fenestration and 91% (78-98%) in patients with fenestration. In 43 patients fenestration was closed. SaO2 increased to median of 96% (88-100%) ($p < 0.01$). On last FU SaO2 was in median 95% (69-100%), 92% (72-99%) in patients who still have an open fenestration and 96% (69-100%) in those without. $SaO2 \leq 85\%$ was noted in 15 patients, of whom 6 died. 142 patients received catheterization during long term FU; vv. collaterals were identified in 56 and closed in 53. A total of 37% ($N = 94$) were cyanotic at last FU. In 47% ($N = 44$) this can be explained by an open fenestration, in 17% ($N = 16$) by vv. collaterals. In 36% ($N = 34$) no reason for cyanosis could yet be found.
Conclusion: Our study shows that a significant proportion of TCPC patients stay or again become cyanotic at long-term FU. More attention should be directed to the development of desaturation during long-term FU to treat and prevent long-term complications.

P2-51

Transcatheter Takedown of the Failing Fontan

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Case 1 underwent staged palliation of tricuspid atresia and truncus arteriosus. Lateral tunnel Fontan was performed at 18 months due to cyanosis secondary to arteriovenous malformations. He returned 2 weeks post Fontan with pleural effusions. Despite fenestration stenting, he developed profuse drainage, low cardiac output and renal failure. He was unlikely to survive to transplant. At catheterisation, a Rashkind balloon was used to interrogate the lateral tunnel for occlusion device sizing. This was then used to control SVC return, permitting fenestration stent dilation to 14mm. A 12mm Amplatzer muscular VSD device was then deployed into the proximal tunnel. The patient had an embolic

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stroke post procedure but recovered to extubation. He continues to require peritoneal dialysis and oxygen, his neurology has recovered, he was discharged home. He has since been assessed as unsuitable for cardiac transplant due to renal failure.

Case 2 underwent staged palliation of hypoplastic left heart syndrome. He acutely failed after extra-cardiac fenestrated Fontan with low cardiac output. Catheterisation at < 24 hours in hybrid theatre revealed Fontan pressures of 24 mmHg. A sheath was placed directly via the right atrial appendage and the fenestration was stented to 10mm. The conduit was oversewn with two layers of continuous proline between two clamps, a third of the diameter of the conduit was occluded with a Ligaclip. Pleural drain losses continued, after 4 weeks he underwent further dilation of the fenestration stent to 15mm. Renal and hepatic function recovered, he was extubated after 6 weeks and drains were removed. He is now weaning off oxygen, mobile and referred for cardiac transplant.

Transcatheter takedown of Fontan may remove some of the mortality associated with further cardiac bypass. Consideration should be made to thrombus formation in the 'dead space' between a fenestration and a conduit occlusion device. In the early failing Fontan, this newly described 'Hybrid' approach may be preferred, it may also act to preserve the potential lumen of the extra-cardiac conduit for future attempts at transcatheter recanalisation to Fontan circulation.

P2-52
Multicenter Off-label use of Nit-Occlud coil in percutaneous closure of small Patent Ductus Arteriosus

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Introduction: The Nit-Occlud coil (NOC) is a nitinol coil-type patent ductus arteriosus (PDA) occluder with a reverse cone configuration, which is implanted antegrade using a controlled delivery system. Off-label retrograde approach of NOC has never been reported.

This study aims to assess the efficacy and safety of the NOC to close small PDAs using the retrograde approach. The need for such modification is justified by the low cost of NOC and the non-availability of other retrograde-approach devices.

Methods: Between Jan/2013-Dec/2015, fourty-two patients with small PDAs less than 3 mm from two centers underwent attempts of retrograde closure of small PDAs by PFM-NOC.

The duct was crossed retrograde and the coil was delivered through the PFM end-hole catheter in 20 cases from the first center (51%). In the remaining 19 cases from the second center, the coil was delivered direct through the Judkins catheter to decrease fluoroscopy and procedure times. We released 1-2 coil loops in PA and the rest in the duct or aorta. The mean follow up time was 15.1+ /- 14 months.

Results: The study included 25 (59.5%) females and 17 (40.5%) males with a median age of 1.5 years and a median weight 9.75 kg. PDA was type A, C, D and E in 19, 13, 4 and 6 patients respectively. The mean sizes of pulmonary and aortic ends were 1.22 and 2.9 mm respectively. NOC size 5 × 4 was used in majority of patients (66.7%) and size 6 × 5 was the second common (23.8%). The mean procedure and fluoroscopy times were 51.6 and 8.9 minutes respectively.

The procedure was successful in Thirty-nine cases (93%). Complete closure in the same or following day was achieved in 38 patients (97%). There were no deaths or serious adverse events. Three patients failed (7%), two due to embolization and coarctation of the Aorta and were retrieved and removed, while one was due to failure to cross the PDA.

Conclusion: Retrograde closure of small PDAs using PFM Nit Occlude coils is safe, effective and feasible. It offers a cheap alternative to conventional retrograde designed devices.

P2-53
Closure of perimembranous VSD using the Amplatzer duct occluder I device

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Introduction: Transcatheter closure of perimembranous VSDs (pmVSD) has been tempered by increasing awareness of procedure-related complications, in particular AV block in young patients. In developing countries, the prohibitive cost of devices specifically designed for pmVSD closure has also limited their use. We report on off - label use of the Amplatzer duct occluder device (ADO I) in this setting.

Methods: 35 patients (20 male), median age 3.5 years (range 7 months - 17 years), median weight 14 kg (5.2-51), BSA 0.57 M2 (0.29-1.39) and median LVEDD z score of 2.6 (-0.7 to +4.9) underwent the procedure, based on a combination of clear clinical indications. In 12 patients a continuous arteriovenous loop through the VSD was developed to deliver the device (median fluoroscopy time of 41 minutes) . In 23, a direct venous approach was used (median fluoroscopy time of 25 minutes).

Results: Median VSD Diameter was 6mm (3-9mm). Device sizes used were 6/4 (n = 6), 8/6 (n = 12), 10/8 (n = 14), and 12/10 (n = 3). Two procedures were aborted after the initial device chosen pulled through the VSD. In the remainder the VSD was successfully closed. Complete closure rates were 91% immediately, increasing to 97% at 6 weeks. No patient developed heart block upto the most recent follow-up. None had new onset aortic or tricuspid valve insufficiency. Antifailure medications were discontinued in all patients following successful closure.

Conclusions: A significant proportion of symptomatic pmVSDs in small patients can be cheaply and effectively closed with the Amplatzer ADO I device.

P2-54
Interventional cardiology in children and radiation-induced cancer risk

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Objectives: Children with congenital heart disease frequently undergo interventional cardiology procedures (ICP) for diagnostic or therapeutic purposes. Despite the clear clinical benefit to the patient, the complexity of these procedures may result in high cumulative radiation exposure. This issue is particularly relevant for children given their greater sensitivity to radiation and the longer life span during which radiation health effects can occur. Epidemiological data are, however, still awaited for providing a lifelong overview of potential cancer risks. In France, an epidemiological cohort study, named Coccinelle (French acronym for « Ladybird »), is carried out. The specific objectives of this

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nationwide study are to characterise the paediatric population that underwent ICP in France; to estimate doses associated with ICP procedures and finally to assess the hypothesis of an excess risk of solid cancers and leukaemia attributable to ionising radiation exposure during ICP in children.
Methods: All children who have undergone at least one ICP before age 10 since 2000 are included from main French hospitals. For each ICP, dosimetric parameters (kerma area product, fluoroscopy time) are retrieved retrospectively. Organ doses, especially to the lung, breast, thyroid, and bone narrow are calculated using dedicated software. Children's exposure to CT scans, which account for a major contribution to their medical exposure dose, is also collected. Long term follow up of the cohort will be performed by linkage with French paediatric cancer registries. In the meantime, a quantitative risk assessment approach allows giving predictions of potential lifetime risks of cancer incidence.
Results: Up to now, about 9,000 children have been included. The cohort profile will be presented as well as reference levels for the main ICP performed. Using this exposure data, excess risks of leukaemia, breast, lung and thyroid cancers will be calculated using dose–response models derived from the Japanese atomic bomb survivors' cohort and from studies of medical exposures.
Conclusion: The Coccinelle study is specifically designed to provide further knowledge on the potential cancer risk associated with paediatric ICP. It will also provide comprehensive information on typical levels of doses for ICP in France.

P2-55
Reducing shunts in congenital heart disease: gentle telescoping pigtail technique for device delivery

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Introduction: AMPLATZER(TM) Vascular Plug II and IV are well-established to reduce collateral flow via superfluous arterial and venous connections associated with congenital heart disease (CHD). However, it can sometimes be challenging to access the target lesion and deliver the optimal device. Compared to the Plug IV, the Plug II is sometimes preferable due to its shorter length, flat profiled retention disks and availability of large diameters, but its profile means the preferred delivery catheter needs a larger inner lumen. In this respect, we present a new simplified telescoping technique for Plug II delivery.
Methods: Retrospective analysis of all CHD patients who underwent a vascular occlusion procedure with Plug II/IV between 12/2012 and 12/2015. Target blood vessels were characterized, measured, and the size and number of implanted devices registered and device-to-vessel diameter-ratio calculated. The refined telescopic technique for Plug II delivery worked by probing the target vessel with a floppy wire, which was simply reinforced by the curved tip of a standard pigtail catheter at the proximal vessel-segment to guide the needed delivery sheath/catheter. Deployment technique, complications and procedural results were documented.
Results: 59 patients with median age and bodyweight of 3.0 years (range 37 days – 75 years) and 13.8 kg (range 2.5 –80 kg) were treated. 37% of the cohort had a bodyweight ≤10 kg. 106 devices (30 Plug II, 76 Plug IV) were implanted in 93 target vessels. Major indications for their use were ductus arteriosus (19%), aorto-pulmonary collaterals (43%) and venous collaterals (34%) and miscellaneous vessels (4%). Complete primary vessel occlusion was achieved in 76% of these patients. In 5 curvy vessels with narrow retention zones, we accomplished vascular occlusion with the Plug II by using our novel delivery technique. No complications occurred.
Conclusion: Collateral-vessel closure via AMPLATZER(TM) Vascular Plug devices is well established, but device-delivery can be challenging in curvy vessels, especially in young children. In this context, we describe a promising, novel and simplified telescopic technique for easy controlled Plug II delivery.

P2-56
Lifetech Cera vs Amplatzer VSD occluders for transcatheter closure of ventricular septal defects

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Objectives: The Amplatzer and modified double-disc Lifetech Cera ventricular septal defect (VSD) occluders allow the transcatheter closure of the VSDs. The Amplatzer membraneous devices are not used any more because of increased complete atrioventricular block risk. Therefore, a comparison of these devices will show us the exact risk of the differences of the devices.
Methods: From May 2009 to November 2015, 128 consecutive patients (mean age 8.6 ± 4.2, range 1.4-26 year) underwent transcatheter closure of VSD. Used devices were Amplatzer membranous in 35 patients (27.3%), Amplatzer muscular in 32 (25%), Cera symmetric in 31 (24.2%), Cera muscular in 18 (14.1%), Cera asymetric in 4 (3.1%) patients. And also 8 patients took Amplatzer ductal occluder I or II.
Results: There were no differences in age, sex, defect type, shunt ratio, and pulmonary artery pressure between groups. Membranous defect ratio was 71%. Amplatzer device sizes (7.2 ± 2.1, range 4-16 mm) were bigger than Cera devices (6.2 ± 1.8, range 4-10 mm) ($p = 0.009$). Pacemaker implantation was performed temporary in 3 and permanently in 1 patient at Amplatzer group and none in Cera group. The follow-up period was statistically longer at the Amplatzer group (21.8 ± 16.6 vs 5.7 ± 6 months, $p < 0.001$). The other complication and residual flow and success rate were similar at both devices ($p > 0.05$). Membranous Amplatzer devices used in 72.9% of membranous defect but Cera symmetric and asymmetric devices used 94.5% of membranous defects ($p < 0.001$), because of increased block risk of Amplatzer devices.
Conclusion: Although success rate was similar, Cera devices compare favorably with lower complete block risk. And Cera membranous devices have more alternative with choosing symmetric, asymmetric and eccentric types.

P2-57
Intervention: Murphy's law in the cath lab: Tuning a disharmonic Melody into a harmonic pulmonary and tricuspid Concert - why we had a Christmas party

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Introduction: Synchronous transcatheter valve implantation in the pulmonary (PP) and tricuspid (TP) position is not an established approach and only performed on an individualized basis. Here we report on interventional maneuvers that were performed when after uneventful pre-stenting of the right ventricular outflow tract (RVOT) the Melody valve (MV) could not be placed in the desired PP and dislocated from the balloon of the 'Ensemble' delivery catheter.
Case Report: A 28-year old male patient with surgically corrected tetralogy of Fallot presented with signs of venous congestion and

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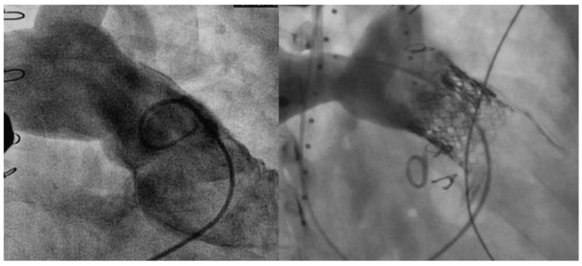
pre-syncope due to severe right heart failure caused by severe stenosis of the surgically implanted pulmonary homograft (24 mm) combined with an obstructed bio-prosthesis (27 mm Carpentier-Edwards) in the TP. Following uneventful preparation of the pulmonary 'landing zone' (placement of one covered, one uncovered CP-stent and one open-cell-designed Andra-stent) we were unable to advance the 'Ensemble' delivery catheter with the pre-mounted MV into PP. The following bailout maneuvers failed: I) partial retraction of the outer shell to enhance the mobility of the MV; II) re-dilation of implanted stents after gaining additional femoral venous access and bypassing the MV with a second guidewire, followed by attempting re-advancement of the MV into the stents; III) retraction of the MV to attempt implantation into the tricuspid position which failed due to dislocation of the MV from the balloon catheter IV) repositioning of the MV onto the balloon by a snare catheter. Implantation was finally achieved by the following technique: the partially funnel-shaped expanded MV was balloon floated within the lower part of the stented RVOT; under rapid pacing and a partially deflated delivery balloon the valved-stent was (via the second femoral vein access) additionally crossed by a coronary balloon catheter. The distal part of the funnel-like MV was gradually dilated by changing the balloons up to a 22mm Atlas catheter and by simultaneous retraction of the Ensemble-system. Finally, a second MV was uneventfully implanted in TP yielding 'two Melodies in harmonic concert'.

Conclusion: Endurance in attempting various bailout maneuvers during disharmonic percutaneous interventions may be rewarded by a harmonic result and a Christmas party.

P2-58
Percutaneous pulmonary valve replacement in the extraordinarily large pulmonary artery: Working at the technical limit

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Introduction: Percutaneous valve replacement in the extraordinary large native or patched right ventricular outflow tract and pulmonary artery remains a critical issue in interventional cardiology.
Case history: We report about a 32 years old female with corrected Tetralogy of Fallot by VSD closure and transannular patch plastic who was lost in follow-up. At the 27th week of gestation the patient presented with critical right heart failure due to severe pulmonary regurgitation and right heart volume overload. Under clinical monitoring pregnancy was continued until the 32nd week of gestation, delivery by caesarean section. After delivery progression of right ventricular dilation combined with left ventricular compression. Development of a progressive postcapillary pulmonary hypertension due to left ventricular diastolic filling disturbance. Surgical pulmonary valve replacement was negated due to severe biventricular failure and high mortality risk.
Angiography showed a severe pulmonary regurgitation and enlarged pulmonary artery (figure). The minimal diameter of the landing zone for PVR was 26mm x 26mm over a length of 10mm with dilation of up to 32mm before and after. After prestenting by two Andrastents crimped on 30 × 40mm Numed Z-med II balloon 29mm Edwards Sapien XT valve was be implanted transfemorally. Overdilation of the XT valve by 1 ml extra fluid in the inflator was used for better hold in the extraordinarily large pulmonary artery. Despite the overdilation Edwards Sapien XT valve function was excellent without regurgitation or stenosis.

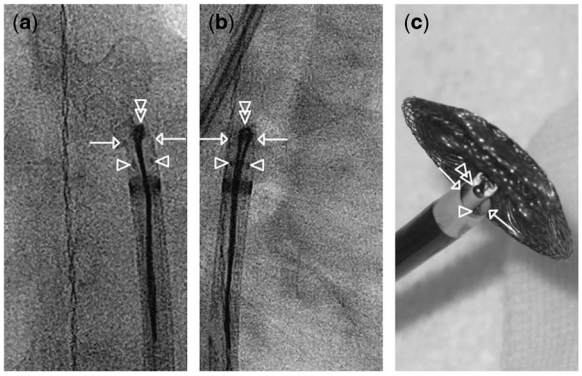


Conclusions: In individual cases a percutaneous valve implantation is possible even in an extraordinarily large and patched outflow tract. The critical limitation of this procedure is made by the diameter of available valves and balloons.

P2-59
Facilitating retrieval of embolized devices: Sheath-in-sheath-technique for device retrieval

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Introduction: Embolization of devices after deployment of the device is a known complication. Sometimes interventional retrieval of an embolized device can be a difficult task. Several techniques using different snares or other retrieval tools are known. Even if the device is perfectly snared at the tip of the device, re-entry of the device in the sheath is often difficult because the tip of the device might get stuck at the rim of the sheath due to insufficient centralization of the device by the snare. Usage of the dilatator as a guide for the snare wire is some cases helpful but has the disadvantage of a relatively stiff system, which can not easily be handled. We describe a novel method that combines full flexibility of almost any catheter as a guide for the snare wire and centralization of the caught device in the sheath.
Method: Retrospective case study.
Results: 15 hours after attempted closure of an ASD with a 10,5mm Figulla Flex ASD occluder (Occlutech) the device embolized to the transverse aortic arch. Due to the anatomy and the position of the device the tip was only successfully snared with a Multisnare wire (pfm) through a 5 F Amplatzer Right 3,5 catheter (Cordis). To achieve a centralization of the catheter with the snared device in the 10 F sheath (St. Jude) we used a second 7 F sheath (Terumo) in the first sheath. The device was retrieved without complications.



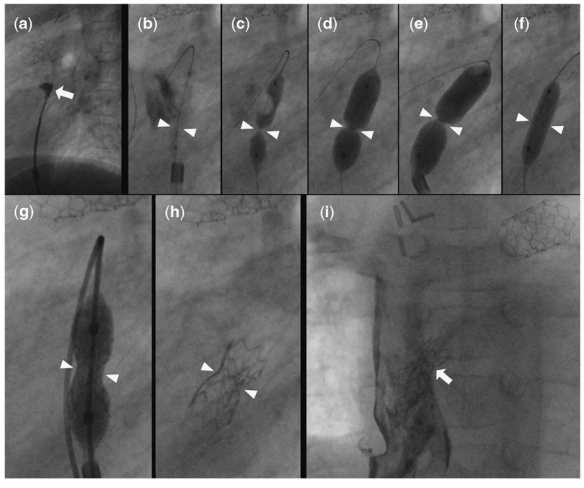
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The figure shows fluoroscopy images in a-p (a) and lateral (b) projection as well as the as the device retrieved with the sheath-in-sheath technique ex vivo. The arrows mark the rim of the outer 10 F sheath, the arrowheads mark the radiopaque marker of the inner sheath (a,b) and the tip of the inner sheath (c) respectively and the double-arrowheads mark the nicely centralized ball tip of the occluder.
Conclusion: The sheath-in-sheath-technique is a helpful way to combine the advantage of a flexible and easily handled catheter and centralization of the snare in the sheath for facilitated retrieval of embolized devices.

P2-60
Creation of a de-novo-fenestration by a cutting balloon technique in a child with failure of an extra-cardiac conduit Fontan circulation

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Introduction: In patients with a failing Fontan circulation based on an increased trans-pulmonary pressure gradient (TPG) at rest or exercise, de-novo creation of a communication between the Fontan conduit and the atrium is a possibility to increase cardiac output and decrease systemic venous pressures even if an extra-cardiac synthetic conduit was used for TCPC (total caval pulmonary anastomosis). However, in some patients this procedure remains challenging. We present a patient in whom a cutting balloon technique became necessary to dilate the conduit-atrial wall junction before a stent could be placed to maintain patency.
Method: Retrospective case study.
Results: In a child with plastic bronchitis and abnormal TPG as symptoms of a failing TCPC, transcatheter fenestration was performed. With mechanical force a trans-septal Brockenbrough needle was advanced through the extracardiac Fontan conduit (gelatin coated polyester) and the atrial wall to place a guide wire. A long sheath could not be advanced because of the rigidity of the material. A series of conventional balloon dilatations with high-pressure balloons did not achieve an increase in size of the novel communication. Thus a cutting balloon was used to successfully create a fenestration. To maintain patency finally a butterfly-shaped stent was implanted. No complications occurred.



The figure shows fluoroscopic images in lateral (a-f) or a-p projection (g-h). a) Puncture with the transseptal needle at the marked position in the Fontan conduit. b-e) Unsuccessful conventional balloon dilatation with balloons of increasing size (b: 3mm, c: 5mm, d: 6mm, e: 8mm) of the newly created communication between the Fontan conduit an the atrium. f) Successful cutting balloon dilatation (4mm). g) Placement of a pre-mounted Palmaz-Genesis stent (7mm balloon). h) Stent with the desired residual waist. i) Shunt from the Fontan-conduit into the atrium.
Conclusion: Cutting balloon dilation can be an option to achieve adequate sized fenestrations in failing Fontan patients when conventional balloon dilation fails, especially when rigid conduit materials were used.

P2-61
Predictors of outcome of transcatheter balloon dilatation of valvular pulmonary stenosis in neonates and infants

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Background: Balloon pulmonary valvuloplasty (BPV) is the treatment of choice for patients with pulmonary valve stenosis (PS).
Methods: An observational cross sectional study including neonates and small infants underwent BPV in the period from 2004-2013 in Cardiac Cath Unit- Pediatric Cardiology Department- Cairo University. Multivariable models were built to report the predictors of outcome of balloon pulmonary valvuloplasty and its complications both during and early after the intervention.
Results: A total of 644 cases were included in the study . They were devided according to age into two groups :neonatal and early infancy ($n = 282$, mean age 26.7 ± 6.9 days, BW 4.7 ± 1.4 Kg), infancy ($n = 362$, mean age 6.8 ± 2.7 months, BW 7.3 ± 1.3 Kg). Procedural success was achieved in 82.7% of the patients, being defined as a drop of right ventricular systolic pressure to less than or equal to 50% of the baseline measurements.
The significant predictors of outcome were: The pulmonary valve annulus measured by angio (successful 8.8 ± 2 mm, unsuccessful 8.2 ± 1 mm, Pvalue $= 0.015$), RV pressure and pressure gradient post BPV ($P = 0.000$, $P = 0.000$), the presence of infundibular stenosis ($P = 0.006$),and the balloon size (successful 10.9 ± 2.7 mm, unsuccessful 10.1 ± 2.5 mm, $P = 0.05$). The balloon/annulus ratio was not different between the successful and unsuccessful group ($P = 0.5$). We studied also the impact of age, BW, RVP before balloon diltation(Critical PS), presence of RVD and the morphology of the pulmonary valve (dysplastic valve) on the results of BPV and no statistically significant difference of these values was found between successful and unsuccessful cases.
*Conclusion:*The PV annulus size, balloon size, absence of infundibular stenosis are strongly correlated to successfullness of BPV ($P < 0.05$).

	SUCCESS						T-test	
	No			Yes				
	Mean	±	SD	Mean	±	SD	t	P-value
Age in days	133.625	±	96.090	134.992	±	87.674	-0.104	0.918
WT	5.878	±	1.734	6.295	±	1.857	-1.702	0.092
PG1	94.250	±	26.421	91.211	±	25.226	0.832	0.408
PVannulus	8.220	±	1.848	8.872	±	2.007	-2.487	0.015*
RVP1	115.906	±	34.890	113.264	±	28.401	0.562	0.576
PAP1	21.734	±	6.496	23.326	±	9.142	-1.608	0.110
RVP2	80.032	±	30.303	50.632	±	12.805	7.482	0.000*
PG2	53.787	±	26.481	24.241	±	9.889	8.575	0.000*
BALL1 S	10.180	±	2.553	10.908	±	2.779	-1.970	0.05*
Ball. Ratio	1.266	±	0.355	1.299	±	0.341	-0.662	0.510

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P2-62

Initial Experience with the Nit-Occlud ASD-R: Short-Term Results with Emphasis on Causes of Device-Related Cardiac Rupture

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Objective: The Nit–Occlud® ASD-R (NOASD-R) is a new device with unique characteristics in terms of the device structure and delivery system. The device comprises two equal-sized circular discs linked by a short waist corresponding to the diameter of the defect. Reported data on the NOASD-R are limited. We evaluated patients who underwent cardiac catheterization with the aim of the device closure of ASD using NOASD-R to assess the safety and efficacy of the device.
Methods: Between 2014 and 2015, transcatheter closure of ASD using the NOASD-R was performed in 30 patients. If standard left upper pulmonary vein (LUPV) deployment technique was unsuccessful as the device not aligned perpendicularly to the septum, right upper pulmonary vein (RUPV) approach was used.
Results: The mean age was 10.9 ± 12.8 (3.5–60, median 6) years and mean weight was 28.2 ± 18 (14–79) kg. 2-D diameter of the defect was 13.7 ± 3.1 (10–22.0) mm and colour flow diameter was 16 ± 3.6 (11.2–26.3) mm. The nature of the defect was complex in seven patients as multiple defects in two, floppy-mobile posterior-inferior rim in two, and deficient posterior-inferior rim in three (< 5 mm). Implantation was successful in all patients. The standard LUPV approach was used in 25 whereas RUPV approach was required in 5. The mean size of devices was 17.1 ± 3.3 mm (12.0–26.0 mm). The mean device size/2D defect diameter ratio was 1.26 ± 0.09 (1.12–1.40). The mean device size/color flow diameter ratio was 1.07 ± 0.06 (1.0–1.22). Releasing problem was encountered in three. Device-related erosion on the day after the closure was observed in one. No embolization, late erosion or dislodgement of the device was encountered during or after implantation. Complete occlusion has occurred in all at a median 10 month follow up.
Conclusion: NOASD-R is a feasible and effective device for transcatheter occlusion of moderate to large secundum ASDs in selected patients. The occurrence of the erosion on the right atrial roof may be due to the high localization of the defect, and the larger size of the right disc.

P2-63

Transcatheter Treatment of Middle Aortic Syndrome (MAS) with Bare and Covered Stent Implantations

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Introduction: Middle aortic syndrome (MAS) is an uncommon cause of arterial hypertension in children and young adults characterized by long segment narrowing of the distal thoracic and/or abdominal aorta. Stenosis of the abdominal aorta may be associated with stenoses of renal and visceral arteries.
Method: Between 2012 and 2015, six patients underwent stent implantation for the treatment of with MAS. In patients with severe sub-atretic stenosis, predilation with smaller sized balloons was required before stent implantations. Balloon size that stents will mount was selected according the diameter of distal aorta (1-2 mm smaller). When first stent does not cover the lesion completely additional stents were implanted by telescopic method. If the lesion is close to the critical vessels bare stents if not covered stents were implanted. After implantation further dilation was performed to optimum size in the same session or subsequent session.
Results: Median age was 15.5 years (8–22 years). None had inflammatory signs of Takayasu arteritis but one had neurofibromatosis and the other had Williams syndrome. Length of the stenosis varied between 19 mm and 105 mm (median 64) and median diameter of the lesion was 3.3 mm (1.5-5.4). Aortic narrowing was isolated in five and coexisted with left renal artery stenosis in one. 4 covered stents were required for long segment subatretic lesion in one, two stents in three and single in two. Covered stents were used in two, both bare and covered stents in one and only bare stents in tree. One had two intervention stages; cutting balloon suboptimal dilation in the first session than bare stents was implanted in the second. Balloon angioplasty for unilateral renal artery stenosis was performed in patient with neurofibromatosis. There was no procedural complication. Redilation was needed due to suboptimal dilation in the first session in one and antihypertensive medication was continued in all.
Conclusion: Transcatheter treatment of long segment middle aortic syndrome is an effective and safe option with excellent results. It improves both vessel diameter and pressure gradient by using multiple stents with telescopic method. Staged dilation may be preferred in some situations.

P2-64

Percutaneous dilation of pulmonary arteries in the patients after Norwood procedure- its utility before the Fontan completion

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Introduction: In a significant number of patients after the Norwood procedure there is observed hypoplasia of the left pulmonary artery, It is the reason of pulmonary hipoperfusion, development of collaterals and elevation of pulmonary pressure in the contralateraly branch.
Material and methods: During last 5 years there were 48 hospitalized children after the Norwood procedure and hemiFontan/Glenn operation. The pulmonary hipoplasia with hipoperfusion were confirmed in 28 patients aged 31.8 ± 12.3 months. They were catheterized with intention of percutaneous dilation of pulmonary arteries (21.7 ± v8,5 months after hemi Fontan/Glenn procedure).
Results: The average diameter of the pulmonary stenosis was 3,5 ± 1,4mm. 30 stents (Genesis- 17, Formula- 7, Valeo- 6) were implanted successfully. The diameter of stenotic vassel increased to 8,4 ± 2,0mm, McGoon index increased from 1,3 ± 0,3 to 1,9 ± 0,4. The pulmonary pressure in the right pulmonary artery decreased from 16.2 ± 2.5 to 15.2 ± 1,8 mmHg. The balloon predilation was performed in 3 patients: in 2 patients it was completly ineffective, and in 1 patient- the dissection of the vessel wall appeared. The stents were implanted subsequently. Simultaneous additional percutaneous procedures were performed in 21 patients: angioplasty of the connection of neoaorta with aorta in 12, stent implantaion in 3, percutaneous closure of collateral in 11 and dilation of interatrial defect in 1 pt.
During follow-up, 7 patients required additional stents implantation (Genesis-2, Formula-3, Valeo-2): in 4-due to the appearance

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of the peripheral pulmonary stenosis, in 1-due to the rupture of the previously implanted stent and in 2 due to stenosis in stent. The Fontan operation was performed in 16 pts, 6 pts have good conditions (after catheterization) and are waiting for this operation and 5 pts were disqualified. There were 6 deaths: 4 in short time after Fontan operation, 1-1,5 year after Fontan operation due to cerebral stoke and 1 after heart transplantation.
Conclusion: Hypoplasia of the pulmonary arteries is a common problem that occurs after Norwood operation. The percutaneous implantation of stents is effective treatment of this complication in majority of patients with this problem.

P2-65

The ability of percutaneous closure of ventricle-pulmonary connections in the setting of cavopulmonary shunt or after the Fontan procedure: our experience

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Background: Low pulmonary pressure and resistance is crucial for the proper function of the Fontan circulation. Maintained anterograde pulmonary blood flow (through pulmonary artery banding (PAB) or recanalization of surgically sutured pulmonary trunk) can cause elevated mean pulmonary artery blood pressure (MPAP) and volume overload of systematic ventricle in the setting of cavopulmonary shunt or after the Fontan procedure.
Objective: To study the utility of occluder devices in ventricle-pulmonary connections.
Material and methods: 5 patients (3pts after Glenn procedure, including 2pts with elevated MPAP, respectively 22 and 19 mmHg; 2pts after the Fontan operation, including 1 pt with failing Fontan circulation), in the age group 2-19 yrs, were treated percutaneously with occluder devices for ventricle-pulmonary connections. We used different types of occluder devices: CP cover stent (n = 1), Amplatzer Muscular VSD Occluder (MuscVSD) (n = 1), Amplatzer Duct Occluder II (ADOII) (n = 2), Amplatzer Septal Occluder (ASO) (n = 1).
Results: Procedural success (with no residual shunt) was achieved in all patients. Although in one case there was a need for implantation of an additional stent and in another case we had to retrieved MuscVSD and deployed ADOII due to its unsatisfactory position. We observed improvement and resolution of pleural effusion in the patient with failing Fontan circulation. One patient with cavopulmonary shunt and elevated MPAP six months later was qualified and scheduled for completion of Fontan circulation due to normalisation of MPAP.
Complications: Two patients had thrombus formation in pulmonary trunk proximally to the device. In one case thrombus formation was detected immediately after the procedure and it resolved with warfarin therapy without complications. Second patient, who was lost in follow up for 10 months, was admitted to our institution with thrombus and cerebral stroke.
Summary: Percutaneous closure of ventricle-pulmonary connections is technically possible. This intervention should be considered in patients with cavopulmonary shunt or Fontan circulation with elevated MPAP and volume overload of systematic ventricle. The use of appropriate anticoagulation is crucial for success.

P2-66

Two cases of successful percutaneous coarctation of aorta repair in pregnant women

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Introduction: The native severe coarctation is a condition in which pregnancy risk – WHO IV, what means – pregnancy contra-indicated. The secondary hypertension during pregnancy is present in 5 to 10% of cases and must be considered in presence of drug resistance. Also, the hypertension is a frequent medical complication during pregnancy (normal pregnancy – preeclampsia 11% in population). Management of hypertension during pregnancy is challenging due to the fetal toxic impact of some drugs, namely ACE inhibitors and ABRs. Treatment of the arterial hypertension is mandatory when the blood pressure is higher than 170/90 mmHg. Although, we should bear in mind, that the aggressive treatment of arterial hypertension must be avoided in the patient with coarctation of aorta to prevent the placental hypoperfusion. To our best knowledge no one case of aortic coarctation repair during pregnancy has been described till nowadays. We are presenting two cases of successful stenting of the aorta during pregnancy, because of the uncontrollable hypertension.
Case 1: A 17-years old woman admitted in her 24-th week of gestation with uncontrollable hypertension. Mid-aortic syndrome was diagnosed for the first time. Severe abdominal coarctation was repaired with CP covered stent. After the procedure the blood pressure decreased from 220/100 mmHg to 140/90 mmHg.The transcatheter gradient decreased from 110 mmHg to 30 mmHg. The spontaneous vaginal delivery occurred in 39 weeks of pregnancy, the healthy male newborn was born.
Case 2: A 28 years old women who had coarctation of aorta repaired in a childhood admitted in her 19-th week of pregnancy with arterial hypertension resistant to medication. Echo data showed hypoplastic transverse arch with the gradiant accross it 70 mmHg. She had transverse arch stenting with Andrastent. The residual invasive gradient was 20 mmHg. The patient did not required any medication during all pregnancy and had uneventful spontaneous vaginal delivery.
Conclusion: Percutaneous intervention for undiagnosed coarctation or re-CoA is possible during pregnancy and should only be performed if severe hypertension persists despite maximal medical therapy and there is the evidence of maternal or fetal compromise
Key words: coarctation of aorta, pregnancy, arterial hypertension.

P2-67

Pfm-Le VSD coils in closing various challenging ventricular septal defects in children

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Interventional VSD closing allows for avoiding surgery, being indicated when operative access is difficult and high risk-associated. The implant type selection depends on VSD location, size, number and interventional team experience. One of the methods used to close various types of VSD is Nit-occlud Spiral Coil system. The objective of the report is presentation of our experience with this system in various VSD types. The

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material consisted of 18 (10 F/8 M) patients aged 2-18 years, x-8 years, body mass 10-64 kg, x-28.2 kg, qualified for intervention after preliminary echocardiographic evaluation. There were 10 pts with perimembranous VSD (pmVSD), 4 with midmuscular VSD (mVSD), including 1 pt with multiple muscular VSDs after a previous unsuccessful PAB, and 4 pts with acquired LV-to-RA shunts as a complication of previous cardiac operations. The implant location, size and number were determined based on angiocardiography results. The procedure was in keeping with the manufacturer protocol.
Results: In 9/10 pmVSD pts and in all children with LV-to-RA shunts, the procedures were successful without complications. In 1 pt with 8 mm-size pmVSD, a spontaneous coil migration to LPA occurred, necessitating surgical removal and surgical VSD closure. In one pt with complex CHD, 2 mVSDs were successfully closed, but the patient died 3 months later for reasons unrelated to the procedure. In 1 pt with multiple mVSD after PAB, seven VSD's were closed in three consecutive sessions at 36, 38 and 48 months of age, with a total of 7 coils (1st session - 2 coils: 8 × 6mm, 2nd – 2 coils 8 × 6 and 10 × 6mm, 3rd-3 coils: 10 × 6mm). In follow-up after 26 months, this patient still had one insignificant residual shunt (< 1.5 mm). The LV systolic function was normal but prolonged IVRT (0.09') and abnormal MPI-0.51 were noted.
Conclusions: Pfm-Le VSD coil system is effective, especially in closing atypical, numerous and surgically difficult to access VSDs. Implant plasticity ensures its effectiveness and prevents significant interventricular septal distortion. It also provides an interventional option in treatment of specific defects, such as LV-RA shunts.

P2-68

Transcatheter Closure of Perimembranous Ventricular Septal Defects with Nit Occluds Leˆ VSD Coil® : Early and Mid-Term Results of a Multicentre Study

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Background and Aim: Transcatheter closure of perimembraneous VSD's is still a challenging procedure in interventional cardiology. Technical difficulties and concerns about possibility of the high percentage of permanent AV block with the double disk designed devices leads to investigate new device designs for percutaneous closure of perimembraneous VSD's. PFM Nit Occlud Le VSD coil ® has a completely different design and closure principle and has been using in the closure of perimembraneous VSD's for years. The data about this device is still limited in literature. We report the outcomes of multicenter retrospective study of PFM Nit Oclud Le VSD coil ®.
Patients and Methods: Between October 2011 and November 2015 82 patients with perimembraneous VSD enrolled the study. The data of four centers (two from Turkey, two from Egypt) retrospectively reviewed.
Results: The mean age of the patients was 5,5 ± 4.2 years (1–19 years), mean weight was 20.5 ± 12.1 kg (8 to 75 kg). Implantation was performed successfully in 81/82 patients (98.7%). In one patient procedure abandoned due to severe tricuspid valve stenosis. VSD size at the left ventricular angiogram 8.2 ± 1.9 mm (4.6-14.6). 70 of patients had ventricular septal aneurism. Five patients had aortic valve prolapses into the defect. The mean value of the Qp/Qs ratio was 1.9 ± 0.4. The median procedure and fluoroscopy time were 72.8 minutes (35–180 min.) and 27 minutes (13.3–67.4 min.) respectively. Immediate total occlusion rate was 67/82 (81%). Complete closure occurred in 54/59 patients (91%) after 6 months follow up. Intravascular hemolysis developed after the procedure in 6 (7,3%) patients. In one patient, mild aortic regurgitation was seen but it did not progress during follow up. In one case moderate tricuspid valvular regurgitation developed. There were no rhythm problems and embolization during the follow-up period of 35.3 ± 6.6 months. No deaths occurred.
Conclusion: The Nit-Occlud Leˆ VSD coil device is a good alternative for the transcatheter closure of perimembraneous VSD's in selected cases. As an advantage,there was no development of a permanent atrioventricular block. Patients with residual shunt should be monitored closely for the development of hemolysis.

P2-69

Percutaneous PDA stenting in newborns with duct dependent pulmonary circulation and Univentricular Heart Physiology

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Background: Surgically Creation of a aortopulmonary shunt is still high mortality and morbidity in newborns. Percutaneous stenting of ductus arteriosus is an alternative way in order to supply blood to the pulmonary arteries in duct dependent circulations. Here we report single center experinces with ductus stenting and investigate the efficacy of procedure in newborns with univentricular heart physiology.
Patients and Methods: Between 2014 july and 2015 November, 52 procedures were performed in 46 patients. Fourty of patients had univentricular heart physiology. In 47 of 52 (90%) stents were implanted sucessfully. Procedure related mortality occured in 2 (% 3,8) patients. Mean follow up duration 134.5 months. Reintervention need due to early stent restenosis in three patients (%6). Seven patients (7/46, 15%) died during interstage period. One patient died after Glenn operation. 15 of patients with univentricular heart physiology still waiting for Glenn operation. In 17 (%42) patients bidirectional cava pulmonary anastomosi was performed succesfully.
Conclusion: Percutaneous stent implantation in ductus arteriosus less invasive, feasible method. Early mortality is lower than the surgical aortopulmonary shunt. However interstage mortality rate is still high and patients should be monitored closely.

P2-70

Longer and wider tunnels may warrant primary choice of transseptal puncture in percutaneous closure of patent foramen ovale

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Introduction: Percutaneous closure of patent foramen ovale (PFO) is routinely performed for secondary prevention of stroke. Rarely, transseptal puncture (TSP) has been described to tackle challenging morphologies that may lead to incomplete closure, and is usually used after failure of the standard approach. We report our

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experience in percutaneous closure of PFO with TSP, and analyze variables that may predict its need as a primary technical approach.
Methods: We reviewed our institution's cases of percutaneous PFO closure with TSP since January 2011, and compared them with a group of random controls (ratio 2:1) who underwent percutaneous PFO closure with the standard technique. We analyzed relevant anatomic variables of the interatrial septum assessed by transesophageal echocardiography (size and presence of aneurysm; width of primum and secundum septa); PFO anatomy (tunnel length and width, baseline and per-balloon interrogation; spontaneous shunt) and presence of large Eustachian valve.
Results: We report six cases of percutaneous closure of PFO with TSP [mean age 45 (range 31-62) years; 3/6 males] and twelve controls (non-significantly different demographics). All cases with TSP had a previously failed attempt with the standard technique, due to device malposition, residual shunt or technical difficulties. Comparison between the two groups revealed a significantly longer tunnel [(mean 18.5; range 15.5-22.4 vs. mean 9, range 5-14.5mm], $P < 0.01$) and larger baseline width (mean 3.6; range 2.7-5.5 vs. mean 1.5, range 0.5-2.5mm, $P < 0.01$) in the TSP group. All tunnels longer than 15mm or wider than 2.6mm required TSP. Baseline left-to-right shunt was present in 6/6 cases and in 7/12 controls, but failed to achieve statistical significance ($P = 0.06$). No other differences were found between the sample and control groups, namely the size or presence of atrial septal aneurysm, procedural efficacy or complications.
Conclusion: In our sample, a larger length and width of the PFO tunnel was associated with the need for percutaneous closure of PFO with TSP. Our study, although limited, suggests that pre-closure transoesophageal evaluation of the anatomy of PFO may warrant selective TSP as a first choice in percutaneous PFO closure.

P2-71
Two-centre experience with the Valeo stent in the treatment of pulmonary artery stenosis

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Introduction: In infants and small children stent implantation for pulmonary artery (PA) stenosis may be challenging and require re-interventions to match the patient's growth. Until recently, the lack of stents, dilatable to adult sizes, has limited the more widespread application of this therapy in the youngest patients. We evaluated the implantation and mid-term performance of the Valeo stent, which can be dilated up to 20 mm, for the treatment of PA stenosis.
Methods: A retrospective analysis of Valeo stents implanted for PA stenosis at two large volume paediatric centres was performed. Patient profile, pre- and post-implant catheterization data, follow-up imaging and re-interventions were reviewed.
Results: Between 11/2012 and 12/2015, 51 patients received 56 Valeo stents. The median age was 4.9 years (7 months - 16 years) and median weight was 14.7 kg (5.7-53 kg). There were 31 patients (61%) with single ventricle physiology and 20 patients (39%) with biventricular circulation. Twenty seven patients (53%) weighed less than 15 kg, including 12 patients (23%) weighing less than 10 kg. Nine patients (18%) had the stent implanted within 30 days post-surgery. In 10 patients (20%) the Valeo stent was implanted in a previously placed stent. Improvement in PA diameters and hemodynamic values was achieved across all patient subgroups. Complications occurred in 4 patients (8%); including stent embolization (2), stent dislodgment from the balloon during delivery (1) and hemodynamic instability (1). In the median follow-up of 17 months (2-42) one patient died and one was lost from follow-up. Of the remaining 49 patients, 7 (14%) underwent catheter re-intervention after a median of 15.9 months (1 day - 20.7 months) due to significant neointimal proliferation (6) and early stent deformation (1). In 25 patients (51%) the stent was visualized either on chest X-ray (23) and/or in fluoroscopy (8). Stent distortion was noted in 8 patients (32%).
Conclusions: Based on our results the Valeo stent provided effective relief of PA stenosis in various clinical settings including low body weight, early post-operative course or stenosis within a previously placed stent. Although stent deformation was commonly observed on follow-up imaging, pronounced neointimal proliferation was the most common indication for re-intervention.

P2-72
Early experience with a novel image fusion software for 3D guidance of complex trans-catheter interventions.

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Introduction: Recent improvements in the development of fusion imaging software have led to the introduction of a 3D roadmap based on preregistered Computed Tomography (CT) or Magnetic Resonance (MR) datasets for live guidance of trans-catheter interventions. Currently there is limited experience available with 3D fusion software, coming predominantly from aortic and peripheral interventions in adults. Despite these limitations the results show promising reduction of contrast and radiation exposure along with shorter procedural times. We describe our initial experience with recently available VesselNavigator (Phillips) for live guidance of trans-catheter interventions in congenital heart defects.
Methods: We performed a retrospective review of all trans-catheter interventions guided with VesselNavigator. Patient characteristics and catheterization data were reviewed with focus on segmentation, fusion and intervention guidance.
Results: Between November and December 2015, 4 patients underwent trans-catheter interventions with VesselNavigator guidance. The median age was 9.7 years (2 weeks – 14 years) and median weight was 32.5 kg (3.5–46 kg). The interventions included stent implantation in aortic coarctation, patent arterial duct and right pulmonary artery and pulmonary valve placement. Existing CT datasets were used to create a 3D roadmap in all patients. The target structures were easily selected with one-click segmentation and additional ring markers were utilized in 3 patients. For registration and fusion of the overlay, fluoroscopy images were acquired in posterior-anterior and lateral projections with spine and vertebrae (4 patients), calcifications (3) or contrast injection (2) serving as reference points for orientation of the 3D roadmap against live fluoroscopy. Accurate overlay was achieved in all patients and enabled successful interventions without additional contrast injections prior to stent placement.
Conclusions: In our early experience, VesselNavigator proved to be useful in guidance of versatile complex trans-catheter interventions. Intuitive segmentation and easy fusion with live fluoroscopy allowed shortening of the diagnostic phase of the procedure and reliable 3D roadmap facilitated interventional treatment.

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P2-73

Percutaneous Intervention by Stent Implantation in Aortic Coarctation in Children Under 30 kg

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Objective: There is vast experience in percutaneous treatment of coarctation of the aorta (CoA) in larger patients.
Our aim was to describe results and evolution in a younger population under 30 kg with (CoA) and recoarctation treated by stent implantation.
Methods: Retrospective review of all patients with native coarctation and recoarctation treated percutaneously by stent implantation in our centre between 2004 and 2015. Patients were divided in two groups according to weight. Groups were compared in treatment effectiveness, complications and need for reintervention.
Results: Fifty-three patients were included in our study, of these, 19 were less than 30 kg (group I) compared to the other 34 (group II). Median time of follow up was similar in both groups (5.1 Vs 5.7 y). Median age of group I was 5.2 years old, range [0.02 to 8.1 y] and median weight 17 kg, range [3.5 to 27.4 kg]. Coarctation diameter was standardized with the descending aortic diameter (ratio CoA/Dao). No significant differences were found between these two groups in ratio CoA/Dao pre- and post- stent implantation. The mean minimum diameters of CoA (group I 4.2 to 9.1 mm; group II 7.1 to 12.4 mm) and the ratio CoA/Dao (group I 0.39 to 0.85; group II 0.45 to 0.76) increased significantly in both groups (all $p < 0.005$). Furthermore, no differences in residual systolic gradient were found between the two groups. Large redilatable stents (CP stents) were used in all cases except in two patients in group I where premounted stents were implanted as salvage therapy. Complications were: one aortic aneurism in each group resolved with covered stent implantation and one stent migration to the thoracic aorta in group II. No patient in either groups required further surgery related to stent implantation. No significant differences were found in percutaneous reintervention (38,5% Vs 32,3%) and neither in time until first stent redilatation (3.5 Vs 2.5 y).
Conclusions: Stent implantation for coarctation treatment seems a successful and safe procedure even in younger patients with lower weights. Nevertheless long-term outcomes of stent implantation in these small patients with CoA are still needed.

P2-74

Serum Nickel And Titanium Levels After Transcatheter Closure Of Atrial Septal Defects With Amplatzer Septal Occluder

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Introduction: The Amplatzer septal occluder (AGA Medical Corporation, Golden Valley, USA) is one of the most frequently used device for transcatheter closure of secundum-type atrial septal defects (ASDs). The Amplatzer septal occluder consists of nitinol wire mesh. Nitinol, an alloy composed of 55% nickel and 45% titanium. It has made ASDs closure safe, easy and successful. However, there is a concern about release of nickel and titanium after implantation of nitinol devices.
Objective: To evaluate serum nickel and titanium levels after transcatheter closure of ASDs with Amplatzer septal occluder.
Methods: The study registered 38 pediatric patients with ASDs and no history of nickel sensitivity have undergone percutaneous closure of ASDs with the Amplatzer septal occluder. Blood samples were drawn 24 hours before and 24 hours, 1, 3, 6 and 12 months after occluder implantation. The serum nickel and titanium concentrations were measured in each sample by atomic absorption spectrophotometry. Serum levels of nickel <2 ng/mL were considered to be normal.
Results: the mean patient age was 6.7 ± 3.72 years and weight 23.24 ± 13.37 kg. The mean size of the implanted occluder was 14.0 ± 4.66 mm. Before occluder implantation, these levels were 0.56 ng/mL. They increased to 1.28 ng/mL 24 hours after implantation and reached maximum levels one month after implantation, with a mean of 1.96 ng/mL. There was a decline to a mean of 1.75 ng/mL 3 months after implantation, to 1.22 ng/mL 6 months after implantation and to 0.66 ng/mL 12 months after implantation. Statistical evaluation with the Wilcoxon test showed that the rise in serum levels of nickel 24 hours, 1 and 3 months after implantation was significant. No allergic or toxic reactions were observed during follow-up. No patients showed a detectable serum titanium level.
Discussion: Our study shows that nickel is released from the Amplatzer septal occluder in the first few months after implantation. Conversely of this, no patients showed a detectable serum titanium level. Although nickel may cause toxic and allergic responses in patients who have nickel allergy, nitinol devices such as Amplatzer septal occluder can be use for transcatheter closure of ASDs.

P2-75

Single center experience: retrieval maneuvers of embolized ASD and PFO devices

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Device embolization after interventional ASD/PFO closure is a rare incident. The degree of difficulty in device retrieval varies with the device design. The aim of the study was to report our experience in retrieval maneuvers.
Method: Retrospective single center study of all patients after catheter interventional ASD or PFO closure (n = 1370).
Results: The embolization rate was 0.7% (10/1370; Amplatzer n = 5, Occlutech n = 3 Cardio Seal n = 1, Helex n = 1). From July 1996 until December 2015 twelve different types of ASD or PFO devices were used (Amplatzer n = 1019, Occlutec n = 101, Helex n = 56, Cardio Seal n = 55, GSO n = 54, Starflex n = 35, Premere n = 23, Ceraflex n = 9, Angel Wings n = 7, Solysafe n = 4, Cardia Star n = 3, Rashkind n = 2, PFM n = 2). Two devices had to be removed surgically (Amplatzer n = 1, Occlutech n = 1). All other embolized devices were retrieved by catheter interventional means. In a bench test a 26 mm Occlutec ASD device was not suitable for snare retrieval, even though a 16 F sheath. Different sized snares slipped off the occlude-hub. Retrieval of this Occlutec device was possible by catching it with the grasping forceps of its delivery cable. This technique was used successfully in the next embolized occlutech occluder.
Conclusion: The flat profile and globular hub of the Occlutec Figulla device causes difficuluties to retrieve an embolized device with current snares.

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P2-76

Treatment of PAIVS: what is the best option?

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Background: Innovations in knowledge and techniques have improved survival of pts with PAIVS. Despite this, it is still difficult to define the best treatment for those pts which are 'within the spectrum'. Methods: The outcome of 24 newborns with PAIVS (prenatal diagnosis n = 13, postnatal n = 11) was reviewed.
Results: The mean age and weight at the observation were 2.5+ 2.4 days and 3.3+ 0.8 kg. The mean Z-Score value of the TV annulus was -3.01+ 1.36. We detected a severe hypoplastic RV (6/24, 25.0%), bipartite RV (8/24, 33.3%) or a hypertrophic tripartite RV in 10/24 (41.6%) pts.
Two associated lesions: Ebstein anomaly in one and mitral valve anomaly in another. In one pt we found RV-dependent coronary circulation. Catheter RF was performed in 18/24 pts (72.0%) with favourable anatomy (mean TV Z-score -2.7). Pts with unfavourable anatomy, 1st intervention was MBT shunt in 3 pts (3/24, 12.0%), and PDA stenting in 3 pts (3/24, 12.0%). Perforation was successful in 16 patients (16/18, 88.8%, 1 pt submitted Brock valvotomy and in 1 with pulmonary artery perforation and cardiac tamponade needing neonatal surgery). No deaths. During the first month all pts submitted to RF needed further pulmonary flow. In 13/18 (72.2%) patients ductal stenting and 5/18 pts (27.7%) required a MBTs placement. At a median FU of 6.2 years (range 1.2 to 13.1), among 18 pts underwent RF procedure, 5/18 (27.7%) have a univentricular connection and 13/18 (72.2%) have a biventricular pathway. At FU, in our 24 APSI pts, 9/24 (37.5%) pts have a univentricular connection (3 pts with partial cavo-pulmonary anastomosis and 6 pts Fontan completion), 15 (62.5%) pts got a biventricular correction (2 coil embolization of MBTs with no further intervention, 5 needed RVOT surgery, 3 pts still have PDA flow, and 5 no procedure after MBT or stent spontaneous closure).
Conclusion: detection of RV anatomical type and the related-Z Score value of the TV is a predictive index for surgical or percutaneous intervention. Catheter RF is feasible, with low mortality, even if all pts need a pulmonary blood flow after the procedure.

P2-77

Effect of percutaneous stent implantation on arterial hypertension and aortic flow dynamics in patients with aortic coarctation: identification of responders and non responders

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Introduction: Arterial hypertension (HT) is a common complication after successfully repaired aortic coarctation and involves complex mechanisms. Moderate to severe recoarctation or late diagnosed native coarctation are accessible to endovascular treatment but the effect of aortic stenting on HT is variable.
We sought to identify the factors contributing to persistent abnormal aortic flow dynamics and hypertension despite optimal endovascular treatment.
Methods: 30 consecutive patients (median age 18.5 years, range 9 to 58 years, 76.7% male) who underwent aortic stenting for coarctation and HT (17 native, 13 recoarctations) between 2007 and 2015 were retrospectively enrolled. Optimal treatment was defined by hemodynamic peak-to-peak gradient of less than 10 mmHg after stent implantation. HT was defined according to current recommendations. Aortic arch shape (gothic, crenel or normal) and aortic arch hypoplasia were identified. All patients underwent exercise echocardiography and ambulatory blood pressure monitoring with QKD assessment 6months after the procedure. Multivariate analysis was performed.
Results: Arm-leg pressure gradient (28.5 ± 15.5 vs 8 ± 8 mmHg, $p < 0.01$), systolic arterial pressure (142 ± 35 mmHg vs 120.5 ± 22.5 mmHg, $p < 0.01$), hemodynamic peak-to-peak gradient (33.5 ± 22.5 mmHg vs 9 ± 9 mmHg, $p < 0.01$), aortic isthmus maximal velocity at rest (3.1 ± 1.5 m/s vs 2.4 ± 1.2 m/s, $p < 0.01$) and at exercise (5.6 ± 0.9 m/s vs 3.9 ± 2.1 m/s, $p < 0.01$) were significantly decreased after optimal endovascular treatment. 14 patients had persistant HT (46.7%) and 12 had exercise-induced HT (40%). Persistent HT at rest was associated with higher body mass index ($p = 0.043$), smoking ($p = 0.043$), and the length of the implanted stent ($p = 0.017$). Normal shape of the aortic arch was associated with the absence of persistent HT ($p = 0.029$).
Exercise-Induced HT was associated with aortic arch hypoplasia ($p = 0.01$). Arterial distensibility assessed by QKD measurement seems to be lower in hypertension patients but not statistically significant ($p = 0.054$).
Conclusions: Identifying clinical, hemodynamic and anatomic parameters associated with residual hypertension can help to define the non responders after stent implantation. The QKD is a non invasive useful tool in the follow up of corrected aortic coarctation.

P2-78

Ostium secundum atrial septal defect percutaneous closure in children: is it always a simple procedure?

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Introduction: Percutaneous closure of atrial septal defect (ASD) has become the first line therapeutic option. We aimed to investigate the influence of anatomic parameters on the procedure progress and the outcome as well as the impact of echocardiographic guidance.
Methods: We retrospectively included 91 consecutive children aged 8.1 y.o [6.4-11.9, min 9 months] weighting 26 kgs [21-39; min. 6] who underwent percutaneous closure of ASD in a single children center. 2D-TEE guidance and balloon calibration were performed in all-cases and 3D-TEE in 71 patients > 15 kg. Complex ASD (n = 73.6%) were defined in cases of multiple holes (n = 14, 15.4%), large defect with ASD diameter > 15 mm/m^2 Body Surface Area (n = 62, 68.1%) and deficient rims other than the retro-aortic one (n = 4, 4.4%).
Results: Disagreement between balloon diameter and ASD diameter assessed by 3D (n = 71) or 2D (n = 15) echocardiography was higher in complex ASD. An absolute difference above 2 mm was observed in 61.2% versus 20.3% ($p = 0.001$). Percutaneous closure was more difficult to achieve in complex ASD. In these cases, multiple repositioning and device change were required in 46.3% versus 8.3% ($p = 0.001$) and 20.9% versus 4.2% ($p = 0.049$) respectively. In 2 cases, 2 devices were implanted under 3D-TEE guidance to close multiple defects. Overall, procedural success remained high (96.7%). Failures (2 large ASD > 28 mm and 1 ASD

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with deficient inferior rim) and complications (1 transitorily pericardial effusion and 1 day-one surgically managed device embolization) were observed only in complex ASD (4.5% and 3.0%).
Conclusion: Complex ASD is observed in around 2/3 children referred for percutaneous closure. Despite a harder procedure, success rate remains high and complications rare. Discrepancies between sizing technique are higher in complex ASD. 3D-TEE may be more useful in these cases to describe precisely the anatomy as well as facilitating the device positioning.

P2-79

Stenting for Coarctation of the aorta (CoA) in young patients (<3 years)-short- and mid-term results

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Introduction: Today, young patients with native CoA are treated surgically. However, surgery for re-CoA is associated with increased morbidity and even mortality. Some children with native CoA present relative contraindications for surgery. CoA-stenting may be an alternative treatment option in these patients. We present the short- to mid-term results after CoA-stenting in small children.
Material and Methods: Between January 1999 and November 2015, 218 patients with CoA were treated with stents. Fifteen of these (female - 9, median age 8 months [3-34]; median weight 5.8 kg [4.6-14.7]) were included into the study (age < 3 years; weight < 15 kg). Diagnoses: re-CoA post-surgery n = 14 (Norwood 8, CoA-end-to-end anastomosis 3, complex arch reconstruction 3), native CoA n = 1 with relative contraindications for surgery; ALL on chemotherapy. Seventeen stents were implanted in 15 patients (Osypka baby stent 3, Cook formula 14). Unsuccessful balloon angioplasty preceded stent implantation in 7 patients.
Results: Procedural success was obtained in all patients. The mean systolic gradient declined from 37 ± 34 mmHg to 6 ± 11 mmHg (p = 0.003). The stenosed aortic diameter increased form a mean value of 3.1 ± 1.5 mm to 6.5 ± 1.8 mm (p = 0.001). In four patients the intervention was performed by venous access. In three patients the left subclavian artery was covered by the stent (re-opening by balloon angioplasty n = 1). There were no serious complications. The mean follow-up time was 3.0 ± 4.5 months, during this time five patients required re-dilatation and one re-stenting. Mean time of re-intervention was 6 ± 4 months.
Conclusion: Percutaneous stent implantation for Re-CoA and in selected patients with native CoA can be performed successfully in very young patients. However, repeated stent angioplasties and further on interventional 'opening' of the stent with a larger stent shall be necessary to augment the aorta to adult size.

P2-80

'All roads lead to Rome': Transradial access as alternative routes of vascular access for performing pediatric cardiac catheterization

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Radial arterial access for coronary angiography and left heart catheterization is widely used in adults. We aimed to determine its safety and feasibility in pediatric practice.Twenty-three children and adolescents (25 procedures) with congenital heart disease (CHD) were studied. We conducted 24 diagnostic and 1 interventional procedures. In all patients functioning of their palmar arch was proven through Allen's test.The radial artery was cannulated with a 5-Fr sheath after subcutaneous injection of 2% lidocaine. In order to prevent an arterial spasm, we injected a Verapamil and Heparin cocktail on all patients. We did a diagnostic catheterization on 21 patients by using a Pig-tail catheter. On all of these patients we concluded all planned procedures (angiography, oximetry and manometry). Selective coronary angiograms were obtained using Judkin or Amplatz coronary catheters in 3 patients. On a patient with coarctation of the abdominal aorta and stenosis of the left renal artery, we stented the artery through a 6 F transradial sheath. All children had a haemostatic bandage with Adheban after the procedure. Median age was 15.4 years (range, 12.10 –17.11 years), and median weight 63.5 kg (range, 46–115 kg). Everyone was premedicated with Midazolam or Midazolam and Bromazepam. In 17 cases (68%) the study was performed under local anesthesia. Five children had a marked arterial spasm. For the first one it was necessary that the catheterization continues through the femoral artery. The other child overcame the spasm after the application of 2,5 mg Verapamil.We had no other complication during or after the procedures. In all diagnostic catheterizations we performed the required volume of angiographies, manometries and oximetries without difficulty of access. In all pediatric patients we did not hesitate to ask for help from an adult interventional cardiologist if technical difficulties occurred. In conclusion, we believe that transradial access is a good and safe alternative for performing diagnostic and interventional procedures on children and adolescents with CHD.

P2-81

Cryptogenic transient ischaemic attack and stroke recurrence in patients referred for patent foramen ovale assessment

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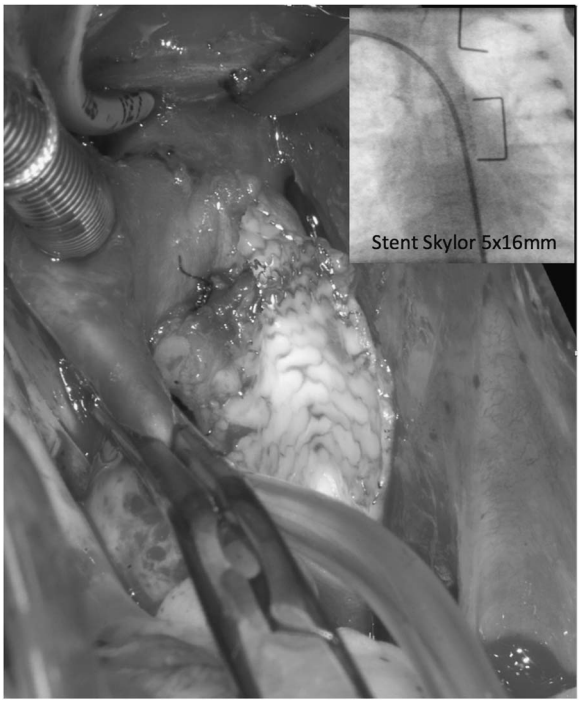
Introduction: Patent foramen ovale (PFO) is associated with cryptogenic stroke (CS). Due to a lack of definitive data, the case for percutaneous PFO closure for the secondary prevention of stroke versus medical therapy alone remains controversial. This study therefore aims to determine the benefits of percutaneous PFO device closure in preventing the recurrence of stroke and TIA.
Methods: Patients who were referred to our PFO clinic following cryptogenic stroke or transient ischaemic attack (TIA) were divided into 2 groups: those that had their PFO closed (closure group) and those that did not have a PFO closure (non closure group), which included patients who did not have a PFO and patients with a PFO who did not proceed to closure. Data was collected from our institution's electronic patient records system as well as by patient questionnaire. Information regarding patient demographics, clinical characteristics and outcomes were recorded. The study end point was a recurrence of stroke and/or TIA during follow up. Data were analysed using chi squared test, or Fisher's exact test where appropriate.
Results: A total of 291 patients was analysed. The closure group contained 199 patients and the non closure group 92 with

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mean (±SD) follow up times of 4.0±3.2 years and 2.9±2.4 years, respectively. There was a significant difference between the rates of stroke and/or TIA recurrence in these two groups, with 8 events in the closure group versus 10 events in the non closure group (4.0% compared to 12.7% respectively, P=0.01).
Conclusion: PFO closure is superior to non closure in reducing recurrence of stroke or TIA in at risk individuals. Further research, specifically additional randomised controlled trials, are required in this field.

P2-82
Coronary Stent Implantation in Critical Aortic Coarctation as Bridging Therapy to Surgery in Very Low Birth Weight Infants
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Introduction: Surgical treatment of critical aortic coarctation (CoA) is extremely difficult in very low birth weight (VLBW) newborns and is usually postponed until at least 2 kg of weight. Sometimes earlier treatment is warranted due to brachiocephalic hypertension, systemic underperfusion or pulmonary overflow. Surgery remains a high-risk option at this weight when prostaglandin infusion had to be discontinued.
Objectives: To review the results of primary interventional coronary stent implantation as bridging therapy to surgery in VLBW newborns with CoA.
Methods: Clinical, echocardiographic, catheterization, surgical and neurodevelopmental data were retrospectively reviewed of all VLBW newborns who underwent primary stent implantation.



Results: Between 2010 and 2015, 5 VLBW neonates underwent primary stent implantation. In all children initial treatment with prostaglandin was discontinued due to severe side effects and/or ineffectiveness. Median age and weight at intervention were 14 days (range 12-16) and 1200 gram (680-1500) respectively. Median invasive gradient was 42.5 mmHg (40-45) before and 2 mmHg (0-10) after stenting. Coronary stent diameter ranged from 3 to 5 millimeter. The femoral artery used for intervention was occluded in 4/5 infants without clinical compromise. There were no other procedural complications. In one infant early restenosis and severe aneurysm occurred 2 months after stenting and was treated with covered coronary stents. To date 4/5 children received surgical correction at a median age of 189 days (111-130) and weight of 5400 gram (4500-6800). No reinterventions were indicated during a median postoperative follow-up of 821 days (186-1622). Neurodevelopmental outcome was unremarkable in all patients. Median Griffiths scores were normal and comparable between patients and their siblings (4/5 patients were gemelli), 89.5 (80-102) and 88 (81-107) respectively.
Conclusions: Coronary stent implantation is a feasible bridging therapy to surgical repair in VLBW newborns with CoA in whom prostaglandin therapy fails.

P2-83
Percutaneous treatment of coarctation of the aorta after Norwood procedure with Valeo Vascular Stent in patients before 2nd stage of univentricular palliation
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Introduction: Postoperative coarctation of the aorta is one of the most common complications of the Norwood procedure with the prevalence reaching 40%. Successful treatment with balloon angioplasty was reported in numerous papers. Implantation of stents was advocated in cases with unfavourable anatomy or unsuccessful angioplasty attempts. Valeo stent is a pre-mounted low-profile, balloon expandable stent with open-cell design and possibility of dilatation up to 13-20 mm, what makes it an interesting option for infants with post-Norwood aortic coarctation (pNCoA).
Methods: A group of 11 patients in whom the Valeo vascular stents were implanted for pNCoA between years 2012 and 2015 was analysed retrospectively. Indications for stenting, anatomic and haemodynamic data, complications, as well as immediate and midterm effects were considered.
Results: Seventy-four Norwood procedures were performed at our institution between 2012 and 2015. In this period 24 patients (aged 1-8 months, median 4.5 month) were treated for pNCoA before Glenn procedure. Balloon angioplasty was successful in 13 cases, in 8 cases it was followed by implantation of Valeo stent, 3 patients received Valeo stent as a primary treatment (because of complex anatomy of the lesion and high risk of stenosis recurrence). Pre-implantation peak systolic pressure gradients across the pNCoA ranged from 12 to 50 mmHg (avg. 29 mmHg). Stents were implanted using anterograde route (8/11) or retrograde route (4/11). Stent nominal diameter was 6 mm (5/11), 8 mm (4/11) and 10 mm in 2/11 cases. After the procedure, the peak systolic pressure gradient dropped to avg. 6 mmHg (0-18 mmHg). The only immediate complication (1 case) was dislodgement of stent

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from the balloon, followed by successful implantation of another stent. No complications were observed in the short- and midterm follow-up. In 3 patients with post-implantation peak systolic pressure gradients of 10 mmHg (2) and 18 mmHg (1) the stents were electively re-dilated with full pressure gradient relief.
Conclusions: Implantation of Valeo vascular stents is the acceptable treatment strategy in patients with post-Norwood aortic coarctation in patients resistant to balloon angioplasty or at high risk of coarctation recurrence. It appears to be safe and effective in short- and midterm follow-up.

P2-84

Are Amplatzer Duct Occluder II Additional Sizes devices dedicated only for smaller children?

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Aim: To present our experience with Amplatzer Duct Occluder type II Additional Sizes (ADOIIAS) for closure of different types of patent ductus arteriosus (PDA) in patients with distinct age compartments.
Methods and results: There were analyzed group of 103 patients, in whom PDA (diameter below 3.5 mm) was closed with ADOIIAS. The median age of treated patient was 3.0 y (from 0.1 till 24 yo)- 55 pts (53.4%) were older than 3 years. Ductal anatomy defined by angiography showed Type A in 42 pts (40.8%), type C in 6 (5.8%), type D in 21 (20.5%), type E in 34 pts (33.0%). Two embolization of the device occurred shortly after implantation.
Both occluders were retrieved percutaneously. One death occurred in neonate 4 days after ADOIIAS implantation not related with the procedure (cause multiorgan failure). In all patients next day after the procedure total occlusion of PDA was confirmed. No protrusion of the device to aorta nor pulmonary artery was seen in any patient in follow-up.
Conclusions: Application of Amplatzer Duct Occluder type II Additional Sizes is good therapeutic option to treat adequate PDA. The implant may successfully substitute coil implantation in all age groups.

P2-85

Emergent diagnostic and interventional cardiac catheterizations within 30 days post congenital heart surgery in children – a single centre 15 years experience

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Background: To assess indications, feasibility, safety and outcome of emergent cardiac catheterization procedures in the early post-operative period within 30 days after congenital heart surgery (CHS) at our institution.
Methods: Retrospective, single centre case review study of all emergent cardiac catheterizations between 01/2001 and 12/2015 within 30 days after CHS.
Results: A total of 207 unplanned (83 diagnostic and 124 interventional) procedures were performed in 168 patients. Median age at catheterization was 4 months (range 0 – 207), median weight was 4.9 kg (range 2 – 85). 115 procedures (56%) were performed in patients with univentricular heart disease. Catheterizations were performed at a median of 7 days (range 0 – 30) after CHS. Previous cardiac surgery was bidirectional/total cavopulmonary connection (BCPC/TCPC) (n = 66, 32%), shunt- or Norwood/comprehensive Norwood I+ II procedures (n = 45, 22%), right ventricular outflow tract surgery/RV-PA graft implantation (n = 29, 14%), arterial switch-operation (n = 15, 7.3%), repair of aortic coarctation (n = 11, 5.3%), and others (n = 41, 19.8%). Indications for catheterization were prolonged postoperative course (13.5%), post BCPC/TCPC (10.1%), hypoxemia (17.4%), ECG-changes/ suspected coronary pathology (12.6%), suspected pulmonary artery stenosis (17.9%), assumed re-coarcations (4.3%) and others (16.4). Diagnostic procedures revealed significant pathology leading to early redo-surgery (n = 16, 19.3%). 124 interventions included stent-implantations/balloon-dilatations in/of pulmonary arteries (n = 56, 45%) or (re-) coarctations (n = 12, 9.7%), coil-embolizations of aortopulmonary (n = 14, 11.3%) and venovenous collaterals (n = 5, 4%), manipulations of shunts and fenestrations (n = 12, 9.6%), and others. 13 patients were catheterized under mechanical extracorporeal support. Immediate success rate was 96%. There was no intraprocedural mortality. There were 3 (1.3%) major complications (SVC rupture n = 1, severe intracranial bleeding n = 1, cardiopulmonary arrest and emergency surgery n = 1) and 8 (3.9%) minor complications (arrhythmias, vessel thrombosis, minor blood vessel dissections, pneumothorax). Risk factor analysis revealed no statistical significant difference for the occurrence of complications for patients' weight (p = 0.194) or underlying uni- versus biventricular heart disease (p = 0.59).
Conclusion: Emergent cardiac catheterizations can be performed safely, with a high diagnostic and therapeutic value in the early postoperative period. Therefore, diagnostic and interventional catheterizations should not be withheld from these patients at any time after CHS.

P2-86

Longterm follow up of interventional closure of atrial septal defect using the Solysafe™ Septal Occluder

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Objective: Since August 2010 distribution and sale of the Solysafe Septal Occluder (SSO) is stopped for interventional closure of atrial septal defect (ASD II) due to reported wire fractures. Long-term follow up data are lacking.
Methods: We analysed the incidence of irregularities associated with SSO during longterm follow up until December 2015 by fluoroscopic assessments as recommended by the company.
Results: SSO was successfully implanted in 51 children (male 24) at an age (mean ± SD) of 8.0 ± 4.6 (range 1.6-17.8) years and a body weight of 29.1 ± 18.1 kg (9.5-86) with a SSO size of 15mm (n = 28), 20mm (n = 13), 25mm (n = 8), and 30mm (n = 2). During longterm follow up all patients were clinically asymptomatic. Fluoroscopy showed after 4.8 ± 1.6 years in 17 patients (33.3%) irregularities, i.e. fractures or dehiscences, not determined by echo before. In eight patients (15.7%) we found single or multiple dehiscences of wire loops disconnected from the central pin, and in nine (17.6%) patients single or multiple fractures. Dehiscence was only found in children with 15 mm devices (n = 8), while the risk of fractures increased with SSO size (15 mm size, 7.1%, 20 mm, 15.4%, 25 mm, 37.5%, 30 mm, 100%).

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The yearly risk of irregularities increases after implantation (3rd year, 5.9%, 4th, 6.3%, 5th, 6.7%, 6th, 7.5%, 7th, 10.5%). The integrity of the device was not altered in 50 patients (98%), despite one patient with three fractures of wire loops on both sides of the device and embolized wire fragments to both pulmonary arteries detected two years after implantation. The fractured wire ends sticked out of the septum and injured anterior mitral leaflet, therefore, cardiac surgery was performed with complete explantation of the device and successful mitral valve repair and uneventful postoperative course.
Conclusions: During longterm follow up the rate of irregularities with fractured or dehiscent wire loops of implanted SSO is high, and progresses continuously, and requires regular fluoroscopic controls. Larger size of SSO is a risk factor for fractures. Despite radiological irregularities the integrity and stability of most of the devices were not affected due to a firm and adequately thick coat of scar tissue, which hopefully will ensure longterm safety for the patients.

P2-87
Treatment of persistent chylothorax after surgery for heterotaxy syndrome
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Objective: Chylothorax is a serious complication after congenital heart surgery and it is mostly encountered after univentricular repair. Although conservative management usually enough for treatment, some patients may need surgical intervention. Ductus thoracicus ligation with right thoracotomy is the preferred approach. Heterotaxy syndrome is a lateralization disorder and there is no data about the localization of the ductus, which makes a confusing situation in those patients who need surgical intervention due to persistent chylothorax.
Methods: Three patients who had univentricular heart and heterotaxy syndrome underwent cavopulmonary anastomosis (n = 2) or Kawashima operation (n = 1). Their ages were 7 months, 3 years and 1.5 years respectively. One patient had previous modified BT shunt and one patient had bilateral banding because of associated aortic arch hypoplasia as a first stage palliation. Dextrocardia was present in two of the patients. Chylothorax developed at 15 days after the operation in 1 patients who underwent Kawashima operation. Others readmitted to the hospital due to chylothorax two weeks after uneventful postoperative period and hospital discharge. Despite maximum medical treatment including high dose of octreotid infusion, high amount of chylouse drainage (>30 ml/day) persisted more than two weeks in all patients. Two patients with right or left sided unilateral persistent chylothorax underwent surgery. Ductus could not be localized and decortication and talk application of the ipsilateral side was performed. MR lymphangiography was done for the last patient who had bilateral persistent drainage to detect the ductus location for proper intervention.
Results: Two patient responded well for surgical intervention. MR lymphangiography revealed both sided, but left dominant thoracic duct in the other patient. Surprisingly, chylothorax decreased dramatically after MR study in this patient. We speculated that lymphatic vessels might be occluded by contrast agent Lipiodol.
Conclusions: Persistent chylothorax in patients with heterotaxy syndrome is difficult to manage, because of the unknown location of the thoracic duct. Decortication and talk application for the ipsilateral thoracic cavity may be an effective intervention. MR lymphangiography might be helpful for ductus localization in patients with bilateral persistent drainage.

P2-88
The efficacy of thoracic USG in postoperative newborn patients after cardiac surgery
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Objectives: The diagnosis of respiratory complications like pleural effusion, atelectasis and pneumothorax is important in postoperative management of newborns after congenital cardiac surgeries. Most frequent method to differentiate these intrathoracic pathologies is X-ray but particulary in newborn patients radiation exposure is detrimental. Thoracic ultrasonography (USG) is a diagnostic tool increasingly used in critical care. Few data are available concerning the pediatric and neonatal populations. In this study the efficacy of thoracic USG during echocardiography was evaluated in newborns.
Methods: 60 newborns were evaluated after pediatric cardiac surgery, successively between 1 March 2015-1 September 2015 in this study. Patients were evaluated for effusion, atelectasis and pneumothorax by USG and results were compared with X ray findings.
Results: 60% (n = 42) of the cases were male, the median age was 14 days (2-30 days), the median body weight was 3.3 kg (2.8-4.5 kg). The median RACHS-1 score was 4 (2-6).Atelectasis was demonstrated in 66% (n = 40) of the cases. 5 of them were determined solely by x-ray, 10 of them only by USG, 25 of them by both USG and x-ray. Pneumothorax was determined in 20% (n = 12) of the cases. Accept a case determined by both methods, all of the 11 cases were diagnosed by x-ray. Pleural effusion was diagnosed in 26% (n = 16) of the cases. 4 of the cases were demonstrated solely by USG, 3 of them solely by x-ray, 9 of the cases by both methods. Pericardial effusion was demonstrated in 10% (n = 6) of the cases. Except 1 of the cases determined by both methods, 5 of the cases were diagnosed by USG.
Conclusion: Thoracal USG is a beneficial non-invasive method to evaluate postoperative respiratory problems in newborns who had congenital cardiac surgery.

P2-89
Use of 3D printing in the evaluation of complex Congenital Heart Disease (CHD). 4 Cases
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Introduction and Objectives: The practice of modern medicine in many medical subspecialties relies on advanced imaging.

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Ultrasonography, Computerized Tomography and Magnetic Resonance Imaging can produce 3D images aiming to provide physicians with improved appreciation of the anatomy. Three dimensional printing (3D printing) is a new tool poised to help surgeons appreciate complex cardiac anatomic features and their interrelationships with surrounding tissues. We sought to explore the utility of creating anatomically accurate 3D printing models for patients with complex or unusual congenital heart defects in facilitating diagnostic understanding and surgical planning.
Methods: Based on contrast–enhanced computer tomography (CT) or magnetic resonance (MRI) images, after appropriate segmentation and processing, accurate 3D printed models of the relevant cardiac anatomy (including appropriately planned sections) were created. These were utilized for the entire surgical team to appreciate the anatomy, for patient family education, and for optimizing surgical planning, including surgical simulation. We present 4 cases, two patients with anomalous aortic origin of coronary artery (AAOCA), and two with double outlet right ventricle (DORV) and TGA, (one with restrictive VSD and the other with complete atrio-ventricular septal defect).
Results: The models proved helpful for diagnostic appreciation of the anatomy, family education, surgical decision making, preoperative planning and surgical simulation.
Conclusion: Printing of 3D cardiac models is a new and much promising tool for the diagnostic assessment and preoperative preparations for patients with complex or unusual congenital heart defects. The opportunity to handle accurate models of the anatomy in ways impossible even at operation enables the care team to appreciate potential procedural difficulties, avoid surprises during operation, and clarify aims and limitations of planned surgical interventions. This new technology can enrich patients', students', and physicians' understanding of structural heart disease with the ultimate result of enhancing the level of care provided to this growing subset of patients.

P2-90
Different prediction of fluid responsiveness by pulse pressure variation in children after surgical repair of ventricular septal defect or Tetralogy of Fallot

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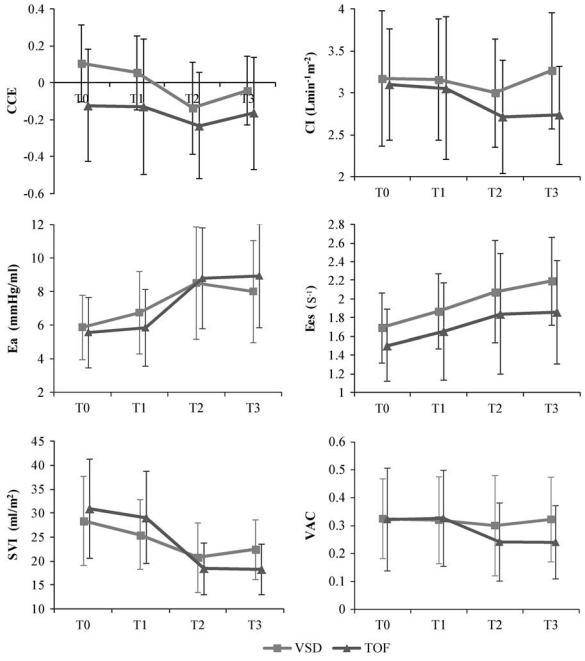
Introduction: Volume expansion therapy is considered the first-line treatment in children after cardiopulmonary bypass (CPB), but excessive volume is harmful. Pulse pressure variation (PPV) derived from pressure record analytical method (PRAM) is based on heart-lung interaction during mechanical ventilation. The heart-lung interaction might be different between ventricular septal defect (VSD) and Tetralogy of Fallot (TOF) due to different right ventricular function and pulmonary vasculature, potentially affecting the predictability of fluid responsiveness using PPV after surgical repair.
Method: Children undergoing CPB for complete repair of VSD (Group VSD, $n = 29$, aged 0.7 ± 0.2 years) and TOF (Group TOF, $n = 36$, aged 0.8 ± 0.3 years) were enrolled. After CPB and before chest closed, mechanical ventilation was set with tidal volume 10 ml/kg. 5% albumin or blood plasma routinely was given (16 ml•kg-1•h-1) over 15 minutes. PPV was recorded using PRAM along with heart rate (HR), stroke volume index (SVI), cardiac index (CI) before and after volume replacement. Patients were considered as responders to fluid loading when CI increased $\geq$ 15%.
Results: In Group VSD, 12 were responders and 14 non-responders. PPV in responders was higher than that in non-responders ($25.7 \pm 6.4\%$ vs. $16.6 \pm 5.0\%$, $P < 0.01$). Area under the curve (AUC) was 0.85 (95% confidence interval, $0.69 \sim 1$, $P = 0.01$) and cutoff value 19% with a sensitivity of 92% and a specificity of 71%. In Group TOF, 15 were responders and 21 non-responders. PPV in responders were not different from that in non-responders ($11.6 \pm 4.6\%$ vs. $10.1 \pm 2.0\%$, $P > 0.1$). AUC was 0.52 (95% confidence interval, $0.31 \sim 0.72$, $P = 0.01$).
Conclusion: PPV by PRAM can be used to predict fluid responsiveness in children after surgical repair of VSD, but not in children with TOF.

P2-91
Pressure record analytical method monitoring ventricular efficiency in children with Tetralogy of Fallot undergoing surgery correction

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Introduction: Energy-based endpoints are proved to be superior indicators in accessing hemodynamic status. Cardiac cycle efficiency (CCE) and ventricular-arterial coupling (VAC) derived from pressure record analytical method (PRAM) are parameters related with ventricular efficiency. We aim to compare Tetralogy of Fallot children (TOF) with control ventricular septal defect (VSD) children undergoing correction surgery on profile of ventricular efficiency.
Methods: 50 children with VSD (1.3 ± 1.0 year and 46 with TOF (1.0 ± 0.8 year) scheduled for complete repair, were enrolled. In operation room, a radial arterial catheter was inserted to allow routine monitoring and the use of PRAM. Heart rate (HR), dicrotic arterial pressure, indexed stroke volume (SVI), cardiac index (CI), and CCE were recorded before surgical incision (T0), after pericardium cut (T1), after removal of aortic cannula (T2), end of operation (T3). Left ventricular end-systolic elastance (Ees), left ventricular end-systolic arterial elastance(Ea) and VAC were calculated.
Results: Intra VSD group comparison, CCE was lower at T2 and T3 compared with T0 ($P < 0.01$), higher at T3 compared with T2 ($P < 0.01$). Ees was higher at T1, T2 and T3 compared with T0 ($P < 0.01$), higher at T3 compared with T2 ($P < 0.01$). Ea was higher at T1, T2 and T3 compared with T0 ($P < 0.01$). Intra TOF group comparison, CCE was lower at T2 compared with T0 ($P < 0.05$), VAC was lower at T2 and T3 compared with T0 ($P < 0.01$). Ees was higher at T1 ($P < 0.05$), T2 and T3 ($P < 0.01$) compared with T0. Ea was higher at T2 and T3 compared with T0 ($P < 0.01$). As compared with VSD group, in TOF group CCE was lower at T0, T1 ($P < 0.01$) and T3 ($P < 0.05$), VAC was lower at T3 ($P < 0.01$), Ees was lower at T0, T1, T2 ($P < 0.05$) and T3 ($P < 0.01$). CCE was significantly correlated with VAC at T0, T1, T2 and T3 ($P < 0.001$).
Conclusions: RPAM could track ventricular efficiency changes in children with TOF. Left ventricular efficiency was deteriorated after surgical repair in TOF children because of the marked increase of the afterload and the slight increase of contraction, and it needs to be optimized.

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P2-92

First experience of a novel decellularized patch material (CardioCel™) for aortic valve reconstruction in congenital heart surgery

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Objectives: The search for optimal patch material is an ongoing challenge in congenital heart surgery (CHS). In this study, we report our first experience with the use of a novel decellularized bovine pericardial patch material (CardioCel™) for aortic valve reconstruction (AVR) in CHS.
Methods: We retrospectively studied 28 patients (median age 10 (1,7-34) years, median weight 28 (9-100) kg) who underwent AVR using CardioCel™ Patch at our institution between February 2014 and August 2015. 9 patients suffered from predominant aortic valve stenosis, 19 patients showed predominant aortic valve insufficiency. Echocardiographic results preoperatively, at discharge and median 14 (2-18) months following the operation were used for the assessment of graft appearance and performance. Data are presented as median values.
Results: Aortic valve reconstruction using CardioCel™ Patch was feasible in all patients without intraoperative difficulties implanting the patch material. There was no perioperative mortality. Median intensive care unit stay was 21 (2-121) hours and the median hospital stay was 7 (5-12) days. Echocardiography at discharge showed excellent patch function, no signs of device calcification, thrombosis or device failure of the presented cases. Mean pressure gradient was reduced in patients with aortic valve stenosis after AVR (47 (30-55) vs 20 (1-30) mmHg) and remained reduced at 10 (2-16) months follow up (22 (6-52) mmHg). In 1 patient restenosis after AVR was seen at 16 months follow up, which was treated by balloon valvuloplasty. Aortic valve insufficiency was reduced after AVR (grade 3 (2-3) vs 1 (0-2)) and remained low (grade 1 (1-2)) at 14 (12-18) months follow up.
Conclusion: This study demonstrates promising early results for the use of CardioCel™ patch AVR in patients with congenital heart disease and aortic valve lesions. Implantation was free of tissue related complications and mid-term patch function showed good results.

P2-93

Colonization with multi-resistant gram-negative (MRGN) bacteria in paediatric cardiovascular surgery – is it an additional risk factor for post-operative morbidity?

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Objectives: The incidence of patients who are colonized with multi-resistant gram-negative (MRGN) bacteria showed a significant increase over the recent years. This finding is paralleled by an increased number of nosocomial infections due to MRGN bacteria. In this study, we aimed to determine the prevalence of colonization with MRGN bacteria in a paediatric population after cardiac surgery and its impact on early postoperative outcome.
Methods: Retrospective analysis of all patients who were treated on our paediatric cardiac intensive care unit (PCICU) between January 2012 and December 2015. The following parameters were obtained: demographic information, MRGN status on admission to the PCICU, type of underlying cardiac pathology, type of operation, duration of postsurgical PCICU stay, systemic-inflammatory-response-syndrome (SIRS) criteria, change in empirical antibiotic therapy and the crude mortality rate (CMR). A subgroup analysis of these outcome parameters was performed in patients after Fontan completion (MRGN-positive vs. MRGN-negative).
Results: During the study period of four years, a total of 150 (8,3%) of 1818 patients operated on with cardiopulmonary bypass were found to be MRGN-positive prior to the admission to the PCICU. Of these, 33 (22%) patients were found to be ESBL-positive, 70 (45.3%) MRGN2-positive, 48 (31.3%) MRGN3-positive and 1 (0.7%) MRGN 4-positive. There was no statistically significant difference in the CMR between MRGN-positive patients and the overall group (0.7% vs. 2.4%). 14(9.3%) MRGN-positive patients fulfilled SIRS criteria. In 49 (32.7%) of MRGN-positive patients, empirical antibiotic therapy was changed, that was associated with prolonged hospital stay. In the subgroup analysis of patients after Fontan completion, there was no significant difference in CMR and morbidity (ICU-Stay, hospital stay) between MRGN-positive (n = 9) and MRGN-negative patients.
Conclusion: Early detection of MRGN-colonization prior to operation can help to manage this challenging clinical scenario. With informed change of empirical antibiotic therapy we were able to manage the problem without clinical deterioration, also in patient with abnormal hemodynamics. Early MRGN screening of vulnerable patient populations seems to be an important measure to reduce morbidity and mortality.

P2-94

Continuous hemodialysis therapy for a very low birth weight (VLBW) premature with acute renal failure and congenital heart disease

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Case description: The premature girl was born at 32 weeks of gestation with a birth weight of 1495 g and Tetralogy of Fallot (TOF) with severe infundibular and valvular pulmonic stenosis as well as hypoplastic pulmonary arteries. Due to progressive hypoxic spells, at the age of 31 days an aortopulmonary shunt (diameter: 3.5 mm) was placed. Postoperatively, the girl developed a pronounced capillary leak syndrome with acute renal failure. Peritoneal dialysis was started at the age of 32 days. Due to an intestinal perforation on day 43, a bowel resection with ileostomy was performed. Nevertheless peritoneal dialysis was continued, fluid retention increased and mechanical ventilation worsend.
The primary technical challenge for initiating continous hemofiltration in this VLBW premature with an actual weight fo 1800 g was the size of the catheter for continous veno-venous hemofiltration and the capacity of the hemofiltration set. We successfully placed the smallest available short term dialysis catheter with a diameter of 6-French (Joline, Hechingen) into the right jugular vein. Hemofiltration was initiated with the Plasauto Sigma-System (DIAMED, Cologne). Following the priming of the hemofiltration set (filling volume of the set: 47 ml) with heparin and sodium chloride, the set was filled with a mixture of red cell concentrate (75%), fresh frozen plasma (25%) and sodium bicarbonate (2 ml). On hemofiltration, the set was run with the blood pump of 7-10 ml/min, an ultrafiltration rate between 10-30 ml/h and a replacement fluid rate of 100 ml/h. Despite effective anticoagulation with heparine and a prothrombin time over 60 seconds and anti-factor Xa over 0.4 IU/ml the major complication was clotting of the hemofiltration sets venous line connected to the patients venous vascular access. As a result of the clotting, the hemofiltration set had to be changed 12 times (in average every 39.15 hours). Uremia and lung funtion improved quickly. Hemofiltration was needed over 29 days, renal function recovered completely. A clipping of the aortopulmonary shunt to reduce pulmonary blood flow was performed 8 days after admission. The patient was discharged home after 88 days.
Conclusions: Continuous hemodialysis therapy is a successful and safe possibility even in VLBW prematures with acute renal failure.

P2-95
Chylothorax after surgery for congenital heart disease - incidence, risk factors and treatment

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Introduction: Chylotorax is a known and severe complication after congenital heart surgery. The incidence of chylotorax ranges from 0.5 to 9% and it is associated with respiratory, nutritional, immunologic, hematologic and metabolic morbidity.
Methods: We present a retrospective analysis of data from our centre from 1/1/2012 to 30/10/2015. We report the incidence of chylothorax after congenital heart surgery, risk factors associated with its occurrence and the therapeutic approach at our centre.
Results: During this period 596 surgeries in 513 patients with congenital heart disease were performed. The incidence of chylotorax was 3%(16 patients). Chylothorax occurred after arterial switch (6 patients), total cavopulmonary connection (3), correction of tetralogy of Fallot (3), correction of aortic coarctation, creation of interatrial communication, mediastinal debridement and cardiac transplantation (1).
The occurrence of chylothorax was associated ($p < 0.05$) with younger age at surgery (1.6 ± 2.8 vs 8.2 ± 15.3years, mean $\pm$ standard deviation), longer extracorporeal circulation (148 ± 46 vs 86 ± 5 minutes), longer aortic clamping (84 ± 48 vs 52 ± 38 minutes), delayed sternal closer (RR 24.9; IC95% 10.7-69.2) and prolonged stay in the intensive care unit (53 ± 40 vs 15 ± 22 days). Diagnosis of chylothorax was also associated with the occurrence of: arrhythmias requiring treatment ($p = 0.009$), seizures ($p < 0.01$), cerebral vascular accident ($p < 0.01$), diaphragmatic paralysis ($p < 0.01$), acute renal injury requiring dialysis ($p < 0.01$) and infectious complications ($p < 0.01$). One patient died without resolution of chylothorax. There was no statistical association between chylothorax and mortality in our study.
Diagnosis of chylothorax was made at a mean of 8 days after surgery (range 4 to 15 days). All patients were treated with hypolipid diet, the only treatment required in two thirds of patients and resolution was in mean after 12 days (4-29 days). Treatment with octreotid was required in 7 patients and was effective in 4(57%). This treatment was initiated in mean at the 5th day after diagnosis (0-18 days) and mean treatment duration was 10 days (2-17 days). In two patients (12,5%) surgery was required (ligation of thoracic duct and pleurodesis).
Conclusions: Chylothorax was a rare complication. It was associated with younger age, complex surgical procedures, higher morbidity and longer stay in the intensive care unit. Chylothorax was not associated with increased mortality. Most cases were treated with hypolipid diet or hypolipid diet and octreotid. A minority of patients required surgery.

P2-96
25 years of the Fontan operation: experience of a referral centre

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The Fontan operation is the final palliative procedure in patients with univentricular hearts. The ideal age to perform the surgery is still unknown and the clinical outcomes of contemporary Fontan survivors need further investigation. We aimed to describe our patients' characteristics and to determine the outcomes after Fontan surgery.
Single-centre retrospective review of 25 paediatric and adult patients who underwent the Fontan operation between 1988 and 2013.
Patients with univentricular hearts with left ventricular morphology predominated (80%). The median age at Fontan operation was 6.2 ± 4.2 years.
Overtime, a shift in surgical practice was observed, the intra-atrial lateral tunnel became the dominant technique after 2000. Overall 68% had an intra-atrial lateral tunnel and 32% a classic or a Fontan variant. About 8% of patients underwent a bidirectional Glenn before Fontan completion, 72% had modified Blalock-Taussig shunts at a median age of 0.27 ± 1.25 years. The mean pulmonary pressures (mPAP) before surgery were 12 ± 1.8 mmHg. Heterotaxia syndromes were correlated with higher mPAP ($p < 0.009$).

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Atrial arrhytmias were present in 32%, tromboembolic events in 8% and protein loosing enteropathy in 4%. No correlation was found between age at surgery or Fontan technique and poorer outcomes.
In the paediatric group (n = 9), median 11.9 ± 3.1 years, mean follow up 6.6 ± 7.4 years, 2/3 of patients are asymptomatic and 1/3 in NYHA class II. 3 patients had atrial arrhythmias, 33% were on diuretics.
In the adult group (n = 16), median 28 ± 6.58 years, mean follow up 18.3 ± 7.44 years, 63% were on NYHA class II, 12.5% on NYHA class III, 24.5% were asymptomatic. In this group, patients with classical or a Fontan variant were on antiarrythmic drugs (p = 0.006) diuretics and angiotensin converting enzyme inhibitors (p = 0.027), comparatively to the intra-atrial lateral tunnel.
Perioperative mortality was 8%. Re-operation after Fontan surgery was correlated with higher mortality (P = 0.04). Freedom from death or transplantation was 88%.
Survival in patients undergoing the Fontan surgery is good, but patients remain at high risk for adverse events. Poor outcomes in Fontan patients are probably not only due to timing, but more importantly related to patient selection.

P2-97
The impact of pulmonary valve-sparing techniques on early postoperative results in tetralogy of Fallot repair

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Introduction: Patients undergoing repair of Fallot tetralogy (TOF) with a transannular patch (TAP), may require pulmonary valve replacement long after the initial operation. However impact of pulmonary valve-sparing (PVS) procedures on early postoperative results is controversial. In this study, we reviewed the impact of pulmonary valve-sparing techniques on early postoperative results in tetralogy of Fallot repair.
Methods: A total of 64 patients with a diagnosis of TOF, operated by the same surgeon from 2010 to 2015 were included in this retrospective study. Mean age of the patients was 19.96 ± 14.23 months. Forty patients (62.5%) were male. Thirty two of the patients (50%) were under 1 year of age. PVS techniques could be performed in 29 patients (46%), while the remaining 35 patients (54%) had TAP. In 15 of them, pericardial monocusp valve was created. PVS techniques were transatrial repair in 9 patients; transatrial-transpulmonary in 8, infundibular patch in 8 and infundibular-pulmonary patch in 4 patients.
Results: There was no early postoperative mortality in the PVS group. Five patients (14.2%) died early after operation in the TAP group (p = 0.058). Of these patients, 2 had pulmonary monocusp insertion (p = NS). The causes of mortality were sudden cardiac arrest (n = 2), multiorgan failure (n = 1), low cardiac output (n = 1) and neurological complications (n = 1). ECMO support was needed for 5 patients in TAP group (p = 0.058). Three of them could be weaned from ECMO and 2 of them were discharged uneventfully. Reoperation was required in 1 patient due to residual VSD in TAP group. Permanent pacemaker implantation was done in 3 patients (2 in PVS and 1 in TAP group). Total postoperative morbidity rate was significantly higher in TAP group (Table 1). Morbidity rate was lower in patients with pulmonary monocusp than those with no monocusp (p = 0.0176).
Conclusions: Early postoperative mortality and morbidity rates are significantly higher in TAP group. Pericardial monocusp insertion might decrease postoperative morbidity. PVS techniques should be used whenever possible during total correction of TOF.

Table 1. Postoperative morbidity

Postoperative morbidity	TAP group (n = 35) (with monocusp n = 15)	%	PVS group (n = 29)	%	p value
Low cardiac output	4 (1)	11.4 (6.6)	0	0	0.12
Prolonged mechanical ventilatory support	6 (2)	17.1 (13.3)	1	3.4	0.12
Peritoneal dialysis	4 (1)	11.4 (6.6)	1	3.4	0.36
Prolonged pleural effusion	4 (0)	11.4 (0)	0	0	0.12
Total	**18 (4)**	**51.4 (26.6)**	**2**	**6.8**	**0.0001**

P2-98
Mid-Term Results of the Ross and Ross/Konno Procedure

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Introduction: The Ross and Ross-Konno procedure may be the only alternative surgical method in infants and children with congenital aortic valve disease. In this study, we aim to show the midterm outcomes of pediatric patients undergoing the Ross operation.
Methods: From 2011 to 2015, a total of 8 patients underwent either Ross or Ross-Konno operation were included in this retrospective study. Median age was 4.5 years (ranged from 3 months to 17 years). Preoperative diagnosis was congenital aortic stenosis in all patients. In 6 of them, aortic regurgitation also present. Four patients had bicuspid aortic valve and 1 patient had Shone's syndrome. Seven patients had previous interventions (balloon valvuloplasty (n = 4), ventricular septal defect repair (n = 1), aortic valve repair (n = 1) and aortic coarctation repair (n = 1)). One patient needed mechanical ventilation preoperatively. Ross-Konno procedure was performed in 5 patients and other 3 patients underwent Ross operation. Pulmonary reconstructions were performed by using Contegra conduit except one patient who had pulmonary homograft. Additional subaortic resection was required in 2 patients, and mitral valve repair in 1. Only one adult sized patient had pulmonary autograft wrapping with dacron conduit.
Results: There was no mortality and major morbidity. Mean cardiopulmonary bypass and cross clamp times were 234 ± 64 and 177 ± 38 minutes respectively. Two patients underwent delayed sternal closure. Three patients had pneumonia. Median intensive care and hospital stays were 4.5 and 13.5 days respectively. All patients were followed up between 3 months and 4 years (mean 29.3 months). Only two of the patients had moderate pulmonary conduit stenosis and one patient had mild mitral valve regurgitation.There was no moderate or severe aortic valve regurgitation seen at last follow-up. The neo-aortic valve and root grew with the patient and median valve Z scores were in the appropriate range for all patients.
Conclusions: Pulmonary autograft and pulmonary conduit functions were good during mid-term follow-up. The Ross and Ross-Konno procedure remains a good option for aortic valve replacement in children and young adults and can be performed with low mortality.

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P2-99

Totally endoscopic robotic repair for sinus venosus atrial septal defect with partial anomalous pulmonary venous connection

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Objective: Robotic repair of sinus venosus atrial septal defect with a partial anomalous pulmonary venous connection (PAPVC) to the superior vena cava offers a less invasive alternative to conventional surgical approaches in selected group of pediatric patients. The aim of this report is to present our experience in robotic surgery in 2 patients with PAPVC, with or without left persistent superior vena cava.
Methods: Between July and September 2015, 2 children (16 and 17 years, one male) was referred to our hospital with a diagnosis of PAPVC and high venous atrial septal defect. The height and weight of patients were 160 vs. 170 cm and 44 vs. 54 kg, respectively. Transthoracic echocardiography examinations confirmed the diagnoses and, in one of these cases, left persistent superior vena cava with the absence of innominate vein was diagnosed. Preoperative biochemical tests and chest X-ray were normal. Patients underwent totally endoscopic robotic surgery with the Da Vinci endoscopic surgery system for repair of these pathologies.
Results: Operations were completed uneventfully. There was no complication developed after operations. Under peripheral cardiopulmonary bypass with moderate hypothermia, lateral cava-atrial incision was made to expose the defects. Left persistent vena cava was drained with a pump sucker through the coronary sinus after right atriotomy incision. Double patch repair was performed using glutaraldehyde-treated autologous pericardial patch (Figure). By this technique, the risk of turbulent flow and stenosis of the right upper pulmonary vein orifice and superior vena cava-atrial junction was eliminated. The mean duration of cardiopulmonary bypass and arrest were 98 vs. 85 minutes and 65 vs. 60 minutes, respectively. Ventilation and intensive care unit stays were 3 vs. 5 hours and 16 vs. 18 hours, respectively. There was no perioperative blood loss and blood transfusion. Patients were uneventfully discharged on post-operative day 3. Follow-up examination were normal with a good clinical status. Echocardiographic examinations showed no turbulence and a superior vena cava-right atrium pressure gradient in both patients.
Conclusions: Robotic repair of PAPVC can be safely managed in selected group of children. The double-patch technique is technically reproducible without any increase in complications.

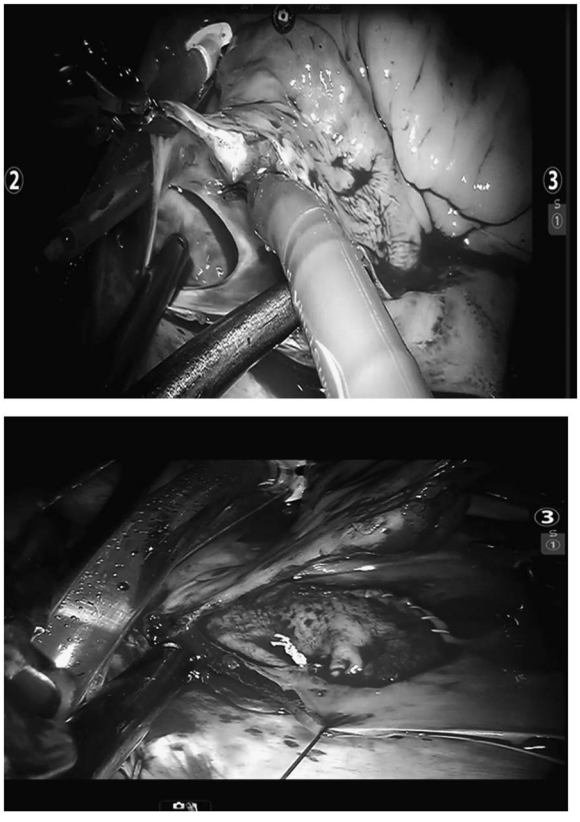


Figure. Endoscopic view shows high venous atrial septal defect through transcaval-atrial incision (left panel) and closure of the defect with pericardial patch (right panel).

P2-100

The Effect of Upper Body Central Venous Catheter Insertion on Superior Vena Cava Anatomy and Systemic Venous Return in Infants with Single Ventricle

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Introduction: The upper-body central venous (UBCV) catheter insertion is a widespread technique during pediatric cardiac surgery but might result in obstruction. This complication may especially have serious results in patients with cavopulmonary shunt (CPS). In this study our aim is to evaluate the effect of UBCVC insertion at first operation on superior vena cava (SVC) anatomy and systemic venous return during inter stage period.
Methods: The data of 73 children, who underwent palliation for single-ventricle (Norwood operation = 6, stage 1 hybrid palliation = 3, systemic to pulmonary artery shunt = 24, PA banding = 22, PDA stenting = 15) and consequently pre-CPS cardiac catheterization between 2010-2015 were gathered retrospectively. Patients were grouped as the ones who had UBCV catheter (group 1) and ones who did not have (group 2). The SVC size evaluated with SVC index and SVC/Nakata index ratio.

Table 1. Patient Demographics

	Group 1 (n = 26)	Group 2 (n = 47)	p value*
Age, months	11.5 ± 7.4	10.2 ± 7.8	0.13
Bilateral SVCs	6 (23.1%)	9 (19.1%)	0.76
Venous obstruction	4(15.3%)	1 (2.1%)	**0.04**
Right internal jugular	1	1	
Right SVC	1		
Left SVC	1		
Left innominate	1		
SVC diameter (mm)	8.9 ± 1.6	8.7 ± 2.2	0.19
Indexed SVC[1]	22.4 ± 4.3	23.8 ± 6.04	0.62
Nakata index(mm/m^2)	243.8 ± 124.5	212.6 ± 101.6	0.38
SVC/Nakata index ratio	0.11 ± 0.09	0.14 ± 0.05	0.30

* p value of < 0.05 was considered statistically significant
[1] SVC size mm/BSA mm^2

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Results: UBCV obstruction occurred in 4 of 26 (15.3%) patients in group 1, and in 1 of 47 (2.1%) patients in group 2 (p = .004). The mean duration of catheterization was 8.9 days for group 1 and this was longer in patients who had UBCV obstruction (12.5 days). The demographic data of the patients were demonstrated at Table 1.
Conclusion: This study showed with significantly higher prevalences of obstruction in group 1. Although statistically not significant, SVC indexes of group 1 patients were lower. UBCV catheter insertion in patients with univentricular cardiac anatomy might not be proper. Future studies with larger sample size will help to clarify this problem.

P2-101
Initial Clinical Experiences with Novel Diagonal ECLS System in Pediatric Cardiac Patients
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Purpose: Extracorporeal life support (ECLS) is a lifesaving mechanical circulatory support that has become an accepted therapeutic modality for neonates and children afflicted with respiratory and/or cardiac failure that is refractory to conventional management. The Deltastream® DP3 (Medos Medizintechnik AG, Stolberg, Germany) is a newly designed rotational pump with a diagonally streamed impeller that can be used in children of all ages. The aim of this study was to analyze the safety and feasibility of this novel diagonal ECLS System.
Methods: Between March, 2011 and May, 2015 89 patients underwent ECLS support. We switched to DP3 after November, 2012, and DP3 was used in a consecutive series of 64 patients (almost 5% of all children undergoing congenital heart surgery at same period). We retrospectively reviewed medical records of patients to study.
Results: ECLS with the DP3 was performed in 64 patients (30 females, 34 males). The patients' age was a median of 90 days (2—4320 days). The patients' weight was a median of 3.8 kg (2.1–40 kg). Indications for ECLS were failure to come off of cardiopulmonary bypass (n = 19), postoperative low cardiac output syndrome (n = 18), E-CPR (n = 19), and respiratory failure (n = 8). Nineteen (29.6%) children had single ventricle, while 45 (70.4%) children had two-ventricle pathology. ECMO indications, durations, and start times had no statistical significance on survival. Complications included cardiopulmonary (n = 41.64%), neurologic (n = 18, %28), infectious (n = 27, 42%), pulmonary (n = 19, 29%), renal (n = 34, 54%), hemorrhagic (n = 35, 54%) and mechanical (n = 6, < 10%). Median ECMO duration was 6.4 (range 4–41) days. ECMO was successfully weaned in 45 (70.3%) patients, 24(37.5%) patients survived to hospital discharge and 21 patients (32.8%) died after successful weaning. While the weaning success rate was 36% and the survival rate 20% before November 2012.
Conclusions: The combination of a DP3 pump and a Hilite 800LT oxygenator in pediatric ECMO circuits may improve durability and reduce circuit-induced complications. As a result of shift from DP2 to DP3, revision of ECMO protocol and increased ECMO experience, significant improvement was observed in our clinical results.

P2-102
Inhaled nitric oxide with a nasal cannula and use of phosphodiesterase inhibitor to facilitate weaning from inhaled nitric oxide for children after congenital cardiac surgery
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Objective: After congenital cardiac surgery, pulmonary hypertension complicates the postoperative management. Inhaled nitric oxide is effective for critical pulmonary perfusion. After extubation, inhaled nitric oxide is usually discontinued and rebound pulmonary hypertension is the problem. The purpose of this study was to evaluate inhaled nitric oxide with a nasal cannula and use of phosphodiesterase inhibitor in facilitating its problem.
Methods: Inhaled nitric oxide (10 to 20 ppm) was administered in the case with critical pulmonary perfusion (central venous pressure >20 mmHg or pulmonary hypertensive crisis) during the operation or in the early postoperative period. In seven cases, inhaled nitric oxide could not be discontinued before extubation. In these cases, inhaled nitric oxide with a nasal cannula was continued after extubation and phosphodiesterase inhibitor was also used. Patients ranged in age from 2 months to 2.5 years (median, 6 months) and weighed from 2.5 kg to 12.7 kg (median, 4.5 kg). Three cases were after bidirectional Glenn anastomosis, two case were after total cavopulmonary connection and two cases were after patch closure of ventricular septal defect.
Results: There was no hospital death. No rebound pulmonary hypertension occurred upon inhaled nitric oxide withdrawal in any cases. The time to discontinuation of inhaled nitric oxide with a nasal cannula after extubation raged from 2 days to 26 days (median, 3 days). In a case after total cavopulomary connection, inhaled nitric oxide with a nasal cannula was continued for 26 days because of prolonged pleural effusion. The dose of inhaled nitric oxide ranged from 1 to 10 ppm (median, 3 ppm). Phosphodiesterase inhibitor (sildenafil in four cases or tadalafil in three cases) was administered before or after extubation. The dose of sildenafil ranged 0.5 mg/kg to 1.5 mg/kg and the dose of tadalafil was 1 mg/kg. All but one cases need oral phosphodiesterase inhibitor after discharge. No toxic side effect was observed in any cases.
Conclusion: Inhaled nitric oxide with a nasal cannula and use of phosphodiesterase inhibitor can facilitate weaning from inhaled nitric oxide for children after congenital cardiac surgery with critical pulmonary perfusion.

P2-103
The Cardiovascular Effects of Erythrocyte Transfusion in Critically ill Children
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Introduction: There are many studies concerning that limited eryhtrocyte transfusion strategy decreases the mortality and organ

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insufficiency however studies about oxygen delivery to the tissues are rare in number. In our study we aimed to evaluate the effects of erythrocyte transfusion on tissue oxygen delivery and cardiovascular system in acute phase.
Material and Methods: Patients were divided into 2 groups (Group A: Hemoglobin < 7 g/dl and Group B: Hemoglobin 7-10g/dl). Echocardiographic measured cardiac output, serum lactate, hemoglobin levels were recorded before transfusion. Then initial, 1st, 2nd, 3rd and 4th hours' oxygen saturation (SpO2), perfusion index, noninvasive total hemoglobin (SpHb), oxygen content (Spoc), heart rate and bloodpressure values were measured. Cardiac output, hemoglobin and serum lactate levels were repeated also after finishing transfusion.
Results: Seventytwo cases were included in the study (40 female, 32 male). Mean age was 63.3 ± 57 months. Hemoglobin levels of 22 cases were < 7 g/dL and the rest of 50 cases were between 7-10 g/dL idi. Median hemoglobin values in Group A was 7.9 ± 0.78 g/dl, in Group B was 6.12 ± 0.65 g/dl. 44 patients had mechanical ventilation support; 5 of those were in Group A, rest 39 of them were in Group B. The ones without mechanic ventilation support: 11cases were in Group B, 17 were in Group A. The level of hemoglobin that was required for tranfusion was 7.8 ± 1.1 g/dl for the ones had mechanical support and 6.8 ± 0.9 g/dl for the others that didnot have ventilation support. It was significantly high in ventilation group ($p < 0.001$).
In each group cardiac output, noninvasive hemoglobin (SpHb), oxygen content (Spoc), serum lactate levels were measured pre and posttransfusion. There was a significant increase after transfusion ($p < 0.01$, $p < 0.01$, $p < 0.01$, $p = 0.009$). But perfusion index was significantly increased in Group A ($p < 0.01$).
Conclusion: Limited transfusion strategy had positive effects on microcirculation and cardiovascular system. Also when adverse effects of liberal transfusion were considered, limited transfusion was the right transfusion model.

P2-104
S100 a potential biomarker for cerebral injury in neonates and infants undergoing surgery for congenital heart disease - association to perioperative cerebral tissue oxygenation

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Objectives: Neurodevelopmental impairment including fine and gross motor deficits or speech and language problems are common in children with congenital heart defects (CHD) requiring palliative or corrective surgery as neonates or infants. Identification of risk factors, monitoring techniques and specific and sensitive markers for perioperative neurologic injury are of special interest. We analyzed serum levels of S100, a protein expressed primarily by astrocytes, in children undergoing surgery for CHD within the first year of life. The association between serum levels of S100 and perioperative cerebral tissue oxygenation was evaluated.
Methods: Serum levels of S100 were measured preoperatively and 0, 12, 24 and 48 hours postoperatively using electrochemoluminometric immunoassay (ELECSYS, Roche®). Cerebral tissue oxygenation (cSO2) was derived by near infrared spectroscopy (INVOS, Covidien®) for 12 hours preoperatively, intraoperative and for 48 hours postoperatively.
Results: S100 and cSO2 were obtained in 32 cases (Repair of Tetralogy of Fallot, n = 6; Arterial switch operation, n = 7; Norwood procedure, n = 5; Hemifontan procedure, n = 5; Aortic arch repair, n = 2; others, n = 7). Median age at surgery was 82 (1-295) days, median weight 5.2 (2.8-9.2) kg. The median duration of cardiopulmonary bypass was 129 (49-208) minutes. None of the patients showed neurologic symptoms during the perioperative course. Preoperative S100 was 0.20 ± 0.10 µg/l. Highest S100 levels were measured directly after surgery (0.88 ± 1.22 µg/l). Overall, S100 declined to preoperative levels in the postoperative period (S100 48 hours post OP: 0.22 ± 0.11 µg/l). However, 48 hours after surgery, S100 was >0.2 µg/l in 17 cases. In those, mean cSO2 values were significantly lower in the early postoperative course (first 4 postoperative hours: 54 ± 13% vs. 68 ± 14%, $p = 0.007$) and within the first 12 hours (58 ± 14% vs. 71 ± 10%, $p = 0.005$). Preoperative and intraoperative cSO2 were not different (59 ± 11 vs. 62 ± 11%, $p = 0.435$ and 70 ± 14% vs. 63 ± 10%, $p = 0.157$).
Conclusion: Infants younger than 1 years of age undergoing surgery for CHD who did not show a decline of serum S100 levels to preoperative levels had lower early perioperative cerebral tissue oxygen saturations. Neurocognitive outcome still needs to be determined, but S100 may serve as a surrogate for subtle cerebral injury.

P2-105
Restrictive enlargement of pulmonary annulus at surgical repair of tetralogy of Fallot: 10 year Follow Up study

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Objectives: Since 1996 our center follows a uniform strategy of restrictive enlargement of the pulmonary annulus at surgical repair of tetralogy of Fallot (ToF). A transanular patch (TAP) is only used if the z score of the pulmonary annulus (PV) is < -4. The rate of TAP was significantly reduced accepting a significantly smaller PV. Whether this strategy leads to reduction of pulmonary insufficiency (PI) and re-operation rate in the long-term has not been studied.
Methods: 95 ToF patients who had their repair between 1996 and 2006 were included in the study. Clinical, echocardiographic, ECG and cardiac MRI data were collected. The cohort was compared to a historic cohort of 110 patients, who had their repair between 1975 and 1996.
Results: 6 patients were lost to Follow Up. Follow Up since repair was 12.6 (5.9-19.4) years. 28 patients (31.4%) needed a TAP. Patients were in a good clinical condition (NYHA (1/2): 78/11) with a mean QRS of 130 ± 21 ms. On echocardiography all patients showed a mild pulmonary stenosis (PS) with a mean Vmax 2.4 ± 0.63 m/s, while 38% showed moderate or severe PI (PI (none/I°/II°/III°: 17/37/32/1). CMR derived volumes were: EDVi 110.4 ± 20.9 ml/m^2, ESVi 55.6 ± 15.8 ml/m^2 and EF 48.8 ± 12.1%. 7 patients needed a re-operation for PS (7.9%) and 5 for PI (5.6%). In 3 patients the pulmonary valve had to be replaced (PVR) in the first re-operation, altogether 10 PVRs were performed in 5 patients. Freedom from re-operation of the PV was 89.3% at 10 years Follow Up. In the historic cohort (69% TAP) significantly more patients needed a re-operation for PI (5/89 vs. 20/110, $p = 0.005$).
Conclusion: The re-operation rate for pulmonary insufficiency is significantly lower in patients with a restrictive enlargement of the pulmonary annulus compared to a historic cohort. To proof

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whether this result holds true when compared to a cohort of patients operated in the same time period but without following a uniform strategy a control group from the Kompetenznetz Angeborene Herzfehler will be recruited.

P2-106
Reduced antithrombin-III-activity in patients with early thrombus formation after Fontan-operation

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Introduction: There is a major risk for thrombus formation (TF) inside the extracardiac conduit and both pulmonary arteries after Fontan-operation. It may configure in the early postoperative period, despite anticoagulation with heparin. Disorders of the coagulation system are possible promotive factors.
Methods: In a retrospective study we analysed the AT-III-a before and after Fontan-operation in n = 29 consecutive patients without and n = 6 with early postoperative TF.
Results: The preoperative AT-III-a was normal (100% ± 11.4%) without differences among both groups (p = 0.16).
Immediately after operation AT-III-a decreased slightly (77% ± 10%), again with no significant difference between the two groups (p = 0.54).
Six to eight hours after the operation we observed a major decline of AT-III-activity (56.2% ± 10.9%) in patients with TF inside, while it fell only marginally in patients without TF (74.3 ± 11%). The difference between both groups was significant (p = 0.014).
There was also a difference in AT-III-a among patients with and without TF in the morning of the 1st (62.2 ± 12.7% vs. 71.2 ± 14.9%) and 2nd (50.5 ± 14.3% vs. 64.0 ± 10.4%) postoperative day, but the dissimilarity was not significant.
In all patients with TF we could not reach a therapeutical aPTT during the first postoperative day, despite high doses of heparin.
Conclusions: AT-III-a is reduced in some patients early after the Fontan-operation. This plays an important role in the pathogenesis of early postoperative TF inside the conduit or central pulmonary arteries.
Thus, AT-III-a has to be controlled and if necessary substituted, to attain a sufficient early postoperative anticoagulation.
Because argatroban is inhibiting thrombin directly and – in contrast to heparin - independent from AT-III-a, it may be an alternative to AT-III substitution.

P2-107
Cone repair of tricuspid valve in Ebstein's anomaly: own experience of anatomical reconstruction

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Ebstein's anomaly - a congenital heart defect in which the septal leaflet of the tricuspid valve (TV) is displaced into the cavity of the right ventricle toward its apex. Its prevalence is 0.5-1% of all congenital heart defects (CHD). The main cause of death I this patients is a heart failure. Complications are serious arrhythmias, which can also lead to sudden death.
Objective: To analyze our own experience of successful cone reconstruction of TV in Ebstein's anomaly.
Methods: From 2012 to 2015 in Ukrainian Children's Cardiac Center 17 patients were operated with Ebstein's anomaly. It was apply the new method for the cone reconstruction of TV.The mean age of patients was 8.6+ 2.7 years. Preoperative diagnosis was made by the anatomy of the tricuspid valve: type 'A' in 3 patients, type 'B' in 6 patients, type 'C' in 5 patients, type 'D' in 3 patients. All patients were conducted electrophysiological study (if necessary, conducted high-frequency catheter ablation) and a cardiac catheterization before surgery. 6 patients had cone reconstruction combined with the Glenn shunt because of right ventricle failure.
Results: Early postoperative mortality was 11.7% (2 patients). Average postoperative follow-up was 7.5 + 2.8 months. During the follow-up visit the clinical condition of the patients was examined, tests with exercises stress were conducted to assess functional status, evaluated data of ECG, echocardiography, chest X-Ray and magnetic - resonance imaging of the heart. As a result of control there is a from mild to moderate insufficiency on the TV, contractile function of the left ventricle is good and mild decreased right ventricle's contractility, indicating good result of correction. In late postoperative period reoperation was performed in 3 patients through a severe tricuspid insufficiency.
Conclusions: Methods of cone reconstruction of TV is a new and effective technique of surgical anatomical treatment of Ebstein's anomaly.

P2-108
Management of low cardiac output following fetal aortic valve dilatation and neonatal Ross-Konno procedure. Report of two cases

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The postoperative treatment of newborn patients with critical aortic stenosis, endocardfibroelastosis and biventricular circulation has to deal with problems caused by a restrictive and stiff left ventricle. This leads to an elevated end-diastolic pressure, an increase in the left atrial and pulmonary artery pressure causing a low cardiac output syndrom (LCOS). Patients, who underwent a fetal aortic balloon dilatation have particular needs because they can develop significant LV hypertrophy with a good systolic function generating a high gradient over the aortic valve but with a impaired diastolic function. After a neonatal Ross-Konno procedure to relief LV outflow obstruction, some of these patients have to go on ECMO due to LCOS. To wean them off ECMO usually positive inotropic catecholamines like epinephrine and dobutamin are applied.
We present two particular cases from our Center with critical aortic stenosis after fetal aortic balloon dilatation at 30 weeks of gestation. In both cases the left ventricle regained its systolic function after the fetal intervention and was again able to generate a high gradient over the aortic valve. Following a neonatal Ross-Konno procedure both patients had to go on ECMO due to LCOS. The weaning from ECMO was successful only by using lusinotropic drugs like milrinon and levosimendan instead of catecholamines. In the case of the first patient three weaning attempts failed using catecholamines to treat LCOS. After three cycles of levosimendan and 19 days on ECMO the fourth attempt was successful using only low doses of dobutamin and milrinon but no epinephrine at all. In the case of the second patient levosimendan was administered on ECMO and - after seven days - weaning was successful using again only lusinotropic drugs like milrinon and nitroprussidnatrium but no epinephrine.

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Fetal aortic balloon valvuloplasty may lead prenatally to significant LV hypertrophy with good systolic but impaired diastolic function, which postnatally needs tailored treatment with lusinotropic medication avoiding inotropic catecholamines.

P2-109
Long-term survival after pediatric ECMO therapy

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Objectives: Extracorporeal membrane oxygenation therapy (ECMO) is commonly used in pediatric heart centers to treat cardio-pulmonary failure which occurs perioperatively or primarily e.g. in cases of cardiomyopathy. In 2007 a pediatric ECMO program was introduced in our center. This study shall examine the short and long-term survival rate of this method and identify potential risk factors for mortality.
Methods: Between 2007 and 2014 86 children were treated with arterio-venous ECMO therapy at our center, 7 of them without previous heart operation. The age of the patients varied between 1 day and 16 years (median 26 days), 47 patients were newborns. Median weight was 6.7 kg (2.2-68 kg). 45 patients had univentricular anatomy, most of them after stage 1 palliation. 41 patients had biventricular anatomy. In 57 cases the indication for ECMO installation was postoperative cardiac failure (including 3 cases of cardiopulmonary resuscitation in the long-term follow up), in 29 cases the indication was pulmonary failure (e.g. severe postoperative cyanosis, suprasystemic pulmonary hypertension, pulmonary infection or diaphragmatic hernia). In 37% of the cases ECMO was implanted during CPR.
Results: 66 out of 86 patients could be weaned off ECMO (76.7%), but in the long-term follow up only 48 patients survived (55.8%). Children with pulmonary indication had a significantly higher survival rate versus the cardiac indication. (79.3% vs. 43.8%; $p < 0.05$). ECMO duration < 12 days showed significant advantage versus prolonged duration (61.8% vs. 10%; $p < 0.05$). Children with single ventricle physiology, newborns, patients with CPR and implantation after the first pod showed worse outcome, but without statistical significance.
Conclusions: The fact, that three quarters of the patients survived a potentially life threatening period of cardio-pulmonary failure due to ECMO therapy, verifies this method as an indispensable postoperative instrumentarium. In the long-term follow up a considerable late mortality and complication rate shows up. In our series we could only identify cardiac indication and longer ECMO duration as significant risk factors.

P2-110
Coronary artery anomalies and their impact on the outcome of the arterial switch operation for the transposition of the great arteries: 25 years' experience

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introduction: Successful coronary translocation was the major issue at the beginning of introduction of arterial switch operation and still remains a challenge in case of complex coronary pattern. The aim of this study was to describe the frequency and anatomy of the coronary anomalies associated with the transposition of the great arteries and their impact on the outcomes of the arterial switch operation.
Methods: For this retrospective case review study we included all 716 patients TGA who underwent ASO in our institution between years 1991-2015. All of the surgical procedures were performed by one cardiac surgery team lead by JJM, using the same surgical technique with his own modifications. The anatomy of the coronary arteries was obtain from surgical protocols and confirmed by further routinely performed coronarography or coronary CT angiography.
Results: Coronary anomalies were present in 227 patients (31.7%), among their 27 different types detected in our study group, circumflex arising from right coronary artery and looping behind pulmonary artery, occurred most frequently (98 patients, 13.7%). Intramural pattern of the coronary artery was detected in 25 patients (3.5%). In 99 patients, there was a complex coronary transfer – to the one sinus, very close to the commissures or high above the sinus, in one patient with common ostium of the coronaries placed anteriorly to the aorta, pericardial tube was used to facilitate proper coronary translocation. As the most of coronary anomaly types were extremely rare and didn't exceed 1% of our cohort, for the data analysis they were grouped into 5 subgroups (1: intramural course; 2: single ostium; 3: anterior looping; 4: posterior looping; 5: anterior and posterior looping), complex coronary transfer was taken to the analysis as the additional risk factor. In multivariate logistic regression model none of the observed anomalies were significantly related to the early mortality, early and late reoperations or catheter interventions. Coronary anomalies were significantly correlated with Taussig Bing anomaly ($p < 0.001$), non-facing commissures ($p = 0.013$) and significant discrepancy of aortic and pulmonary valves ($p = 0.033$).
Conclusions: Currently coronary anomalies associated with TGA didn't impact on the early results of the arterial switch. The further fate of the transferred coronaries, especially in case of their complex anatomy still remains uncertain.

P2-111
Early Repair of Tetralogy of Fallot and Postoperative Outcomes

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Introduction: While mounting data indicate a significant risk for postoperative morbidity when surgical repair of Tetralogy of Fallot (TOF) is performed in neonates, there is far less consensus in terms of the optimal timing for repair beyond the neonatal period. Particular concern remains with regard to the postoperative care and the risk for reoperation in patients with repair before the age of 3 months.
Methods: 207 consecutive patients with TOF referred to our institution for non-conduit repair between 1996 and 2013 were reviewed for neonatal, demographic and postoperative data as well as data regarding cardiac reoperation during follow-up. Patients with repair below 3 months of age (Group A) were compared with the remaining cohort (Group B).

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Results: Thirty-one patients had repair before the age of 3 months (Group A). There was no mortality in Group A. One patient in Group B died 6 months after repair. There were no differences between the groups with regard to prematurity ($p > 0.9$), birth weight ($p > 0.5$) and chromosome anomalies ($p > 0.9$). BT shunt was used in 1 patient in Group A and in 47 in Group B ($p = 0.01$). During repair, transannular patch was used in 17 patients (55%) in Group A and in 93 patients (53%) in Group B ($p > 0.9$). Of these, monocusp valve was used in 9 (29%) and 46 (26%) patients, respectively ($p = 0.8$). There were no differences between the groups in ventilation time, ICU stay, pleural effusion drainage and hospital stay ($p > 0.5$). During the follow-up (median duration 9 ± 8 years in Group A and 11 ± 9 years in Group B; $p = 0.1$), 1 patient (3%) in Group A and 32 patients (18%) in Group B required reoperation with pulmonary valve replacement (PVR) ($p = 0.07$). Six patients (19%) in Group A and 6 patients (3%) in Group B required reoperation due to RVOT obstruction ($p = 0.02$).
Conclusion: The postoperative hospital resource utilization and the risk for later PVR in patients with TOF repair under the age of 3 months are comparable to those in patients with repair beyond this age. Reoperation due to RVOT obstruction appears to be more common in patients with early repair.

P2-112
Porcine pulmonary prostheses versus bovine jugular vein to repair the dysfunctional right ventricular outflow tract in children and teenagers

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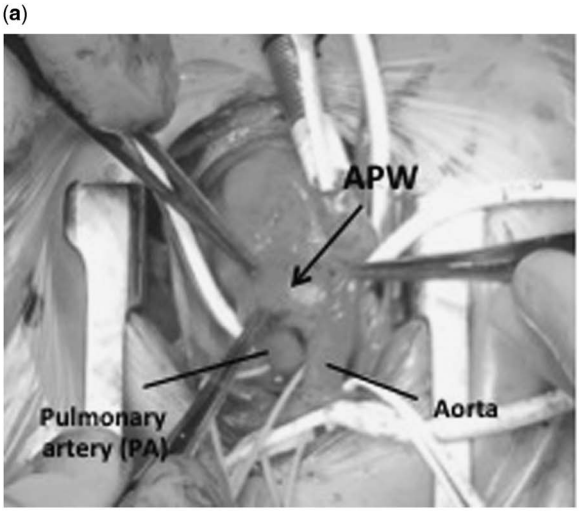
Objectives: Residual dysfunction of the right ventricle outflow tract (RVOT), due to congenital reconstructive surgery, is usually reoperated into adulthood. Sometimes, symptoms and/or dysfunction of the right ventricle (RV) during childhood may condition an earlier pulmonary valve replacement .
Our target is to compare the results of the valved bovine jugular vein (BJ) versus the stented porcine pulmonary prosthesis (PPP), implanted in patients aged 1 to 18 years old.
Methods: All reoperation performed for prosthesis interposition (BJ or PPP), in patients aged 1-18 years with dysfunction of the RVOT after previous congenital cardiac surgery. Study period 2003-2015. Prosthetic dysfunction criteria: surgical/percutaneous reintervention, gradient >50 mmHg or severe prosthetic regurgitation. Statistical Analysis with SPSS 20.0.
Results: 21 PPP/20 patients and 15 BJ/15 patients. 60% male. Fallot, most common primary disease in both groups. From 24 preoperative variables studied, statistically significant differences occur in 11, highlighting previous surgery on RVOT ($p < 001$, more transannular in PPP group), degree of previous pulmonary regurgitation (PR) ($p = 0.011$, more PR in PPP), RV function ($p = 0.016$, lower in PPP) and RV diastolic volume ($p = 0.026$, more dilated PPP group)
No hospital mortality. From 15 perioperative variables, significant differences in: average age of the implant ($p = 0.044$, lower in BJ group, 8 vs 11 years), implanted valve diameter ($p < 0.001$, lower in BJ group) and need of aortic cross-clamping ($p = 0.015$ higher in BJ).
No late mortality. BJ mean follow-up 4.8 years, vs 2.4 in PPP group ($p = 0.046$).
From other 16 postoperative variables, were also statisticaly significant peak transprosthetic average gradient ($p < 0.001$), degree of residual PR ($p = 0.009$) and prosthetic dysfunction ($p = 0.006$, 60% BJ vs 23% PPP), ever favourable for PPP group.
Conclusions: Although only a longer follow-up will confirm the hipothesis, it seems reasonable to chose a PPP to improve RVOT functionality in children above 1 year, being significantly lower its percentage of dysfunction in tracking.

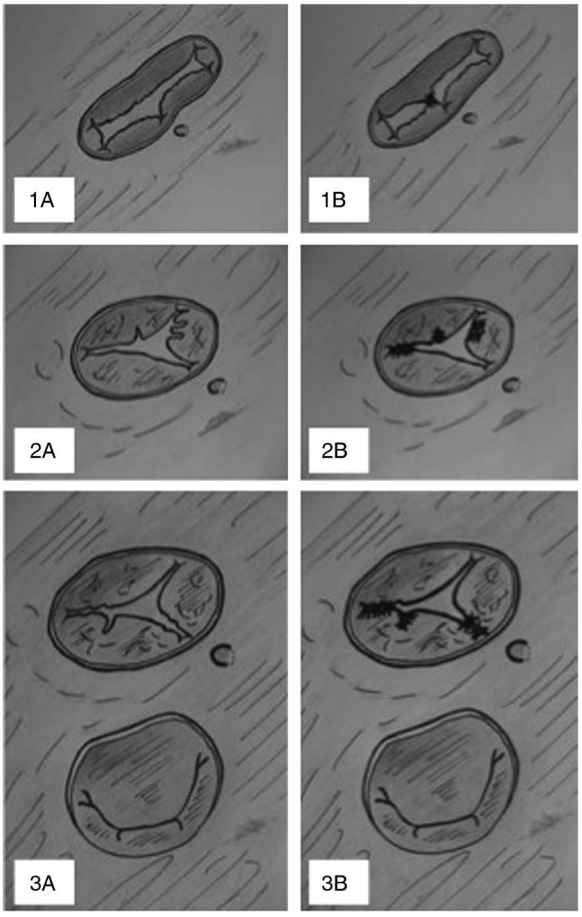
P2-113
A 10-year experience with aortopulmonary window repair in children: an uncommon anomaly with an excellent outcome

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Background: Aortopulmonary window (APW) is rare and single institutional experience is low. The purpose of this study is to describe our approach to the management of APW and its associated lesions and to assess the outcomes over a 10-year time period.
Patients/Methods: We conducted a retrospective review of all children (n = 8, 50% males) who underwent surgical repair of APW between 2006 and 2015. Clinical features, surgical approach and early and late outcomes are included.
Results: Median age at surgery was 19 days (range 4-30) for 6 neonatal patients. Two patients were older than 1 month (3.5 month and 6 year-old respectively). Median weight was 4.9 kg (range 2.5-15.7). Type I (proximal) APW was detected in 5 and type II (distal) in 3 of them. Simple APW was present in 3 patients (37.5%) and 5 patients (62.5%) had concomitant cardiac defects. Associated cardiac lesions (n = 6) included interrupted aortic arch type A (n = 2), perimembranous ventricular septal defect (VSD) (n = 1), atrial septal defect (ASD) (n = 2) and congenital mitral valve regurgitation (n = 1). The APW was repaired by direct closure in 2 patients (25%) and transaortic patching (bovine or autologous pericardial patch) in 6 patients (75%). Single-staged repair of APW and aortic arch repair (n = 2), VSD closure (n = 1), ASD closure (n = 2) and mitral valve repair (n = 1) was performed in those patients with associated anomalies. Cardiopulmonary bypass time was 81 +/-22 minutes (range 54-120); aortic cross-clamping time was 35 +/-21 min (14-57). Median length of stay (intensive care unit) was 9 days (range 4-19). Operative/in-hospital mortality was zero. Median follow-up was 4.3+/-2.5 years (range 1-7): overall survival was 100%; freedom from reoperation was 87.5% (n = 7) (1 patient underwent mitral valve replacement 3.5 years following surgery); freedom from percutaneous procedures was 87.5% (n = 7) (1 patient required ballooning due to supravalvular aortic stenosis 7 months following surgery). All patients demonstrated normal pulmonary artery and aortic growth, normal pulmonary pressures, no residual shunts and NYHA functional class I/II.
Conclusions: Despite being a rare anomaly, the current strategies for APW repair using cardiopulmonary bypass are associated with excellent outcomes. A single-staged repair of associated cardiac anomalies is advised.

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P2-114

Atrioventricular valve repair concomitant with Fontan procedure: 'simple' surgical strategies for single-ventricle patients

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Introduction: Atrioventricular (AV) valve regurgitation is widely known as a risk factor for the Fontan completion in patients with univentricular physiology. To date, indications and timing for the AV valve repair remain unclear and different surgical techniques have been advocated. We present our initial experience by using individualized 'simple' techniques for repairing the AV valve along with Fontan completion.
Patients/Methods: Since 2013-2015, 53 consecutive patients underwent extracardiac Fontan completion in our institution. AV valve repair (avoiding the use of prosthetic ring) was performed as concomitant procedure in three of them due to moderate or severe tricuspid regurgitation. Case 1: 9 year-old, 26 kg. Diagnosis: unbalanced atrioventricular septal defect. AV valve: common AV valve. Procedure: closure of the left-side cleft (cardioplegic arrest) (1 A/1B). Case 2: 5 year-old, 15 kg. Diagnosis: hypoplastic left heart syndrome. AV valve: tricuspid. Procedure: the antero-septal commissure was closed, as well as two gaps in the posterior valve (beating-heart repair) (2 A/2B). Case 3: 5 year-old, 18 kg. Diagnosis: double outlet right ventricle, non-committed ventricular septal defect. AV valve: mitral and tricuspid. Procedure: both anteroseptal and posteroseptal tricuspid valve commissures were closed (cardioplegic arrest) (3 A/3B). In the three cases, following the valve repair, the Fontan pathway was completed uneventfully with an extracardiac 18-20 mm conduit.
Results: On-table echocardiography, trace-to-mild regurgitation was noticed for all the cases. No post-operative complications were detected. Patient 1 underwent percutaneous embolization of venous collaterals on early post-operative period. Average in-hospital stay was 19+/-6 days. At follow-up (18+/-4 months), they remain asymptomatic with unchanged echocardiography findings (trace-to-mild regurgitation).
Conclusion: The AV valve repair concomitant with Fontan procedure when moderate or severe regurgitation is present may be performed at a low risk with excellent short-term results. Beating-heart surgery can be accomplished in selected cases and surgical techniques should be simple and reproducible, avoiding AV valve replacement in these single-ventricle patients.

P2-115

A 10-year experience with arterial switch operation in a single medium-volume institution: improving outcomes over the years

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Background: Several factors have been identified as carrying additional risk in patients with transposition of great arteries (TGA) following arterial switch operation (ASO). Due to the evolving surgical strategies/perioperative management, the purpose was to analyse the outcomes and morbi-mortality trends at our institution.
Patients/Methods: Between 2006-2015, 115 ASO were performed [age 11+ /-7 days (range 1-150); weight 3.2+ /-0.3 kg (range 1.8-7.4)]. 'Simple'-DTGA 59.1% (n = 68); DTGA+ CIV/DORV 30.4% (n = 35); Taussig-Bing 8.7% (n = 10); DTGA+ partial AVSD 0.8% (n = 1); DTGA+ hypoplastic aortic arch 0.8% (n = 1). Surgical era/impact of preoperative factors on early mortality and follow-up analysis were conducted.
Results: Single-stage repair was performed (all patients). The coronary pattern was: normal (type A/D) (80.8%, n = 93), single coronary artery (9.5%, n = 11), intramural pattern (3.4%, n = 4), inverted pattern (0.8%, n = 1), single sinus pattern (2.6%, n = 3), others (2.6%, n = 3). Cardiopulmonary bypass time: 212.6 +/-34.5 min (range 134-448). Aortic-cross-clamping time: 111.9 +/-23.2 (range 55-270). Peri-/postoperative ECMO: 17 patients (14.7%). PICU stay: 18+/-4.5 days (range 5-179). Mechanical ventilation duration: 192+/-87.8 hours (range 48-1752). Delayed sternal closure: 62.6%. 30-day mortality: 13% (n = 15) (causes: severe ventricular dysfunction and/or refractory hypoxemia). 30-day mortality was influenced by surgical era and factors showed in the table. In-hospital percutaneous procedures (6 in 5 patients, 4.3%): ballooning (neopulmonary/PA's/neoaorta) (n = 5), coarctation stenting (n = 1). In-hospital reinterventions (8 in 5 patients, 4.3%): left coronary artery reconstruction (n = 1), aortic/mitral repair (n = 3), mitral replacement (n = 2), LV-free-wall rupture repair (n = 1), pacemaker implantation (n = 1). Median in-hospital stay: 28.8+/-9.3 days (range 11-128). Median follow-up: 59.1 +/-31.8 months (range 2-120). Overall survival (100 hospital survivors): 99%; one mortality case 3 months following ASO (severe biventricular dysfunction). Freedom from catheter-based interventions at 2, 5 and 8 years: 91%, 76% and 73% [ballooning/stent RVOT (n = 12)]. Freedom from re-interventions at 1 and 5 years: 94% and 89% [RVOT/PA patching (n = 4)]. Significative lesions (last follow-up): 13 patients: moderate aortic/pulmonary regurgitation (n = 7), moderate PA's stenosis (n = 2), multiple VSD (n = 2), moderate supravalvar pulmonary stenosis (n = 1).
Conclusions: Early outcomes following ASO have improved in recent years. Older patients or those with associated lesions/abnormal coronary pattern may not be at increased risk of death at medium-volume institutions in the current era. Medium-term outcomes are excellent, but follow-up focused on right-sided lesions is needed.

Variables – 30-day mortality, n (%)	2006-2010	2011-2015	p
Age >21 days	2 (2.7)	0 (0)	0.04
Associated cardiac/great vessels lesions	5 (6.9)	2 (4.6)	0.038
Abnormal coronary pattern	4 (5.5)	0 (0)	0.0021

P2-116
Aortic valve sparing surgery in congenital heart disease

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Objectives: Aortic valve lesions requiring surgery in patients with congenital heart disease are a real challenge. These valves present more complexity in comparison with acquired valve lesions, owed to more severe structural anomalies of the valve itself, and the wide range of size of the patients from infant to adult. In order to delay the time of the valve resection and prosthesis interposition, our first surgical option in each patient is trying to spare his aortic native valve. We present here our experience with conservative aortic valve surgery in our congenital heart unit.
Methods: Retrospective study:27 opertions performed in 26 patients, during 2010-2015. Statistical analysis with SPSS 15.0.
Results: Median age 7 years. (IQR 0.6-12), and 37% were ≤6 moths. Previous percutaneous aortic valvuloplasty was done in 26%. Clinical symptoms: asymptomatic 59%, dispnea 30%, angina 7%. Aortic valve anatomy: bicuspid 48%, monocuspid 11%, cuadricuspid 4%. Aortic valve main functional pathology: stenosis 11 (41%), insufficiency 6(22%) and double lesion 5(18%).
Operations were performed with cardiopulmonary by-pass (CPB) and aortic clamp (AoC), with transaortic approach. Surgical techniques employed comissurotomy in 5(18%), aortic leaflet plasty in 7(26%), David operation in 4(15%). Associated surgery was done in 22(81%) patients, mainly consistent in closing a ventricular septal defect and subaortic membrane resection. Median CPB time 96 minutes (IQR 77-185) and AoC time 67 minutes (IQR 55-143). Hospital mortality 1(3.7%) patient. Median intubation time 7 hours (IQR 3-52); median intensive care unit stay 4 days (IQR 3-6) and median hospital stay 8 days (IQR 6-14).
Follow-up is complete, median 16 months (RIC 4-29). There is no late mortality, and 2 patients were reoperated during this time. Nowadays, the majority of our patients are asymptomatics, with normal function of their aortic valve.
Conclusions: Aortic valve sparing surgery in patients with congenital heart disease presents more complexity related to valve anatomy, the size of the patient and the associated pathology. If we achieve the aortic valve conservation, our results are good related to short and medium term follow-up

P2-117
Outcomes after surgery for Anomalous Origin of The Left Coronary Artery From The Pulmonary Artery In Infants

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Introduction: Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is one of the most common causes of myocardial ischemia in children. Excellent results are obtained by early surgical correction. We sought to determine outcome after surgical correction of ALCAPA.
Methods: Data from 26 consecutive patients with ALCAPA who underwent coronary reimplantation between 1993 to 2013, were retrospectively analyzed. Surgery re-established a two-coronary system in all. Concomitant mitral valvuloplasty was performed in 2. Initial and follow-up echocardiographic data were analyzed in order to assess LV function and mitral regurgitation.
Results: The median age at repair was 3.4 [2.2-16] months, median weight was 5.6 [4.2-7.9] kg. The median preoperative left ventricular ejection fraction (LVEF) was 28% [20-40] and 67% of our patients (n = 17) presented with LVEF < 35%. Young age at presentation was significantly correlated to impairment of LVEF (p = 0.001). Mitral insufficiency was moderate to severe in 10 patients (38.5%). Extracorporeal membrane oxygenation (ECMO) support systems were required in 2 cases (7.7%) before surgery and in 5 cases (19.2%) after correction. There was

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significant correlation between age and ECMO requirement ($p = 0.02$).The median follow-up time was 63 [30-153] months. There were 3 postoperative death (at day 7, 60, 120 post-operatively, in children respectively aged 7 days, 3.5 and 4 months) with an overall mortality rate of 11.5%. Six months after the operation, LVEF had improved to a median of 55% [50.5-61]. Only 1 had persistent LV dysfunction that only normalized after 2 years. At latest follow up, the median ejection fraction was 65% [61.5-70]. Regression of mitral regurgitations (MRs) was noted in all patients. There were no early or late reinterventions on the reimplanted coronary arteries.
Conclusions: ALCAPA is a severe disease and young age is significantly associated with a more severe presentation and more ECMO requirement. This may be explained by lesser development of coronary collateral circulation. After establishment of a two-coronary circulation both LVEF and MR tend to rapidly normalize over time in most patients.

P2-118
Idiopathic dilatation of right atrium in young children: case report of 7 successful repairs
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Objectives: idiopathic dilatation of right atrium is an extremely rare congenital heart disease which is characterized by significant increase of right atrium in absence of other heart malformations. The aim of our study was to present successful right atrium enlargement repair in 7 young children.
Methods: 7 children with idiopathic dilatation of right atrium (RA) was operated at the median age from 1 month to 5 years, median age 5 months, Interquartile Range (IQR) 14 months, BSA 0.385 m^2 (IQR 0,43) in Bakoulev SCCVS from 01.2005 to 12.2013. Examination included routine physical cardiovascular examination and X-ray finding, ECG with 12 leads and 24 hours monitoring, contrast-enhanced MSCT, EchoCG and angiography.
Results: Right atrium enlargement was diagnosed on 30-32 gestation age (weeks) in four of seven infants, two of them were newborns. Heart failure was preponderous in the clinical picture for all patients, and right atrium and ventricle overload was diagnosed by 12 lead ECG and X-ray finding. NYHA class was 3 (IQR 1). Cardio-thoracic index ranged from 0.57 to 0.91 (median 0.74, IQR 0.12). Arrhythmia in the form of paroxysmal atrial flutter was up to 300 beats/min in 1 of 7 patients and one of 7 infants was diagnosed with supraventricular trigeminy. According to MSCT findings RA volume varied from 145 ml/m^2 to 322 ml/m^2 (median 215.7 ml, IQR 113.7). Partial resection of right atrium to the extent of normal tissue by cardiopulmonary bypass during moderate hypothermia was performed to all patients. Patient mortality was 0%. Morphological examination of resected RA portion uncovered high-grade dysplasia of atrial myocardium with diffuse thinning, endocardial fibroelastosis and local muscle layer atrophy. One of 7 cases was accompanied with calcareous degeneration patch. Three of seven children were under long-term follow-up from 3 months and up to 4 years (median 16 months, IQR 24). Late outcomes were successful.

P2-119
Short-term results of surgical treatment of Coarctation of the aorta with small left ventricle
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Goal: to perform 10-years experience of treatment infants with coarctation of the Aorta
Materials and methods: retrospective analysis of 880 patients of the 1st year of life with Aorta Coarctation, who had no significant septal defects, treated in Bakoulev center. Among all operated infants 7,5% were newborns (n = 66) with left ventricle end diastolic volume index (LV EDVi) less then 30 ml/m^2, where Z-score of at least one of LV structures were lower than -1.65. Age at the hospitalization time ranged from 4 hours to 28 days of life (av. 8 ± 7.5 days), weight 1.65-4.86 kg (av. 3.23 ± 0.71), length 40-62 cm (av. 51 ± 4.2), Z-score LV end diastolic size (Z LV EDS) from -6.63 to -0.51 (av. -2.45 ± 1.22); Z-score mitral valve (Z MV) from -3.91 to -0.15 (av. -1.71 ± 0.91); Z-score aortic valve (Z AV) from -5.09 to -0.91 (av. -0.77 ± 1.12). 28 newborns (42.4%) had entered hospital in critical condition. We operated 3 premature newborns from 26 to 32 weeks of gestation. Only 5 newborns (7.8%) had Coarctation with Aortic stenosis, before resection of the Coarctation we performed aortic valvuloplastics.
Results: 34 newborns (51.5%) had complicated postoperative period, among which we observed mainly heart failure (n = 28, 42.4%), respiratory failure (n = 16, 24.2%) and necrotic enterocolitis (n = 8, 12.1%). Progressive heart failure caused death of 3 pts., in other 7 cases it was generalization of the infection. Total mortality rate was 15.2% (n = 10). There were no reliable influence of age, z-score AV, AV stenosis and ASD size on surgical results. Newborns with LV EDVi < 22.5 ml/m^2 (33.3%), weight less 2.8 kg (37.5%) and who had critical condition before operation (29.6%) had higher mortality.
Conclusion: z MV < -1,7 ($p = 0.009$) is a predictor of postoperative heart failure. LV EDVi < 22.5 ml/m^2 (33.3%), weight less 2.8 kg (37.5%) and critical condition before operation reliably increases the mortality rate ($p = 0.015$).

P2-120
Evaluation of a light activated and elastomeric tissue sealant for vascular surgery
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Objectives: Tissue adhesives (TA) and sealants may offer advantages over sutures during surgery, particularly in the reduction of operative time and procedural simplification, improving patient safety and faster recovery. However, commercially available TA still show limitations such as low adhesive strength and sealing

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performance, limited control over activation and poor biocompatibility. GB02 is a novel hydrophobic light activated sealant developed and provided by Gecko Biomedical (Paris, France) that aims to fill this unmet clinical need. In this study, the performance of GB02 for sutureless closure of vascular incisions is compared with a commercially available sealant.
Methods: 2 mm defects of the carotid artery (CA) and jugular vein were created and closed with GB02 or bovine serum albumin and glutaraldehyde (BSAG) without the use of sutures in an in vivo porcine model. Animals were survived for 7 days and 5 weeks (n = 4 per time point and experimental group). Vessel diameter, wall thickness and flow velocity were determined after 5 weeks by means of magnetic resonance imaging. Histopathology of the explanted vessels was performed after 7 days and 5 weeks.
Results: In vivo closure of the defects was successful in all cases, with immediate hemostasis after clamp release. Neither postoperative bleeding nor thrombi have been observed. For CA closure, there were signs of stenosis for BSAG in contrast to GB02 as revealed by differences in vessel diameter (6.4 ± 0.4 mm for GB02, n = 4 and 4.2 ± 0.1 mm for BSAG, n = 2), wall thickness (0.8 ± 0.1 mm for GB02, n = 4 and 2.2 ± 0.1 mm for BSAG, n = 2) and flow velocity (71.8 ± 0.1 cm/s for GB02, n = 4; no residual flow for BSAG in three cases, 102 cm/s in the fourth case). In two CA closures with BSAG, there was a complete occlusion of the CA. Closure with GB02 was associated with a low inflammatory reaction limited to the adventitia. In contrast, BSAG caused a moderate to severe inflammatory reaction (p = 0.010).
Conclusions: GB02 effectively seals 2 mm defects of vessels. Preliminary results demonstrated no signs of stenosis and a low inflammatory reaction for closure with GB02 in contrast to BSAG. Thus, this study revealed benefits of GB02 for the surgical practice.

P2-121
iNO delivery and monitoring during different ventilation modes

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Introduction: Complex congenital heart surgery may induce long duration of mechanical ventilation.
This MV has to be protective avoiding VILI during PSV, VCV HFO, NAVA, and NIV. As a high percentage of these patients experiences pulmonary hypertension, the delivery of pulmonary vasodilators has to be available and accurate in all the ventilation modes.
Methods: With a new device able to deliver and monitor inhaled NO, 51 patients were included, with a median of 4 [2 to 6] measurements per patient. Of the 195 measurements, 115 were performed on pressure controlled ventilation, 9 on volume controlled, 3 on high frequency oscillatory ventilation and 68 on non-invasive mechanical ventilation. The Bland Altman methodology was used to evaluate the agreement between the set and the measured iNO concentrations.
Results: The Bland-Altman plot is shown in Figure 1. The measurement error, as assessed by the mean value of the difference, was significant (mean -1.01, [-3.27–1.24] ppm, one sample t-test p value < 0.001), indicating the presence of a fixed bias. We could not identify any proportional bias, and the slope of the linear regression of differences on averages was not significant (p = 0.14). The mean bias on controlled ventilation was -1.13 [-3.65–1.36] ppm and was -0.76 [-2.36–0.84] ppm on non-invasive ventilation.
Comments: There was a -8.6%, 95%CI [-26.0–8.8] difference between the delivered concentration and the concentration set at the device.
Conclusions: iNO delivery is possible safely and accurately in different ventilation modes including PSV, VCV, HFO, NAVA, and NIV.

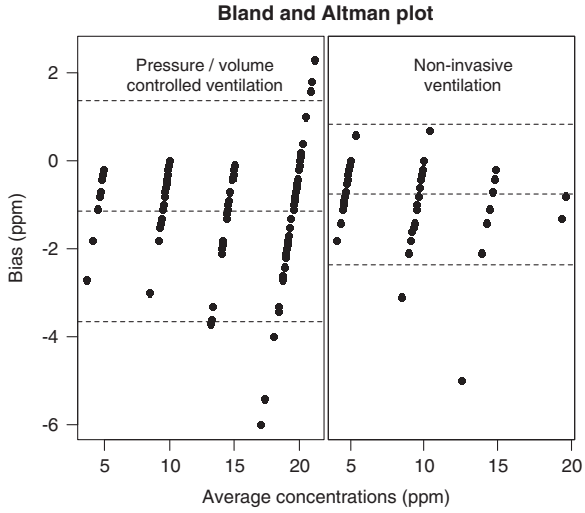


Figure 1. Bias: the difference between the set and delivered concentrations; Average concentration: the mean of the set ad delivered concentrations; in dotted lines the mean and 95%CIs of the difference.

P2-122
Operative outcomes and indication of surgery for primary cardiac tumor in children

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Objectives: To investigate indications and outcomes of surgically managed cardiac tumors in children.
Methods and results: 43 consecutive patients aged < 18years, were retrospectively analyzed from January 1995 to December 2015 in two cardiac centers. The diagnosis was prenatal in 19 patients (44%). For the remaining patients, median age at diagnostic was 89days (0days to 14.9 years). Histologic examination of the tumors showed rhabdomyoma in 16 patients, teratoma in 8, fibroma in 6, myxoma in 6, Inflammatory myofibroblastic tumor in 4, hemangioma in 1, lipoma in 1 and fibrosarcoma in 1. 37 patients had surgical resection of the tumor: 27 complete (73%) and 10 partial resection (27%). Reason for partial resection was infiltration of the tumor in the myocardium in 4 patients, in a cardiac valve in 3, in a coronary artery in 2 and in the sinus node in 1. 16/37 patients had additional procedures during surgery. The 6 remaining patients had conservative surgery without resection (2 implantable pace-maker or defibrillator, 1 Blalock-Taussig shunt, 2 pericardial windows, 1 mitral valvuloplasty). Achievement of preoperative goal regarding primary indication for surgery was 76% for patients with hemodynamic compromise (22/29): intracardiac obstruction in 16/19, pericardial effusion with tamponade in 4/5,

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valve impairment in 2/4 and left ventricular dysfunction in 0/1; and 80% for patients with rhythm or conduction disorder (4/5). Other indications of resection were systemic embolization of the tumor with stroke in one patient, respiratory distress du to bronchial compression in one and prophylactic surgery in 7. Conservative surgery and partial resection were not associated with lower rate of achievement of preoperative goal. Major post-operative adverse events occurred in 6 patients (14%) with 4 patients needing early re-intervention and 2 deaths (4.7%) before discharge. We did not find any clinical or surgical predictive factor of freedom from post-operative death or major adverse event.
Conclusion: Surgical management of cardiac tumor in children is indicated in 80% of the cases for hemodynamic or rhythmic disorders and provides good operative results. Total resection of the tumor is not the only way to achieve therapeutic aim and conservative surgery should be considered when necessary.

P2-123
Ross Procedure in Children and Young Adults: 21 years of follow-up
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Background: The Ross procedure is used to treat aortic valve disease in children. The advantages include autograft growth, long-term durability, and avoidance of anticoagulation. Long-term follow-up of Ross procedure in infancy is limited.
Methods: We performed a retrospective analysis of prospectively collected data in a population of patients younger than 18 years who underwent Ross operation at Policlinico San Donato between January 1994 and December 2015. Echocardiographic, surgical and clinical data, including survival and need for reoperations, were obtained and analyzed.
Results: A total of 136 patients underwent a Ross procedure at a median age of 11 years (range 27 days to 17.9 years). Twenty patients (15%) were infants and pre-school children. The indication for the Ross procedure was aortic valve stenosis, regurgitation or both, which was observed in 34, 55 and 11% of patients respectively. Seventy-three patients (54%) had a bicuspid aortic valve. Seventy-two patients (53%) had previous surgical procedure for associated congenital malformations or interventional procedure on the aortic valve. Fifteen patients (11%) uderwent Ross-Konno procedure to relieve a left ventricular outflow tract obstruction. Concomitant surgical procedures were performed in 10 patients (7%), 5 of whom underwent resection of subaortic membrane. Mean follow-up was 8 years (range 6 days to 20.5 years). There were 3 late deaths and 1 heart transplant. Twenty patients underwent reoperation after the Ross procedure. At 20 years the freedom from right ventricular outflow tract reintervention was 88%, and the freedom from autograft reintervention was 77%. On average, patients were reoperated 7.8 years (range 5 months to 18 years) after the Ross procedure. The most common indications for surgical reintervention were isolated failure of the neo-aorta in 11 cases (55%), regurgitation and/or stenosis of the pulmonary conduit in 11 patients (55%). The surgical technique used is strongly related to the risk of reoperation in the follow-up.
Conclusions: The long-term outcomes of the Ross procedure in children and young adults are favorable. The low incidence of neo-aorta and pulmonary homograft dysfunction at 20 years demonstrate that the Ross procedure is an attractive option for the treatment of aortic valve disease in children and young adults.

P2-124
The impact of aortic arch repair with lower body circulatory arrest on the incidence of acute kidney injury in neonates
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Introduction: Acute kidney injury (AKI), which is associated with increased mortality, is a common complication in children undergoing complex cardiac surgery for congenital heart disease. Aortic arch repair with lower body circulatory arrest in neonatal patients might be associated with a higher risk of AKI after cardiac surgery. The aim of the present study was to assess the incidence of AKI using the Acute Kidney Injury Network (AKIN) system in neonates with complex congenital heart disease who underwent aortic arch surgery with lower body circulatory arrest. We also sought to describe the risk factors for the development of AKI, as well as the association of AKI with the clinical outcomes, including the duration of mechanical ventilation, the length of stay, and in-hospital mortality.
Methods: The cases of 126 neonates who underwent aortic arch repair under cardiopulmonary bypass with lower body circulatory arrest between January 2006 and December 2014 were reviewed. AKI was classified, according to the Acute Kidney Injury Network (AKIN) system, as stage I-III. We analyzed the incidence, clinical outcomes and risk factors for AKI.
Results: Postoperative AKI was observed in 66 patients (52%). The classifications were as follows: stage I, n = 31 (25%); stage II, n = 6 (5%); and stage III, n = 29 (23%); 21.4% of the patients required renal replacement therapy within 3 days after surgery.
The age at the time of surgery, the surgical procedure (palliative or collective), a RACHS-1 score of ≥ 4, the intraoperative use of epinephrine, the duration of CPB and the 75th percentiles of lower body circulatory arrest (odds ratio [OR]: 2.396, 95% confidence interval [CI]: 1.016-5.652) were significantly associated with the development of a stage III AKI. The development of an AKI was associated with a longer duration of mechanical ventilation and a prolonged intensive care unit stay.
Conclusions: Longer durations of lower body circulatory arrest were associated with a high incidence of AKI in neonates who underwent aortic arch repair under cardiopulmonary bypass with lower body circulatory arrest and may be a marker of case complexity.

P2-125
Outcomes of primary surgical repair of Tetralogy of Fallot in patients under 3 months of age
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Introduction: Classical management of symptomatic babies with Tetralogy of Fallot has involved placement of a Blalock-Taussig shunt followed by complete repair. Some have suggested advantages to early complete surgical repair instead of initial palliation. Here, we compare early and mid- term outcomes for babies undergoing very early complete repair of Tetralogy of Fallot.
Methods: Patients undergoing complete surgical repair of Tetralogy of Fallot at our institution from 2005 to 2015 at < 3 months of age were retrospectively reviewed and compared to a control group of older children matched by anatomical diagnosis and outflow tract intervention.
Results: Fifteen index cases (group A) and 15 controls (group B) were indentified. At surgery median age was 42 days and weight 4.2 kg in group A, and 132 days and weight of 6.1 kg in group B. Comorbidities were more common in group A (5/15) than group B (3/15). One patient in each group had a diagnosis of Double Outlet Right Ventricle. None had significant coronary abnormalities. Mean pulmonary valve z-score was -3.1 (95%CI -3.6 to -2.5) in group A, and -2.1 (95%CI -2.6 to -1.6) in group B ($p = 0.01$). In each group, 12 had transannular patches placed.
Peak inotrope score (23.3 vs 12.5, $p < 0.01$) and PICU length of stay (4.6 days vs 2.7 days, $p = 0.01$) were found to be higher in group A, compared to group B. However, bypass and cross clamp times, duration of intubation and total length of stay did not differ. On multivariable analysis operation at < 3 months of age, pulmonary valve z-score or presence of comorbidites were not associated with increased duration of intubation, length of PICU stay or total length of stay. Pulmonary valve z-score and presence of comorbidities, but not age at the time of repair, were found to be independent predictors for increased peak inotrope score.
Conclusions: Very early complete repair of Tetralogy of Fallot is associated with increased initial inotrope requirement and duration of PICU stay. However, mortality, and total duration of admission are comparable to those of older patients. We suggest that early complete surgical repair is an important management option in these patients.

P2-126
Advantages of hybrid management for pulmonary atresia, ventricular septal defect, major aorto-pulmonary collateral arteries and completely absent central pulmonary arteries
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Objectives: To review early and long-term clinical outcomes for pulmonary atresia, ventricular septal defect (PA/VSD), major aorto-pulmonary collateral arteries (MAPCAs) and completely absent central pulmonary arteries (cPAs)
Methods: Of all 120 patients with PA/VSD, MAPCAs who underwent surgical intervention between 1981 and 2011, 15 patients (12.5%) with completely absent cPAs were enrolled. Median age at initial surgery was 1.7 years-old (range, 0.1-17.6). Median number of unifocalized MAPCAs were 3.5 (2-6). Before 2005, various surgical approaches had been applied (Other group, $n = 10$), including staged unifocalizations via thoracotomy followed by RV-PA conduit placement and complete VSD closure in 8 patients, primary definitive repair via median sternotomy in 1, and the other in 1. Since 2003, hybrid management has been applied (Hybrid group, $n = 5$), where complete unifocalization and RV-PA conduit placement via median sternotomy as the 1st surgical palliation, then which was followed by VSD closure with one-way fenestrated patch and conduit replacement about 1 year later. For hybrid group, percutaneous trans-catheter balloon angioplasty (PTA) was aggressively performed after, and also before the 1st surgical palliation. After the definitive repair, stent implantation in addition to PTA was repeated for both group patients, if necessary. Follow-up was completed on all patients and median follow-up period was 7.8 years (0.3-21.7).
Results: Whereas all 5 patients in Hybrid group survived long after the definitive repair, 5 patients died before or early after the definitive repair in Other Group ($p = 0.05$). Uncontrollable bleeding complications were related to death in 4 of all 5 mortalities. Actuarial survival rates at 10 years after the initial surgery were 100% in Hybrid Group and 40.0% in Other group (log-rank, $p = 0.08$). For 10 patients survived long after the definitive repair, catheter intervention for unifocalized MAPCAs were repeated in 6 patients and latest catheter examination at median duration of 4.8 years after the definitive repair showed that RV/LV pressure ratio was maintained from 0.60 (0.45-0.73) at early after the definitive repair to 0.57 (0.35-0.81).
Conclusions: Although statistical significance was not proven, maintaining patency of MAPCAs by hybrid management can be expected to provide better early and long-term clinical outcomes.

P2-127
High values of NT-proBNP after re-surgery is related to decompensation of right ventricle and weakening of left ventricle in repaired patients with tetralogy of Fallot
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Background: On remote time after definitive repair of tetralogy of Fallot (TOF) patients were sometimes redone right ventricle outflow reconstruction (re-RVOTR) because of severe pulmonary valve regurgitation or severe stenosis of RVOT. Some patients were discharged with high NT-proBNP, though they seem to recover from intensive cardiac stress. We investigated what factors before re-RVOTR were connected with high NT-proBNP after re-surgery.
Methods: The medical records of 26 TOF patients were reviewed who underwent re-RVOTR between 2010 and 2015. We performed cardiac catheterization before surgery to judge candidate for re-RVOTR. We defined high NT-proBNP as values of NT-proBNP on discharge after re-RVOTR 400 pg/ml or over ($n = 10$) with which patients had potential to fall into heart failure (Japan heart failure society). Cardiac performances before surgery were compared between patients with or without high NT-proBNP.
Results: There were no differences in RV volume between high NT-proBNP and non-high NT pro-BNP, such as on end-diastole (180 vs. 143 ml/m^2) and on end-systole (118 vs. 76 ml/m^2); no differences in RV pressure, such as on end-diastole (10 vs. 10 mmHg) and RV to LV pressure-ratio on end-systole (0.58 vs. 0.59). In contrast, ejection fraction of RV (RVEF) was significantly decreased in high NT-proBNP group (0.36 vs 0.48); particularly, the ratio of patients who had RVEF under 0.40 was higher in high NT-proBNP (70% vs. 12%, $p = 0.0085$). As LV performances, volumes were significantly larger in high NT-proBNP group, such as on end-diastole (119 vs. 86 ml/m^2) and on end-systole (58 vs. 38 ml/m2), although end-diastolic pressure and EF were almost equal between two groups. Clinically, the ratio of patients with having symptoms before re-RVOTR was not different between two groups.

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Discussion: Our study showed RVEF had declined substantially before re-RVOTR in repaired TOF patients, if patients discharged with high NT-proBNP after re-RVOTR. Likely, compensation of expanded RV had fallen in patients with reduced RVEF. Moreover, patients with high NT-proBNP had suffered LV expansion before re-RVOTR. Not all these degenerations improve on remote period after re-RVOTR. We should perform re-RVOTR before RVEF was reduced too much or LV started to transform with or without symptoms.

P2-128

Predicting fluid responsiveness in cardiac postoperative children: what about electrical cardiometry ?

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Background: Postoperative fluid management is a milestone of post operative period concerning children with congenital heart disease. Electrical Cardiometry (EC), (ICON®, Osypka Medical) is a continuous noninvasive hemodynamic monitor used routinely in our unit.
Objective: This study try to evaluate reliability of one of its parameters, stroke volume variation (SVV), to predict fluid responsiveness of our patients compared to classical parameters.
Design/Methods: Patients were prospectively included in post-operative period. Stroke volume (SV), SVV on EC, cardiac output, central venous pressure, left auricular (LA) pressure, curve variation of invasive blood pressure, central venous pressure, saturation, LA and echography velocity time integral variation were noted. Results are median (interquartile). Responders to volume expansion (VE) had an increase in SV of at least 15% after VE.
Results: 90 patients were included (6.5 months (44), 6.4 kg (9.8)) and 46 had VE. Before VE, SVV (18(9) vs 10(5) $p < 0.0001$) and pulse oximetry curve variation (0.55(0.74) vs 0.03(0.18) $p < 0.0001$) were significantly higher for patients with than without VE. In our cohort SV is significantly higher ($p = 0.0221$) in VE group but not SV weight index as in other team cohorts. Concerning SV difference, it can be explain by patients younger (3 vs 15.5 months) and with significantly low weight (5.2 vs 8.2 kg, $p = 0.0099$) in VE group. But, concerning responders vs non responders groups after VE, SVV is the only parameter which is significantly different with area under curve and cut off (fig. 1) of 0.696 and 19% ($p = 0.015$) versus and 0.809 and 13% ($p < 0.0001$) between patients with or without VE.
All others parameters (clinic, echocardiography or invasive measures) have no significant difference and too low AUC, including delta aortic peak flow velocity on echography and delta invasive blood pressure.
These preliminary results (first ones in electrical cardiometry) confirme first ones on other bioimpedance device on reliability of SVV, which seems to be more practical and reliable than classical and invasive parameters.
Conclusions: Noninvasive measures of SVV and SV using ICON seems to give reliable data to guid fluid management in post-operative period. Of course, these are preliminaries data, and larger cohort is necessary.

P2-129

Cardiac involvement in children with TMEM70 deficiency

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Introduction: TMEM70 deficiency is the most common nuclear-encoded defect affecting the mitochondrial ATP synthase. Cardiac findings typically reveal hypertrophic cardiomyopathy. Congenital heart defects, WPW syndrome and pulmonary hypertension of the newborn with cardiac failure were also reported. Objective of the study was to determine the significance and course of cardiac involvement.
Methods: Retrospective analysis of cardiac findings and clinical outcome in Slovak children with confirmed TMEM70 deficiency.
Results: Sixteen Roma children who all were homozygous for the common mutation c. 317-2 A > G were evaluated. Median gestational age at birth was 37th week (range 31-40) and median birth weight was 2.0 kg (range 1.5-2.8). Neonatal onset of metabolic deterioration was documented in 75% of patients. Eleven (92%) neonates required intubation and 5 (42%) had severe pulmonary hypertension with haemodynamic compromise. Echocardiography revealed two patients with mild valvular aortic stenosis and one neonate with coarctation of aorta. Eleven (69%) children were diagnosed with non-obstructive hypertrophic cardiomyopathy, and in 4 (25%) left ventricular outflow tract obstruction (LVOTO) was documented. Neonatal mortality was 25% with two early deaths and two deaths after initial stabilization. Two children died at age of 3 and 20 months, respectively. Symptomatic neonatal pulmonary hypertension was positively associated with mortality ($p = 0.017$).
Cardiologic follow-up was performed during median of 14.5 months (range 0.1-153). Echocardiography at birth, 3, 6, 12, 24 and 36 months of age revealed median Z–scores of inter-ventricular septal thickness of +10.4, +6.9, +8.3, +5.5, +8.3 and +8.6, respectively and median Z-scores of left ventricular posterior wall of +5.0, +5.2, +7.9, +4.4, +5.2 and +2.6, respectively. Of 4 patients with LVOTO, one neonate died and in 3 children gradient subsided during follow-up. Mild dilation of left ventricle and mild mitral regurgitation developed in 3 and 3 patients, respectively.
One patient had WPW syndrome and in all but 2 infants ECG findings of left ventricular hypertrophy were documented.
Conclusions: Cardiac involvement of patients with TMEM70 deficiency is characterized by infantile non-progressive non-obstructive or regressive obstructive hypertrophic cardiomyopathy. Persistent pulmonary hypertension of the newborn with cardiac failure is common and it is associated with increased mortality.

P2-130

Small intestinal bacterial overgrowth in Fontan patients with protein-losing enteropathy: preliminary results

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Objective: Small intestinal bacterial overgrowth (SIBO) is characterized by excessive proliferation of colonic bacterial species in

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the small bowel. Symptoms related to SIBO are abdominal discomfort, intestinal inflammation, malnutrition, and growth failure, which are also common features of protein-losing enteropathy (PLE) in Fontan patients. The aim of this study was to test Fontan patients with PLE for the presence of SIBO.
Methods: Six Fontan patients (n = 5 chronic relapsing PLE, n = 1 newly diagnosed PLE) were scheduled for a glucose hydrogen breath test (GHBT) between June 2013 and December 2015 at our institution. SIBO was diagnosed if a fasting breath hydrogen concentration was ≥ 10 ppm or an increase in hydrogen levels of ≥ 12 ppm above the baseline value was measured after ingestion of glucose.
Results: One patient was not able to drink the glucose solution and was excluded from the study. The remaining 5 patients (median age 4.9 yrs, 80% male) successfully underwent a GHBT. Median hydrogen concentrations at baseline, and 1 hour, and 2 hours after glucose ingestion were 8 ppm (range 3-31 ppm), 4 ppm (range 1-27 ppm), and 6 ppm (range 5-10 ppm), respectively. Two patients (40%) were diagnosed with SIBO (baseline 16 ppm and 31 ppm). Clinical features and albumin levels improved after increasing the daily steroid dose in patients without SIBO. Interestingly, the patient with chronic PLE and SIBO did not respond to increasing steroid doses. Both patients with SIBO improved after treatment with rifaximin, a non-absorbable antibiotic, and oral probiotics.
Conclusions: SIBO may complicate the course of disease in chronic or newly diagnosed PLE. We identified an additional mechanism that might contribute to long-term outcome in these patients. Further studies are needed to elucidate the clinical role of SIBO in Fontan patients.

P2-131
Investigating the role of altered organ blood flows on enantiomeric disposition of carvedilol in adult and pediatric chronic heart failure patients by using a physiologically based pharmacokinetic approach

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Introduction: Chronic heart failure (CHF) is associated with reduction in blood flow to different organs and has the potential to alter the pharmacokinetics (PK) of the drugs being administered. Carvedilol is a racemic mixture of R and S-enantiomers and shows extensive stereoselective first pass metabolism. The organ blood flow changes occurring in CHF can affect the stereo-selective disposition of carvedilol and may lead to increased incidence of adverse drug reactions.
Methods: Previously quantified organ blood flow reductions (liver, kidney, bone, skin, muscle and adipose) with respect to severity of CHF were incorporated into a whole body physiologically based pharmacokinetic (PBPK) model using population based PBPK simulator, Simcyp®. The developed model was used to predict carvedilol PK in adult CHF patients and after evaluation in adults, it was scaled to pediatric population on physiological basis using pediatric module of the program. The visual predictive checks and ratios(observed/predicted) for the PK parameters with a 2-fold error range were used for model evaluation.
Results: The predictions in healthy adults after intravenous and oral administration of carvedilol were comparable with the observed data. The predictions after incorporation of reduced organ blood flows in adult CHF patients categorized according to New York Heart Association (NYHA) functional classification of heart failure were in close agreement with the observed data and the ratios (observed/predicted) for the PK-parameters were within the 2-fold error range. The predictions in pediatric patients between the age range of 1 month and 14 years, who were classified according to Ross score, showed no improvement with incorporation of reductions in organ blood flows. While, amongst the adolescents above 17 years of age who were classified as adults, according to NYHA system, two out of three patients were better described with incorporation of reduction in organ blood flows.
Conclusion: The incorporation of reduced organ blood flows in the adult CHF patients resulted in significant improvement of the predictions but for the pediatric patients included in model evaluation, improvements were seen only in the adolescent age group and the infants and young children were better described without incorporating organ blood flow reductions.

P2-132
Development of a physiologically based model to support the choice of paediatric enalapril dosing regimen for orodispersible minitablets

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Introduction: Efforts have been increasingly dedicated towards paediatric clinical trials since the EU-paediatric regulation in 2007. The EU's Seventh Framework Programme under grant agreement n°602295 will perform paediatric clinical trials aiming at the safety and the pharmacokinetics of enalapril in children with heart failure using an innovative formulation of orodispersible minitablets. The paediatric committee (PDCO) of the European Medicines Agency required in silico simulations using physiologically based pharmacokinetic (PBPK) models to inform the dosing regimen of these paediatric trials and to be included into the paediatric investigation plan.
Methods: Drug-specific parameters as well as PK experimental data were collected from the literature for model parameterization and evaluation. A coupled PBPK-model of enalapril and its active metabolite was then developed using Simcyp®. The model was evaluated and refined in adults before scaling to children on physiological basis. The ontogeny profile of the main metabolising enzyme, carboxyesterase 1 (CES1), was additionally assigned based on protein expression data. Using the scaled model, drug exposure in different pediatric age groups was predicted and the generated information were used to support the design of the dosing schedule.
Results: Model predictions in adults were comparable with the observed data for both enalapril and enalaprilat, which indicate the model ability to capture the drug PK behavior. The relative precentage error of expoure predictions were < 10%. The collected information of relative CES1 expression at different pediatric ages indicated that CES1 reaches about 50% of the adult activity within the first 6 months of life. The pediatric simulations showed an age dependent differences of drug exposure, with high variability in young children. The data generated were used to derive a proposed dosing regimen for the paediatric clinical trials according to the requirements, which was accepted by the PDCO.
Conclusion: Simulations of drug exposure in virtual subjects delivered valuable information on the expected drug pharmacokinetic behaviour in the paediatric population on the basis of already available data in adults and thus safeguard the designed dosing regimen. PBPK-models can save time, effort, and resources when planning paediatric clinical trials, however, simulation informed decisions should yet be confirmed by experimentally obtained data.

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P2-133

Survival of Children after Weaning from Mechanical Circulatory Support

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Introduction: Since myocardial recovery during ventricular assist device support is infrequently observed in general, clinical data following weaning in children is even more limited at present. Thus, we aimed to evaluate the outcome of all patients weaned from the Berlin Heart EXCOR Pediatric Ventricular Assist Device (VAD) at our institution.
Methods: A total of 125 children received implantation of the Berlin Heart EXCOR pediatric VAD at our institution between 1994 and 2014. The study group consisted of all 20 children (16%), (13 female, 7 male) who have been weaned from VAD support. Event free survival was defined as freedom from death or transplantation.
Seven children suffered from cardiomyopathy, seven presented with myocarditis, further indications included congenital heart disease and status post cardiac transplantation. Fourteen patients received a left ventricular assist device, one patient a right ventricular assist device, and five patients a biventricular assist device. The median age at implantation was 0.7 years (range 0–14), the median duration of support was 32 days (6–124).
Results: At present 16 of the 20 children who could be weaned are alive. One patient who was transplanted two months after weaning died from chronic graft failure. Three children died after 2.5, 3.3, and 15.5 years, respectively. Causes of death included bronchiolitis in one child, two patients died suddenly of unknown cause. The median interval between VAD explantation and follow-up was 9.0 years (range 1–18). The Kaplan-Meier analysis showed an event-free survival rate after 5, 10 and 15 years of 83.7%, respectively. Mean survival was 14.3 years (CI 11.5-17.1). At follow-up, echocardiographic median ejection fraction (EF) was 53.0% (range 28–64). There was no significant difference between the median EF at discharge following weaning and at follow-up ($p = 0.25$).
Conclusions: The survival analysis of our cohort reveals very good outcome in general. Survival after 5, 10 and 15 years was superior in children weaned from VAD support compared to children after orthotopic heart transplantation (ISHLT registry data, 2015). Moreover, cardiac function is remarkably stable over years.

P2-134

Remodeling in Experimental Right Ventricular Volume Overload: a Systematic Review

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Introduction: The improved treatment options for children with congenital heart disease (CHD) leads to a growing group of survivors with longstanding residual lesions. The most prevalent is increased right ventricular (RV) volume load, e.g. after correction for Tetralogy of Fallot. Increased volume loading plays a significant role in the development of RV failure in patients with CHD. It is unclear how to recognize, treat or prevent failure in the volume loaded RV.
Objective: The purpose of this systematic review is to evaluate the pathophysiological and hemodynamic adaptation mechanisms involved in the development of RV failure after volume loading investigated in animal models.
Methods and results: Using a pre-specified search strategy, a priori published on the online platform CAMARADES, we identified 1221 unique citations. 102 were eligible for full text review, after which we identified 18 relevant studies. We found two models for RV volume loading in five species: 1) shunt or 2) regurgitation. After data-extraction and meta-analysis, we confirmed effective volume loading, as RV end diastolic volume/area significantly increased in all studies ($p < 0.001$). Cardiac output (CO) increased significantly only in shunt models (shunt: $p < 0.001$; regurgitation: $p = 0\,737$). Increased RV end diastolic pressure in combination with stable systolic parameters (dP/dt max, TAPSE) suggests diastolic dysfunction. Initial increase of preload recruitable stroke work implies a positive adaptation mechanism of the RV, which declines over time ($p = 0\,012$). Direct measurement of diastolic function is lacking. Patterns in structural and molecular changes show that fibrosis is a long-term effect in the volume loaded RV, in contrast to hypertrophy, which showed to be unrelated to volume load duration. Systematic analysis of other pathways is lacking, thus specific patterns cannot be discerned from the included studies.
Conclusions: This first systematic review of experimental RV volume load showed a consistent pattern of RV dilatation and diastolic dysfunction rather than systolic failure. In the absence of systolic dysfunction, RV dysfunction is most likely due to diastolic failure, which can be related to the development of fibrosis. Standardized volume load quantification and identification of involved molecular pathways is mandatory to refine the characterization of the volume loaded RV.

P2-135

Heart transplantation in infants and children on mechanical ventricular support

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The aim of this study was to analyze the posttransplant outcome of children and infants on ventricular assist device (VAD) successfully bridged to heart transplantation (HT).
Methods: This is a retrospective analysis of demographics, clinical data and short and long-term outcomes of all patients < 18 years of age who underwent VAD support as a bridge to transplantation
Results: 21 patients (8 males) who were placed on VAD for uncontrolled HF, from 2005 to 2015 were included in the study. Mean age at VAD was 5.6 years (median 3y), mean duration of VAD was 30 days, and mean age at HT was 5.8 years (median 2.8y). Two cases experienced stroke while on VAD and 1 severe digestive haemorrhage occurred. Underlying cardiac disease was a dilated cardiomyopathy in 19 and congenital heart disease in 2. Three deaths occurred (14.2%): one early posttransplant from primary graft dysfunction and 2 late at 2 and 8 years posttransplant from severe sepsis. No early acute rejections occurred. One patient had severe late humoral acute rejection and developped donor-specific HLA antibodies. All the other patients have normal graft function and are in NYHA classI at a mean follow up is 4.5 years, range 3 months to 9.5y. One-year, 5 years and 10 years survival rates are respectively 95%, 89% and 65%
Conclusion: Post-transplant outcome of children and infants on VAD support is favourable with very low incidence of infection and acute rejection.

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P2-136

Acute myocarditis in children: single center experience

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The objective of this study was to assess the outcomes of infants with acute myocarditis.
Methods: Patients < 2years diagnosed with acute myocarditis (proved by virology and/or MRI and/ or complete recovery of myocardial function) were included in the study. Clinical data, echocardiographic parameters and outcomes were collected.
Results: 43 patients were included (1983 to 2012), aged 2 months to 2 years at diagnosis (median 6 months). Heart failure was present at onset in 97.6% of the cases, severe in 56% and cardiogenic shock occurred in 14.6%. LVSF at diagnosis was 16.1 ± 5.9% and sub-aortic VTI was 8.1 ± 2.9 cm. Mitral regurgitation was present in 76.5%, pericarditis in 16.4%, thromboembolic in 4.6%, arrhythmias in 14.6%. Virus was positive in 37.2%. Eight patients died (18.6%) within 2 days to 8.6 months after diagnosis, 1 was transplanted (3rd month). Follow-up is 5.5 ± 5.6y (med 4y). Inotrope support was needed in 52% of the cases. Three patients (7%) needed mechanical circulatory support within day-14 from onset. The duration of support was 4d to 3mos. Ten-year survival was 81.4%. LVSF improved from 18.4 ± 8.9% (med 16%) at onset, to 24.6 ± 10.3% (med 23.5%) at 1stmonth, 26.5 ± 8.6% (med 26.5%) at 3rd month, 30.7 ± 8.6% (med 29.6%) at 6th month and 38 ± 7% (med 37%) at last FU.
Conclusion: Acute myocarditis in infants has favourable outcomes despite early mortality. Myocardial contractility can progressively improve within the first 6months after onset of disease.

P2-137

Use of the Heartware® Ventricular Assist Device for Left Ventricular Dysfunction at bodyweight below 30 kilograms

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Introduction: Ventricular assist devices are well-accepted for prolonged mechanical support in children and adults with end-stage heart failure, with smaller devices allowing improved mobility and functional capacity. However, patient size is often a limiting factor. The Heartware® ventricular assist device (VAD) is officially approved for patients above 18 years of age and body surface area of 1.5 m^2, and few cases have been described of its use in children.
Objectives and methods: We aim to describe the feasibility of Heartware® left ventricular (LV) assist device implantation in a child with a bodyweight below 30 kg and to provide a review of the cases available in the literature of the use of this VAD in small children.
Results: We implanted a Heartware® VAD in an eight year old boy with severe aortic insufficiency after neonatal arterial switch for transposition of the great arteries. Attempted aortic valve repair and mechanical valve replacement brought about severe left ventricular dysfunction, requiring mechanical support. Patient weight was 28 kg, height 125 cm, BSA 0.98 m^2, left ventricular diastolic diameter 5.4 cm (z-score +5.5), and mitral to LV apex distance 8.4 cm on echo. The VAD was implanted in the LV apex via median sternotomy along with bioprosthetic replacement of the aortic valve after 5d (days) of ECMO support and 8d on the Levitronix CentriMag® short term VAD. The device was set at 2500 rpm yielding a cardiac output of 2.7 L/min. This allowed weaning of mechanical ventilation (16d), inotropic support (18d), and oxygen (21d), hospital discharge (53d), and return to school (100d), maintaining anticoagulation with oral Warfarin and Aspirin while awaiting heart transplant. Literature review has shown 10 previous children with BSA < 1.0 m^2 in whom the Heartware® VAD has been implanted with no mortality, eight undergoing successful transplant and four being discharged home while listed for transplant.
Conclusions: Heartware® VAD implantation is feasible in small children with a dysfunctional, dilated left ventricle, due to relatively small device size. Its portability allows improved quality of life and return to school for children awaiting heart transplant.



P2-138

Prevalence and Clinical Characteristics of High Cardiac Output Failure in Patients after the Fontan Operation

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Background: Hepatic disease, such as liver cirrhosis, is a post Fontan complication and may reduce systemic artery resistance (Rs) and lead to heart failure (HF) with high cardiac output (HHF).
Methods and Results: This study was to examine the prevalence of HHF post Fontan and clarify its clinical profiles in Fontan patients. We defined HHF based on the hemodynamics of 32 excellent long-term survivors and examined the prevalence throughout a follow up of 389 survivors. We also clarified HHF with clinical variables (study 2). The prevalence from 1 to 25 years after the operation ranged from 2.9 to 8.9%, equivalent to that of 2.9 to 6.9% in patients with low cardiac output (LHF). Of the 33 late deaths, 5 (15%) and 6 (18%) patients died of LHF and HHF, respectively. In study 2, pulmonary arteriovenous fistulae and high hepatic vein wedge pressure (HWP) independently predicted HHF ($p < 0.05$ to 0.0001). Rs was independently determined by hemoglobin levels and HWP, which was independently determined by central venous pressure and γ glutamyltransferase levels ($p < 0.05$ to 0.0001). There was no difference in exercise capacity between the two HF groups. During follow-up, 53 patients experienced clinical events and the hazard ratio of HHF was equivalent to that of LHF ($p = 0.42$) and 3.9 times higher than the non HF groups ($p = 0.0027$).
Conclusions: Prevalence and prognosis of HHF patients were equivalent to those of LHF and the pathophysiology was associated with non cardiac organ abnormalities, including hepatic disease. Hemodynamics guided management strategies, including a focus on Rs, may be mandatory to improve long-term outcome of Fontan survivors.

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P2-139

Diagnostic performance and reference values of novel biomarkers of heart failure in children and adolescents

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Introduction: Biomarkers play a pivotal role in the management of heart failure (HF). In adults, several parameters are known to provide accurate information on diagnosis, prognosis and treatment response. However, reference values and insights from validation studies in adults cannot be extrapolated to the paediatric age group as it differs substantially from the adult population in pathophysiology, comorbidity and compensatory reserve.
Our aim was to assess the diagnostic performance of 3 novel biomarkers in paediatric HF and to elucidate their accuracy differentially in patients with cardiomyopathy (CMP) and congenital heart disease (CHD), respectively. Reference data from healthy children are presented.
Methods: Mid-regional pro Atrial Natriuretic Peptide (MRproANP), sST2 and Growth Differentiating Factor 15 (GDF15) were measured cross sectionally in 111 HF patients and 87 controls (mean age 7.9 ± 5.9 years; range 5 days to 24 years). N terminal pro B Natriuretic Peptide (NTproBNP) was used as a reference standard. A receiver operating characteristic (ROC) was plotted to assess the diagnostic performance of the novel biomarkers alone or in combination with NTproBNP. This was performed in all patients and, separately, for subgroups of patients with CHD (n = 68) and CMP (n = 43). In 38 patients with dilated CMP, left ventricular ejection fraction (LVEF) and diastolic volume were measured by echocardiography or magnetic resonance imaging. Associations with biomarker levels were assessed by logistic regression analysis.
Results: MRproANP and NTproBNP showed good diagnostic accuracy in the general population as well as in the CMP and CHD subgroups. By contrast, GDF15 and sST2 did not perform sufficiently. None of the novel parameters improved the accuracy of NTproBNP alone when combined with it. In the subgroup with dilated CMP, only NTproBNP was associated with an impaired LVEF. Moreover, NTproBNP and sST2 were predictive of left ventricular dilatation.
Conclusions: MRproANP can detect HF with accuracy comparable to that of the reference standard, NTproBNP. This was was the case in, both, CHD and CMP as the causal diagnosis. By contrast, neither sST2 nor GDF15 were of any diagnostic usefulness, alone or in combination with NTproBNP.

P2-140

The Effects of Iron Treatment upon Viscosity for Children with Cyanotic Congenital Heart Disease

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Introductıon: This study was planned to determine the effects iron treatment on viscosity in children with cyanotic congenital heart defect.
Methods: A total of 39 patients with cyanotic congenital heart defect involving 20 girls (51%), 19 boys (49%) were evaluated. Their average ages were 9.9 ± 6.2 years, and average weight was 33 ± 18.4 kg. The patients were categorized into two groups with iron deficiency and without iron deficiency according to their ferritin levels. Iron treatment with two values was applied to the group diagnosed with iron deficiency for three months. Clinical and laboratory findings of both groups were evaluated initially and after three months and their viscosity measurements were carried out.
Results: Iron deficiency was identified in 21(53.8%) out of 39 patients. Average hemoglobin and hematocrit values increased from 14.8 ± 2.4 gr/dl to 16.0 ± 2.0 (p = 0.003), from 45.8 ± 7.5% to 47.6 ± 7.2% (p = 0.052) respectively. Viscosity value, however; decreased from average 5.6 ± 1.0 c poise to 5.5 ± 1.0 c poise by displaying very little reduction (p = 0.741). However, following iron treatment; O_2 sat value increased from average 71.7% to 75% and complaints such as headache, visual haze, frequent sinusitis attacks decreased.
Conclusion: It was observed that iron treatment increased hemoglobin and hematocrit levels without raising viscosity and enabled an amelioration in clinical symptoms of patients with cyanotic congenital heart defect.

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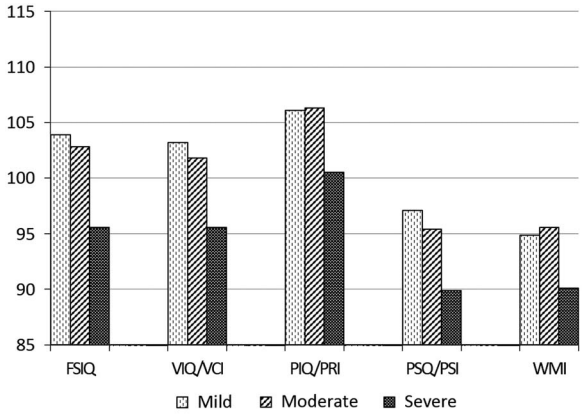
Intellectual functioning in children with congenital heart defects

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Introduction: Studies on children with congenital heart defects (CHD) have indicated that children with mild diagnoses have lower incidence of neurodevelopmental problems than children suffering from severe defects. However, conclusions have been difficult to make since research has been limited to small groups of patients with specific diagnoses or undergoing particular surgical procedures. This study aims to describe and compare intellectual functioning in children with CHD in relation to severity of CHD.
Methods: A total of 531 patients were invited to this study. Families of 235 children agreed (44.3% participation). Participants met a clinical psychologist at their local hospital and underwent a psychological evaluation including testing with the Wechsler scales of intelligence (WPPSI-III or WISC-IV). Participants were divided into three groups depending on severity of diagnosis (Mild, Moderate or Severe). Included in the groups, e.g.; ASD, VSD, PDA and CoA (Mild group); TGA, ToF and AS (Moderate group); SV and PA with VSD (Severe group).
Results: Five analyses of variance were done to assess differences between the severity groups on five intelligence indexes (see figure). For Full Scale IQ (FSIQ), children with Severe CHD had a significantly lower IQ than children with Mild or Moderate severity levels. For Verbal IQ/Verbal Comprehension Index (VIQ/VCI) the Severe group had significantly lower scores than the Mild and Moderate groups. No significant differences in FSIQ or VIQ/VCI were shown between the Mild and Moderate groups. For Performance IQ/Perceptual Reasoning (PIQ/PRI) the Severe group had significantly lower IQ than the Moderate

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group while the Mild group did not differ significantly from the other to. For Processing Speed (PSQ/PSI) the Severe group had significantly lower IQ than the Mild group while the Moderate group did not differ from the other two. For Working Memory (WMI) there were no significant differences between the severity groups.
Conclusions: When looking at differences between the CHD groups, the group Severe stands out as presenting lower levels of intellectual functioning. Although as many as 58.7% of the Severe group have average IQ results, and 10.7% show results clearly above average, there were as many as 30.6% showing results clearly below average.



P2-142
Executive Function Impairment in 108 GUCH Patients
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Introduction: Congenital heart defects (CHD) and repairment procedures create a of risk of cognitive dysfunction. It is still not specified what is the prevalence of certain types of cognitive impairment. In our preliminary study of 45 GUCH patients post-surgery with/without extracorporeal circulation executive dysfunction was the most prevalent. We have therefore assessed the prevalence of executive impairment as compared to other types of cognitive dysfunction in a larger, more diverse GUCH patients group. The objective of the study was neuropsychological assessment of 108 consecutive GUCH patients with CHD leaving paediatric cardiology care after reaching 18 years of age in the years 2011-2014. The material: 108 patients aged x 17.0 ± 19.4 years, 56 M/52 F, after surgical, interventional, pharmacological treatment or no-treatment (14CoAo, 27ASD, 27VSD, 15AVS/AVR, 8PVS, 1Cor Triatr., 4dTGA, 10TOF, 1CAVC, 1 DMGA.DORV.).
Methods: Wechsler Intelligence Scale, Wisconsin Card Sorting Test, Verbal Fluency test, clinical trials assessing memory, attention, praxis, abstract thinking and visuospatial functions.
Results: Among all cognitive disorders frontal impairment was the most common dysfunction in all patients, both as compared to population normative data and to other types of cognitive impairment. Only 9 out of 108 pts (8.3%) achieved normal results on all executive function measures (Wisconsin Card Sorting Test; Formal Verbal Fluency Test; praxis examination; Wechsler subtests: Block Design, Similarities; Rey-Osterrieth Complex Figure Test – copy trial score). Formal verbal fluency generated the lowest scores, with 89,8% scores below normal (97 pts). WCST results were higher, with 31% scores below normal (Total Errors score), difficulties were noted in praxis examination- 61 pts (56.5%), Similarities- 26 pts (24%), and Block Design- 17 pts (15.7%). The distribution of executive disfunction had no significant correlation with the type of CHD, whereas the frequency and intensity of other cognitive dysfunctions (visuospatial abilities, memory, learning) show a tendency to appear more often in conjunction with certain types of CHD (eg. visuospatial/verbal memory dysfunction in 85.4% CoAo pts compared to 60% in other CHDpts).
Conclusions: Regardless of CHD type, complexity, or treatment (surgery, catheterization, pharmacology, no-treatment) executive dysfunction is the most common neurodevelopmental abnormality among GUCH patients, as compared to other cognitive dysfunctions.

P2-143
Mid-term follow-up and quality of life in patients after Fontan surgery
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Background: The Fontan procedure (atriopulmonary Fontan) and total cavo-pulmonary connection are designed to treat univentricular heart. Whereas peri-operative mortality has declined, the current challenge is mid and long-term outcome.
Objectives: To evaluate the outcome and quality of life of survivors with Fontan circulation.
Methods: This retrospective monocentric study aimed patients who had follow-up after Fontan surgery at the University Hospital of Lille. Data were collected on medical records. The quality of life was evaluated between June and October 2014 by two scales: Paediatric Quality of Life Inventory TM (PedsQL) before 26 years of age and Medical Outcome Study Short Form 36 (MOS SF 36) after 26 years.
Results: Among 96 patients who underwent Fontan procedure, median follow-up was 9.6 (6.1-12.5) years after the last intervention. Nine-year global survival was 93%. 95% of patients had total cavo-pulmonary connection and 5% had atrio-pulmonary connection. Arrhythmia occurred in 27.1%, single ventricle dysfunction in 87.4%, leak of the atrio-ventricular valve in 58.9%. Protein-losing enteropathy affected 4.2% of patients and thromboembolic events appeared in 17.7%. Total score of quality of life was 66.5% according to the PedsQL and 62.5% to the MOS SF36.
Conclusion: This French cohort of survivors with Fontan circulation has the same initial characteristics than which described in the literature. The level of quality of life was inferior to general population. The question of global rehabilitation of these patients must be raised.

P2-144
Altered cortisol regulation and reactivity in pre-school aged children who had surgery early in life for congenital heart disease
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Introduction: Early life stress is associated with alterations in the physiological stress response system, and poorer neurodevelopmental and psychosocial outcomes. The Hypothalamic Pituitary Adrenal (HPA) axis, which regulates cortisol secretion, undergoes critical

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development during the first months of life and may be reprogrammed through excessive stress. Cardiac surgery causes significant stress. In this study we compared cortisol diurnal circadian rhythm and cortisol response to a stressor in preschoolers with congenital heart disease (CHD) who had early surgery (before 6 months of age) with those who had no or later surgery.
Methods: Twenty eight preschoolers with CHD (Early surgery = 14, Control = 14) were enrolled. Diurnal cortisol regulation was measured in saliva samples collected from the children at prescribed intervals (30 minutes after waking, 11am, 3 pm and bedtime) over two weekend days. Cortisol response to a stressor was measured in saliva samples collected at commencement of, and 30 and 60 minutes after, routine review echocardiogram. Multilevel analysis (hierarchical linear modelling) was employed to analyse the data.
Results: Diurnal cortisol levels and those in response to a stressor differed between groups. The early surgery group had lower cortisol concentrations throughout the day ($p = 0.03$) and a different slope ($p = 0.002$) compared to the control group. T tests demonstrate that the early surgery group had significantly lower mean waking cortisol levels than controls ($M = 2$ng/mL, $SD = 0.99$ vs $M = 4.35$, $SD = 1.79$, $p = 0.001$) but similar mean bed time values ($M = 0.97$, $SD = 0.7669$ vs $M = 1.192$, $SD = 0.885$, $p = 0.49$) indicating a flatter decline throughout the day. The early surgery group demonstrate a different response to the echocardiogram when compared to controls (group x time interaction, $p = 0.007$). Cortisol levels of the early surgery group increased 30 and 60 minutes post echocardiogram vs. controls who demonstrated the normal recovery pattern of declining levels.
Conclusions: This study provides preliminary evidence that the experience of cardiac surgery in the first 6 months may alter HPA axis, and therefore stress, regulation. Given the importance of stress regulation in neurodevelopmental and psychosocial outcomes, further study may be warranted.

P2-145
Psychosocial needs of children undergoing an invasive procedure for a congenital heart defect and their parents
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Introduction: Up till now, research has focused mainly on psychosocial problems in children with congenital heart disease and their parents, but has largely neglected the need for and implementation of specialized psychosocial care from the parents' and patients' perspectives.
Objective: to investigate the psychosocial needs of both parents of children with congenital heart disease (aged 0-18 years) and patients themselves (aged 8-18 years) in the week before cardiac surgery or a catheter intervention.
Patients: 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 were assessed by a disease specific questionnaire designed for this study, consisting of a 53-item parent version and a 39-item child version (for children >8 years), each covering 5 domains: physical/medical, emotional, social, educational/occupational and health behaviour). 4 items assessed from whom and in what format psychosocial care was preferred. Quality of life was also assessed.
Interventions: if parents/ patients reported a need for psychosocial care, referral to adequate mental health professionals was arranged.
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. Quality of life was relatively high for both parents and patients. Psychosocial care interventions in our hospital increased significantly due to 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.

P2-146
Reference values for children´s pulse wave velocity and central blood pressure using the Mobilograph device
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Objectives: The pulse wave velocity (PWV) and the central systolic blood pressure (cSBP) as indicators for arterial stiffness are of increasing interest for the determination of adults' cardiovascular risk. Although children exhibit a very low absolute risk of experiencing a cardiovascular event, this risk starts developing at a young age. Especially children with congenital heart disease, e.g. after the repair of a coarctation, are subject to an increased risk of developing hypertension and atherosclerosis. Therefore the risk assessment during childhood is an important factor for the prevention of cardiovascular events in later life.
The aim of the study was to establish reference values for PWV and cSBP for children aged between 3 and 5 years.
Methods: The PWV and the cSBP of 128 children (52.3% girls; age 4.77 ± 0.74) were measured with the oscillometric cuff- based mobilograph device (I.E.M. Stolberg, Germany). Reference values and percentiles were calculated with the LMS method.
Results: The mean PWV is 4.32 ± 0.24 m/s and 4.28 ± 0.30 m/s in the male and female subgroup. The cSBP average is 92.15 ± 6.68 mmHg among the boys and 91.67 ± 7.55 mmHg among the girls. Both parameters show no significant differences between the sexes (PWV P■ 0.379; cSBP P■ 0.708).
The mean PWV in 3-3.99-year-old children is 4.23 ± 0.34 m/s, 4.32 ± 0.26 m/s in 4-4.99, and 4.3 ± 0.26 m/s in 5-5.99-year-olds. The cSBP ranges from 90.39 ± 8.91 mmHg (3-3.99 years), 92.7 ± 6.36 mmHg (4-4.99 years) to 91.81 ± 7.01 mmHg for the group of 5-5.99-year-olds. The smoothed percentiles of the reference values are presented based on height and age.
Conclusion: The PWV and cSBP were measured for our population of 3-to 5 year old children with the Mobilograph device in order to define reference percentiles. In particular for children with congenital heart disease and an increased risk for cardiovascular events both parameters are expected to be of great prognostic value.

P2-147
Computer based Digital Phonocardiography Screening for Heart Disease in Childhood
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Objectives: To evaluate whether an automated computer-based algorithm for digital phonocardiogram (PCG) interpretation could detect systolic murmurs and differentiate innocent from abnormal murmurs, thus being capable of supporting large scale screening systems for structural heart disease in childhood.
Methods: 1) Database: Retrospective study including anonymized PCGs obtained from 820 children, (age 1-14yrs) either during school screening program (S.P) or during their visit in a pediatric cardiology outpatient clinic (P.C). In each case 3-5 recordings were obtained from apical, lower and upper left sternal border positions, by using a commercial digital stethoscope, allowing for 3-lead ECG and PCG channel simultaneous recording. PCGs have been off-line labeled by an expert pediatric cardiologist as corresponding to absence of a murmur (A), presence of innocent murmur (I.M) or of abnormal systolic murmur (A.M). All P.C cases and all S.P cases with abnormal murmurs had confirmatory echocardiographic evaluation data, from normal to a wide range of CHD. Recordings with unacceptable noise artifacts were manually removed. 2) Automated PCG analysis: ECG channel R peaks and an envelope-based detection algorithm were used to define the systolic interval. Following band-pass filtering of PCG signals a classification scheme using Support Vector Machines have been used. System training was performed by a dataset of 450 subjects with I.M (n = 329) and A.M (n = 121). Sensitivity (Sens.) and specificity (Spec.) in detecting the presence of a murmur and of an abnormal murmur has been estimated in various scenarios.
Results: 783 cases (95%) with 2677 recordings of acceptable quality have been analyzed, belonging either to A (n = 256), I.M (n = 352) or A.M (n = 175) group. When validating the complete database, the Sens / Spec. of the automated classifier was 93% / 88% to detect the presence of a systolic murmur (against absence of a murmur) and 95%/35% to detect abnormal systolic murmurs (against innocent murmurs). When validating exclusively S.P cases (70 I.M, 7 A.M) the corresponding Sens./Spec. was 84% / 72%.
Conclusions: Automated PCG classifiers could serve as useful means in supporting pediatric cardiac auscultation interpretation. Further software developments and large prospective studies could allow for cost-effective heart disease screening systems in childhood.

P2-148
Cardiovascular Risk in children: outpatient clinics allow early and timely stratification and treatment

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Introduction: The increasing prevalence of cardiovascular risk factors in children needs a structured approach with both early identification and treatment of these patients.
Aim: We aimed at characterizing the methodology and the results in a Paediatric Cardiology centre with a specialized outpatient clinic in this field.
Methodology: Retrospective study was performed from chart review, and classic risk factors, anthropometry, atherogenicity índex and insulin-resistance índex (HOMA-IR) were analysed. Every patient is evaluated clinically and ECG, ecochardiogram and peripheral arterial tonometry (PAT) are routinelly obtained. Patients with suspicion of high blood pressure or association of 2 or more risk factors had additional ambulatory monitoring of blood pressure (AMBP); renal and abdominal ultrasound.
Results: We studied 201 patients, 60% females, mean age 13.9 years (range 6-19) and mean BMI 29.7 (50 patients had BMI > 30). They were referred because of: obesity (63.7%), high blood pressure (53%), hypercholesterolemia (12.4%) and first degree relatives with major cardiovascular events. In the studied population, 2 or more risk factors were prevalent in 42% of patients, 4 patients presented congenital heart defects and 2 Marfan's. The lipid mean values were: total cholesterol: 171.4 ± 38.4 mg/dL; HDLc: 49.2 ± 15.9 mg/dL; LDLc 109.6 ± 35.6 mg/dL and triglycerides: 93.5 ± 62.1 mg/dL. In 32 patients, mean total cholesterol was elevated (233 mg/dL), with high atherogenicity index in 11. Familial Hypercholesterolemia was confirmed in two patients, with mutation of LDLR and suspected in 7. Mean fasting glucose was 97.7 mg/dL, and 27 patients had high levels, but only 4 had Type 1 Diabetes Mellitus. AMBP performed in 73 patients showed high blood pressure in 58 with 19 patients exclusively non dipper. We found endothelial dysfunction in 38 patients with had renal failure and dislipidemia. Pharmacologic treatment was started accordingly to international guidelines, after a period of lifestyle changes.
Conclusions: A structured evaluation of cardiovascular risk factors in paediatric age in a selected population combining several indexes, TAP and AMBP was accurate to evaluate and follow these patients. This methodology also permits selective genetic testing for Familial Hypercholesterolemia allowing identification of 4 cases of FH in 2 families.

P2-149
Endothelial function in children with white coat hypertension

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Introduction: White coat hypertension (WCH) is defined as the observation of high blood pressure (BP) levels in the doctor's office and normal BP during ambulatory monitoring. Endothelial dysfunction is considered to be an early indicator of atherosclerotic changes preceding the morphological alterations, commonly associated with elevated blood pressure. Several studies have demonstrated endothelial dysfunction in patients with essential hypertension. However, the presence of endothelial dysfunction in children with WCH has not been studied. Therefore, we aimed to study the endothelial function in children with WCH and essential hypertension using a novel metod based on assessment of flow mediated dilation (FMD).
Methods: One hundred thirty eight children – 46 (23 boys) children suffering from WCH, 46 (23 boys) patients with essential hypertension and 46 age/gender matched healthy controls - were examined under standard conditions.The age-period of all children ranged from 14 to 18 years. Vascular ultrasound scans were performed with a Prosound F75 Aloka ultrasound machine. Flow increase was induced by inflation of a pneumatic tourniquet to 200 mm Hg for 5 minutes. Diameter of right brachial artery was measured basally and 60 seconds after cuff deflation. Diameter changes were expressed as the percentage change relative to the average baseline scan. Diameter changes < 5% at 60 seconds were considered as a deficient FMD.
Results: Statistical analysis revealed significant differences in the FMD between the both hypertensive groups (WCH, EH) and control group (9% vs 13%; $p < 0.01$). Despite the fact that deficient

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FMD was found in both hypertensive groups (WCH, EH), no significant differences were found between both WCH and essential hypertensive groups. None of the patients in the control group had deficient FMD.
Conclusions: The presence of endothelial dysfunction in children suffering from white coat and essential hypertension suggests that hypertensive children have early atherosclerosis associated with increased cardiovascular risk. Importantly, WCH should not be considered a harmless trait and has common features with essential hypertension.

P2-150
Arterial stiffness evaluated by cardio-ankle vascular index (CAVI) is lower in hypertensive children with overweight
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Introduction: Cardio-ankle vascular index (CAVI) represents a novel marker for early diagnosis of increased arterial stiffness associated with atherosclerosis. However, the studies related to interaction between CAVI and hypertension/overweight in children and adolescents are rare. Previous study found increased arterial stiffness indexed by CAVI at the onset of essential hypertension [1], and another study reported lower CAVI in children with overweight compared with control subjects [2]. Based on these studies, we aimed to evaluate the arterial stiffness using CAVI in children with essential hypertension with normal weight and overweight.
Methods: The studied groups consisted of fifteen normal-weight adolescent boys with newly diagnosed essential hypertension (16.9 ± 1.7 years, body mass index (BMI) $22.3 \pm 1.3\,kg/m^2$), fifteen overweight boys with newly diagnosed essential hypertension (16.9 ± 1.6 years, BMI: $28.0 \pm 1.1\,kg/m2$) and fifteen normal-weight healthy controls (16.9 ± 1.1 years, BMI: $21.5 \pm 1.3\,kg/m^2$). Arterial stiffness indexed by CAVI was examined using the system VaSera 1500 N (Fukuda Denshi, Japan) between 8.00 and 10.00 a.m. under standard conditions after 15 minutes of rest in supine position.
Results: Statistical analysis revealed significantly increased CAVI in normal-weight hypertensive group compared with overweight hypertensive subjects and control group ($p = 0.003$, $p = 0.005$, respectively). No significant difference was found between overweight hypertensive group and healthy controls.
Conclusion: Our findings revealed higher CAVI in normal-weight children with hypertension compared with controls and overweight hypertensive children. In contrast, no significant difference was found between overweight hypertensive group and control children. We suggest that the effect of essential hypertension on arterial stiffness could be masked by the effect of increased weight in hypertensive children indicating potential impact of higher arterial elasticity associated with body composition. This finding is different to studies in adult obese hypertensive patients characterized by higher arterial stiffness; therefore, it could help to elucidate the pathophysiological mechanisms in essential hypertension in children and adolescents.
Support: VEGA 1/0087/14.

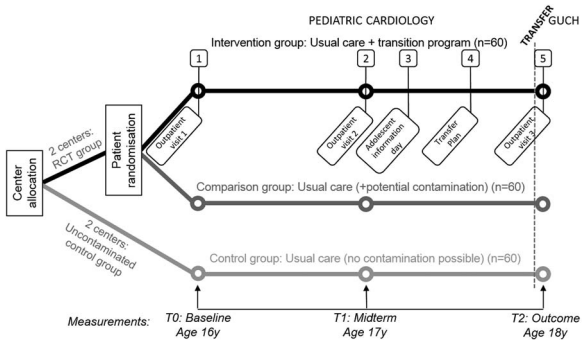
PN-1
Responsibility, knowledge and trust – Important aspects in the preparation to the transfer to adult care
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Introduction: For a majority of children born with Congenital Heart Disease (CHD) the medical and surgical progress has resulted in increased lifetime expectancy. To maximize the potential and lifetime functioning of this growing group of adolescents, lifelong care is needed. In order to ensure a lifetime follow-up, the Paediatric-to-adult transfer of care should be preceded by a preparatory transitional phase for the adolescents. During the transition process the young persons need to learn about their health and step by step take over responsibility for their own health care. However, this transition is also a part of a wider developmental transition process for the young person, which includes occupational choices, personal, family and social aspects.
Objective: The objective was to describe how to prepare adolescents born with CHD before adult care.
Methods: Data was collected through four group interviews. The participants were adolescents between 14 – 18 years old born with CHD from four paediatric cardiology centres in Sweden. The interviews were transcribed verbatim and analysed with content analysis.
Results: Three categories were revealed, Responsibility, Knowledge and Trust. The adolescents emphasised that the responsibility for their health situation had to be shared between themselves, the parents and the paediatric caregivers. Further they wanted to learn about the new adult caregiver, their health, their health situation and how to communicate their health with others. Finally, continuity and a trustful relation to the paediatric health care team were important aspects during the preparation for transfer. The adolescents requested to be included in the transition planning and receive relevant information. Important aspects to consider for the paediatric health care team when giving information and meeting adolescents are maturity and age. These factors are crucial in how adolescents handle different situations related to the transition process.
Conclusion: A trusting relation to the paediatric health care team is a prerequisite for a successful transfer for the adolescents with CHD. Further, maturity and age are other essential aspects to consider when meeting the adolescents need of increased knowledge and the shared responsibility in the transition process.

PN-2
Effectiveness of a transition program to empower adolescents with congenital heart disease in the transition to adulthood: Rationale and methods of the STEPSTONES-ConHD project
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Introduction: Ninety percent of children born with congenital heart disease (ConHD) survive into adulthood. To prepare adolescents with ConHD to take on new responsibilities for their health and to anticipate a transfer to adult care, transition programs have been developed. However, hard evidence on the effectiveness of transition programs is lacking. In STEPSTONES-ConHD (Swedish Transition Effects Project Supporting Teenagers with chrONic mEdical conditionS – Congenital Heart Disease), we test the hypothesis that adolescents with ConHD who receive a structured, person-centered transition program over a 2-year period have a higher patient empowerment score than adolescents who receive usual care. This abstract describes the rationale and design of STEPSTONES-ConHD.
Methods: We employ a hybrid experimental design, in which a randomized controlled trial (RCT) is embedded in a longitudinal, observational study. This design allows to account for potential contamination of the comparison group. The study is conducted in 4 centers in Sweden (Gothenburg, Lund, Stockholm, Umeå). Two centers are assigned to the RCT group and two centers to the control group (see figure).
Literate, Swedish-speaking adolescents with ConHD, aged 16 years, and their parents are eligible for inclusion. A multi-component intervention, comprising 8 key components, is implemented: These components are implemented in five steps (see figure). The intervention is performed by specialized nurses at the outpatient clinic of pediatric cardiology, after being trained to become transition coordinator. Sample size calculation indicated that 60 patients are needed in each arm of the study. Interim analyses will be undertaken during the project, to check if adjustments to the sample size are needed. The primary outcome is Patient Empowerment, as measured with the Gothenburg Young Persons Empowerment Scale (GYPES). The secondary outcomes are transition readiness; knowledge; health behavior; patient-reported health; quality of life; illness perception and parental uncertainty. Data are collected at baseline (T0); midterm (T1); and after transfer (T2). Recruitment of patients starts in May 2016, and will last until the end of 2017. The study will be completed in 2019.
Conclusion: The STEPSTONES-ConHD project is designed to provide evidence on the effectiveness of a transition program for young people with ConHD



PN-3
The Transition Clinic in CHD: an educational project in a Referral National Centre

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Children with a Congenital Heart Disease (CHD), nowadays, may be treated promptly. An increasing number of children born with cardiac anomaly reach adolescence and adulthood as the result of progress that has been made in recent years by cardiac surgery and interventional cardiology. At the same time, also different problems related to this condition are highlighted.
The transition from adolescence to adulthood is a peculiar developmental phase and becomes more extended and less defined. This period is more difficult for adolescents with CHD, because the transition of care does not avoid the complexities associated with this developmental stage. These people need specialized treatment and follow-up, often lifelong. Therefore, a transitional care program should be guaranteed in order to ensure an integrated care. The aim of this program is to let adolescents with CHD responsible for themselves and their health.
In our hospital Phisicians, nurses, psychologists and peer counselors have developed an educational path in order to facilitate the transition. These professionals have designed: informative and training material in order to help young patients and their families in transition; psychological support during all the stages of hospitalization; a communication channel with 'former' patients in order to share their experience. Psychological tests are also used to assess personality and to better understand the impact of the disease on this population. The goal is to optimize the transition's process.
Studies have proved that there is a high risk of discontinuity of care and lost to follow-up of the adolescents with CHD and that families want to be followed and supported during this step of their life. Our project is a model that tries to ensure a suitable and high-quality healthcare for the patient provided by a multidisciplinary team.
The continuity of the follow-up is an important goal of the transition and should be completed even with the transfer, that is, the chance to move adolescents and young adults with CHD from Pediatric Cardiology to an Adult Specialized setting. It is important that transition's project become national, standardized and, therefore, transferable to all national and international Centers who deal with this kind of assistance.

PN-4
A Case Report - Infant Delirium After Cardiac Surgery

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Introduction: Post-operative delirium is a significant but underestimated complication in pediatrics. Impairment in concentration, perception, language, memory and orientation is frequently observed in children as they awaken from anesthesia. Restlessness with inconsolable crying and fiddling is seen in infants. At present, in our cardiac intensive care unit, we do not routinely use any instruments to detect delirium in infants and children nor do we have an interprofessional program for the prevention and treatment of delirium. As a result, there are no structured preventive measures being carried out for the high-risk patients, and delirium often remains unrecognized and inadequately treated.
Method: Based on a literature review and our observations of a 2 ½ year-old girl post cardiac surgery who was treated in our intensive care unit, we identified the main symptoms and triggers of infant delirium.
Results: The literature shows that children and the elderly have an increased risk of delirium. The delirium cascade can be triggered by systemic diseases or iatrogenic interventions. Delirium in children is often associated with accidental extubation and removing catheters, prolonged ventilation time, prolonged hospital stays and can elevate the risk of mortality. Children have delusional memories and experiences of their stay in ICU, which may result in a post-traumatic stress syndrome. We observed restlessness in the 2 ½ year old girl and she stopped verbal communication over several days, only whined and gritted her teeth. Delirium presents a higher risk of long-term damage to the child's perception and development.

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Conclusion: A coherent treatment concept is required in order to improve the early detection and treatment of delirium in children. Delirium presents in a wide variety of symptoms and characteristics depending on the child, his/her age and medical condition. This necessitates the routine use of a sensitive and highly specific instrument by healthcare professionals in order to distinguish delirium from a withdrawal syndrome. Measures such as noise reduction, co-ordination of medical and nursing interventions, involvement of parents in the care of their child, keeping favourite toys close to the child and the use of a primary nursing system positively affect the duration and outcome of delirium.

PN-5
What about life experiences of Congenital Heart Disease adolescents' parents? A literature review and meta-synthesis

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Introduction: Congenital Heart Diseases (CHD) do not preclude the possibility to become adult due to the different innovations in medical and surgical treatments. The transition from childhood to adulthood is a complex process in the lives of all young people, and it is particularly important for CHD adolescents, considering the consequences of their diseases and the need to be adherent with their follow-up indications. In this process, parents play an important role, being a landmark for their children. For this reason, the knowledge of their experiences is important to address a tailored and efficient health-care delivery for the whole family. However, the current scientific literature is mainly focused on quality of life of CHD patients, while the experiences of CHD adolescents' parents are less studied and the literature about their life experiences appears fragmented. Therefore, the aim of this study is to synthesize qualitative papers of life experience of CHD adolescents' parents.
Methods: A review of the literature was performed, considering as main topic the CHD adolescents' parents experiences through a literature search in the last 20 years, in accordance with the PRISMA guidelines and PICOs method. Databases searched included PubMed, CINAHL, PsycINFO and Google Schoolar and keywords used are 'Congenital heart disease', 'Parents', 'Adolescents' and 'life experience'. Only the qualitative papers were included.
Results: The search yielded 405 potentially relevant studies for screening, and only 7 articles met all the inclusion criteria. These papers were analyzed, discussed and a qualitative meta-synthesis was performed, according to the Noblit & Hare methodology. The meta-synthesis results were 4 main themes, exploring 4 main contradictions that characterize the CHD adolescents' parents experiences: 'fear and uncertainty of the future versus positive coping strategies'; 'parents hyper-responsibility and over-protection versus adolescents' independence desire'; 'desire to give support, but not to be supported'; 'normality desire versus awareness to live with particular conditions'.
Conclusions: The role of the CHD adolescents' parents is difficult and they experience some contradictions. This study explore their life experiences in an preliminary way, but further analysis and studies are needed.

PN-6
Brachial venous access for electrophysiological studies in pediatric patients

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Femoral vein is the most used vascular access to perform electrophysiological studies (EPS) to explore conduction tissue. In certain patients, though, femoral access is not possible, such as severely impaired neuromuscular disorders. Adult pneumologists have been using brachial venous access to perform pulmonary hypertension studies, avoiding discomfort and sedation, and short hospital stay.
We aim to analyse the use of brachial venous access to perform EPS in pediatric population whose femoral access is not feasible.
We performed a prospective study of all 14 cases cases that femoral venous access was not possible in paediatric population. In all cases neurological impairment with severe hyperflexion of the pelvic area was the reason for performing an alternative access. Brachial access was obtained using a peripheric catheter and exchanging it by a 5 F introducer. In all cases 5 F tetrapolar stimulation catheter was used. Venous valvules could be passed with the stimulation catheter. In case of sharp angulation, abduction movement of the arm helped to advance the catheter. EPS could be easily performed in all cases either by direct movement of the catheter or whole-circle in the atrium. Hiss analysis was the most difficult to achieve, and the catheter is not made thinking of this special accees, but preceding pre-angulation of the tip of the catheter could help to improve the shape of the angle to obtain nice Hiss signals. Finally, patients could be discharged 1 hour after the procedure with no complications.
Alternative venous accesses as brachial can be easily adapted to pediatric population and used in special cases when femoral access s not feasible.

PN-7
Hybrid Clinic

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Introduction: Given the extensive nature of our area of expertise, the increasing number of patients undergoing Hybrid Procedure for Hypoplastic Heart Syndrome in our Centre and the need of a and the need for careful clinical follow up was created a communication network that allows the parents and pediatrician to immediatly communicate with us through the creation of a 'Hybrid Clinic'.
Methods: The project Hybrid Clinic is composed of four phases consisting in the creation of Hybrid Equipe composed by multi-disciplinar team consisting of a cardiologist, pediatrician, nurse pratictioners;the definition of clinical alarm bells; hospital monitoring with a phase of education and training of the parents performer by the nurse pratictioner and a phase of home monitoring. During the hospital monitoring the parents learn how to check saturation through the use of a dedicated oximeter portable that is assigned for the duration of the monitoring.
Besides the routine outpatient appointment, where we perform the complete cardiac evaluation and overall assessment of the newborn / infant, including nutritional status, anthropometric parameters and assessment of the degree of neurological development we created a dedicated telephone line with a telephone appointment weekly where the parent will confront the health-care workers on the overall condition of the child.
In detail, during the telephone appointment, the nurse pratictioner register on a dedicated database anthropometric parameters, the nutritional capacity and any changes arising in the patient. In case of occurrence of 'alarm bells' we decide the need for specialized assessment. A dedicated mailbox was created and checked daily by dedicated staff to which parents/pediatrician can send updates related to weights, growth curves, blood tests / instrumental performed at home or simple requests for clarification.

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Conclusion: The creation of the Hybrid Clinic improved outcomes related to this already complex pathology leading to the centralization of the patient and the family. We strongly believe that such an approach for complex heart disease represent an example in the clinical scenario of other pediatric cardiac centre.

PN-8

How to teach children a lifetime treatment in only 10 days ?

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Introduction: Heart surgery can, in some cases, lead to a lifetime anticoagulation treatment. In our pediatric cardiac unit, Vitamine K Antagonists (VKAs) are prescribed following cardiac valve replacement and total cavopulmonary connection.
Oral anticoagulant drugs require a steady supervision. International Normalized Ratio (INR) balance is harder to obtain in pediatric population, leading to frequent dose adjustments throughout life. Therefore, a self-monitoring device is available and reimbursed for children since 2009 in France. To be sure children and their family will be able to handle this system at home, we have to teach them how to use the machine and how to live and grow up with this treatment.
Methods: In 2008, a nurse and a cardiologist from our unit followed a specific training which allowed the medical and paramedical team to use this new device and set up an education program.
Our challenge is to educate patients and families during the few days of the hospitalization and to adapt our message to the patient age and comprehension level.
This educational program begins after discharge from ICU. The treatment, its link with the surgery and its surveillance are explained. Two days later, we show the device to the family. The nurse performs the first fingerstick test, then the parents, under nurse supervision, during the next session. The last session focuses on daily life (food, school, sports…). These interviews are evaluated and summarized in a booklet we made for the families. The parents were asked to call us or their cardiologist every time the INR was not in the range.
In 2014, a nurse followed a specific course in educational therapy which led to the implementation of a dedicated patient file for VKA education.
Results: Since 2009, among 187 children (2-18 years) treated by VKAs after surgery in our institution, 160 were eligible for the device. For most patients interviewed, the surveillance was well understood, steady and rigorous.
Conclusion: Our objective is to make the children and their family responsible, autonomous and confident with their treatment. This education is based on mutual trust to make life easier and safe for children under VKAs.

PN-9

Regionalization of a Pediatric Cardiac Department:the experience of Mediterranean Pediatric Cardiologic Centre - Nursing Leadership

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Introduction: Regionalization is a regulatory approach to rationalization of resource allocation, especially for highly specialized medical services or technologies. For high-risk surgical procedures such as cardiac surgery, regionalization may improve outcomes by consolidating surgical programs and increasing the case volume of surgical centers.
Congenital heart surgery is one such highly specialized field. Several studies have shown improved perioperative outcomes for complex congenital heart surgery at centers with greater surgical volumes and that this relation is stronger as the complexity of the surgical procedure increases.
Methods: Sicilian Regionalization of a Pediatric Cardiac Departement was carried out through a partnership between Regional Health Government and Pediatric Hospital 'Bambino Gesù', Joint Commission International. The clinical course of such agreement started in November 2010 at San Vincenzo Hospital of Taormina. The Meditteranean Pediatric Cardiology Center is the only cardiologic tertiary care centre in Sicily and it serves a population of 6 millions inhabitants with about 60,000 newborns per year. Our hub center CCPM is connected to the first and second level centre (spokes) located in Palermo, Catania and Messina.
The highly complex pediatric patients with congenital heart disease require interprofessional teamwork and collaboration to ensure high-quality outcomes with low mortality and morbidity (Congenit Heart Dis. 2013;8:3–19)
Specifically,how nursing and organizational characteristics provide a protective effect on pediatric mortality for congenital heart surgery patients?
Chief nurse works in a peculiar scenario: low level of nurses'of experience in Congenital Heart Surgery, along with elevated turn over of nurse staff operating in the department, also coexistent of private and public in the same structure.
Results: Processes of care included: training, clinical practice guidelines standardized communication, procedural checklists, unit-based medication safety.
The nursing leadership models included a combination of directors, nurse managers, and clinical coordinators.
Conclusion: Howewere the most significant requirement for creation of a cohesive team is in creating a collective sense of responsibility towards the patient.

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